

# Late-Onset Multiple Primary Metastasising Melanomas in a Female Patient with CDKN2A Mutation – Report of an Atypical Case

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## **Introduction & Objectives:**

Frequently denoted as p16 or INK4a/ARF, mutations in the CDKN2A gene are a less-explored aspect within the context of melanoma when compared to BRAF alterations. The CDKN2A gene, functioning as a tumor suppressor, undergoes inactivation through homozygous deletions situated in the 9p21 chromosomal region, that leads to cellular proliferation and disruption of pro-apoptotic pathways. It has been demonstrated that these genetic changes are linked to cases of multiple primary melanomas (MPM) and the maximum number of primaries reported in a patient was 13, with a mean per individual ranging from 2.1 to 2.8.

## **Case Presentation:**

We present the rare case of a female 72-year-old Caucasian patient with a constellation of nine metastasising melanomas distributed across various anatomical sites, posing a diagnostic challenge. Lesions exhibited diverse characteristics, with the initial diagnosis in 2022 revealing multiple ulcerated superficial spreading melanomas [pT3b(m)] in the left mammary, left shoulder, right lateral cervical and left calf cutaneous regions. Immunohistochemistry displayed consistent markers, including positive reactions for S100, PAN MELANOMA, p16, Ki-67, SOX-10, HMB-45, and CD8. By early 2023, the malignancy progressed, manifesting as intradermal and papillary dermal populations with features such as neurotropism and angiotropism, suggestive for multiple low-CSD nodular melanomas in the scalp, left lateral cervical and mandibular area. Notably, lymph node metastases in the right mandibular region were identified, classifying the condition as pT3b N3b. Subsequent assessments in April 2023 revealed the presence of clinically suspicious melanocytic lesions that have been histopathologically diagnosed as intradermal and traumatized junctional nevi, in the right forearm and right calf, respectively. In late 2023, the patient exhibited clinical signs of malignancy in two cutaneous pigmented lesions, that have been excised and confirmed as nodular nevoid low-CSD multiple melanomas located in the right thigh and left foot, as well as two subcutaneous metastases on the anterior thorax (pT2a pM1a). Due to the complexity of the case and the high number of primary melanomas, CDKN2A genetic testing was performed and exhibited the mutated status of the patient, with homozygous deletion. The patient's care plan involves close collaboration between oncologists, dermatologists, and pathologists, to optimize immunotherapy efficacy, while regular dermoscopic follow-ups and imaging studies are crucial for tracking treatment response and adjusting interventions as needed in this intricate and evolving clinical scenario.

## **Conclusion:**

In conclusion, this case report underscores the challenges in diagnosing an elevated number of primary melanomas within a single patient. Emphasizing the significance of immunohistochemistry and, notably, CDKN2A genetic testing, our findings highlight the crucial role of these diagnostic tools in accurately discerning malignant

melanocytic proliferations from nevi and in attaining a proper characterization of MPM cases.



# The Moroccan Hammam: A Purifying Ritual and Its Relationship with Triggered orAggravated Dermatoses: Insights from a Cross-Sectional Study.

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# Introduction & Objectives:

The "hammam" is a bath of humid steam. In Moroccan Araibc, it literally means "hot water" or "hot spring," as it purifies the body and mind. It is also called the Moorish bath and holds significant social importance. It is synonymous with hygiene and beauty.

Due to the popularity of this traditional bath in Morocco, it is important to know the benefits and risks of this ritual on the skin in order to inform and advise the population to prevent any type of skin-related complications.

# **Materials & Methods:**

This is a cross-sectional study conducted within the dermatology department of Mohammed VI University Hospital in Tangier, over a period from March 2023 to July 2023. Using a questionnaire distributed through social networks and aimed at the general population, the data was collected and analyzed on Google Form.

# **Results:**

We received a total of 160 responses. Over half of the participants were women (63.1%). Regarding age, 40.63% were in the 21 to 30-year-old age group. Among the participants, 70% reported a history of dermatosis. The most commonly mentioned types of dermatosis were atopic dermatitis (32, or 18.6%), cutaneous mycoses (31, or 18.2%), cutaneous xerosis (29, or 16.86%), eczema (21, or 12.21%), rosacea (19, or 11%), psoriasis (10, or 6%), and vitiligo (10, or 6%).

A majority of the participants, over 50%, regularly visit the Hammam (more than once a month). On average, they spend about one hour and thirty minutes in the bath. Regarding preferences, 46% of respondents choose to stay in the hot room throughout the session. Moreover, 80% of respondents apply black soap to their skin before the friction procedure, while over 90% use a loofah glove to exfoliate their skin. 75% reported skin irritation after friction.

A significant proportion of the questionnaire participants, exceeding 55%, reported developing a dermatosis after their visit to the Hammam. 56% of respondents reported transient cutaneous xerosis after their bath.

The most frequently mentioned types of dermatosis were cutaneous mycoses (59 individuals, or 42.5%), warts (21 individuals, or 15%), bacterial infections (20 individuals, or 15%), and cholinergic urticaria (8%).

More than half of the surveyed individuals believe that the Hammam worsens their dermatosis. Among them, 30% suffer from rosacea, 25% from psoriasis, 16% from atopic dermatitis, 12% from eczema, 10% from vitiligo, and 7% from cholinergic urticaria. Interestingly, 55% of the participants have consulted a dermatologist regarding their dermatosis.

Regarding post-Hammam care, 80% of the participants use skincare products, including moisturizing creams

(47%), natural oils (12%), lemon (10%), and lightening creams (7%).

# **Conclusion:**

In conclusion, our study highlights the association between visiting the Hammam and the development or worsening of dermatoses in a significant proportion of the questionnaire participants. It is essential to inform patients about the potential risks, especially for those with pre-existing dermatological conditions. Moreover, consulting with a dermatologist is strongly recommended for individuals with pre-existing dermatoses to receive personalized advice and tailored recommendations for each situation with appropriate follow-up.



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## Disseminated Pseudomyogenic Haemangioendothelioma - An unusual case of a rare condition

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## **Introduction & Objectives:**

Pseudomyogenic hemangioendothelioma (PHE) is a rare intermediate-grade (rarely metastasising), vascular neoplasm that most often presents as distinct, solitary nodules on the lower limbs. PHE is a relatively new condition, having only recently been included in the World Health Organisation's (WHO) Classification of Soft Tissue and Bone Tumours in 2013. It more commonly affects young adult males with approximately two-thirds of cases presenting as multifocal tumours that involve different tissue depths in the same anatomic region, such as the dermis, subcutis, muscle and bone. Its presentation can mimic a variety of clinicopathologically similar conditions, one of which is epithelioid sarcoma.

On immunohistochemistry staining, PHE is positive for cytokeratin AE1/AE3, CD31 and ERG but negative for CD34. PHE diagnosis is characterised by FOS-B gene reaarangements in particular (with the partners being SERPINE1 and ACTB).

While PHE can be difficult to distinguish from other high grade malignant tumours, its unique clinical and histopathological appearance can help distinguish it amongst other differentials. It's clinical presentation has been commented on in the literature, however to our knowledge, multuple cutaneous lesions across the body have rarely been described. In this report, we present a rare case of widespread disseminated PHE.

## Materials & Methods (Case):

An otherwise-healthy 23-year-old male presented with an unresolving right arm papule which evolved over one month into multiple widespread, intermittently painful, erythematous papules, nodules and plaques with associated myalgia but no other systemic symptoms. On examination, these were distributed in the upper and lower limbs, hands, feet, trunk, neck and face. PET/CT scan confirmed widespread hypermetabolic cutaneous and subcutaneous lesions as well as multiple intramuscular lesions, and multiple destructive lytic lesions in the skull, axial and appendicular skeleton. MRI brain demonstrated a large lytic frontal bone lesion extending into the dura. There were no solid organ or lung lesions.

Histology from right arm and right knee skin punch biopsies showed an infiltrating undifferentiated epithelioid cell tumour. Immunohistochemistry demonstrated vascular differentiation with a pattern consistent with PHE. Staining was positive for ERG and CD34. Diagnosis was confirmed by next generation sequencing molecular study identifying ACTB-FOSB fusion.

There is currently no established standard of care for PHE. For multifocal involvement, systemic therapies have been described including cytotoxic chemotherapy, bone antiresorptive agents, mammalian target of rapamycin (mTOR) inhibitors, tyrosine kinase inhibitors (TKIs) and vascular endothelial growth factor (VEGF) inhibitors. Our patient was treated with combination sirolimus and zoledronic acid with resolution of symptoms and complete regression of several lesions, but ongoing fluctuating PET hypermetabolism in most lesions elsewhere.

## **Conclusion:**

PHE is a uncommon vascular tumour that most often presents as multiple discontiguous nodules in the same anatomical region, most often on the legs of young adult men. This a rare case of widespread disseminated PHE with minimal symptomatology on mTOR inhibitor and bone antiresorptive agent, which should be distinguished from high-grade malignancy.



# Subcutaneous panniculitis-like T-cell lymphoma and lupus erythematosus profundus: a diagnostic dilemma

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# **Introduction & Objectives:**

A white caucasian woman in her 30's presented with a 1-year history of a tender, indurated lesion on her right upper arm with no history of trauma or exposure to new medication. She was known to have Sturge-Weber syndrome with associated epilepsy, significant left-sided weakness and cutaneous changes throughout her right. Physical examination revealed an irregular indurated plaque on her posterior right upper arm. This measured 13x13cm with an area of hypopigmentation superiorly, superimposed over her existing capillary malformation.

# Materials & Methods:

Laboratory tests at presentation, including rheumatoid factor, anti-Sm, Lyme serology, erythrocyte sedimentation rate, and mitochondrial antibody, were negative. However, a positive antinuclear antibody (ANA) titre was found (1:320) at the time of the first incisional biopsy, and this remained weakly positive (1:80) at the time of the second incisional biopsy.

The lesion regressed spontaneously in the subsequent 5 months followed by the development of a new area of induration anteriorly. An incisional biopsy was performed and a diagnosis of subcutaneous panniculitis-like T-cell lymphoma (SCPTL) rendered. Treatment was with oral prednisolone followed by locally directed radiotherapy with only partial effect. A second incisional biopsy was taken from a persistent area in the right upper deltoid area.

## **Results:**

Following initial diagnosis of SPTCL, the patient was treated with oral steroids. She received 40mg Prednisolone for 4 weeks followed by reducing regime over the subsequent 2 months, reaching 5mg per day on which she was maintained. At this point, 12Gy radiotherapy given in three factions was initiated to treat residual area of disease over the right deltoid. The second biopsy was performed of a persisting area of disease. The patient undergone further radiotherapy, designed to deliver 20Gy over five fractions.

The second biopsy showed changes consistent with lupus erythematosus profundus (LEP) with concomitant microscopic foci of SPTCL. The rationale for continuing to treat as SPTCL rather than lupus with a second course of radiotherapy was based on regional multi-disciplinary team (MDT) discussion, taking account of patient's choice and the ambiguity of treatment response in a dual pathology.

The patient was very reluctant to consider further alternative treatment due to her poorly controlled epilepsy and hence, a further round of radiotherapy was deemed appropriate. Subsequent to the second course of radiotherapy, there has been further softening and improvement of pain in the arm. This patient is continually reviewed in clinic and hydroxychloroquine has been considered as an alternative treatment if disease persists.

## **Conclusion:**

This patient highlights the importance of considering both possibilities. In a patient presenting with SPTCL, an

associated autoimmune disease should also always be considered and appropriate serology undertaken. When LEP is the dominant histological appearance, the pathologist should look carefully for tiny foci suggestive of SPTCL, particularly as the latter disease may be associated with subsequent development of HLH.



# Cutaneous vasculitis as a COVID-19 manifestation: A cross-sectional study with detailed histopathological evaluation

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# Introduction & Objectives:

Although coronavirus disease 2019 (COVID-19) primarily affects the lungs, dermatologic lesions can present as one of the extrapulmonary manifestations. Multiple cutaneous manifestations have been linked with COVID-19, including urticaria, maculopapular rash, vesicular rash, petechiae, perniosis, livedo racemosa, and distal ischemia and necrosis. Cutaneous vasculitis can also occur, ranging in severity from mild or asymptomatic to fulminant disease.

# Materials & Methods:

This study examines COVID-19-associated cutaneous vasculitis/vasculopathy as a defining feature of a virusinduced cutaneous disease, presenting its detailed histopathological studies and clinical follow-ups. Also, we report the impact and efficacy of the different medications used. Patients who presented with typical cutaneous vasculitic manifestations were selected. COVID-19 was confirmed by RT-PCR for SARS-CoV-2 from a nasopharyngeal or throat swab. Skin biopsy specimens were done for 33 eligible patients and were reviewed by an experienced dermatopathologist.

## **Results:**

The most prominent histopathological features for cutaneous vasculitis rashes associated with COVID-19 included endothelial cell swelling, erythrocyte extravasation, vascular ectasia, and a lymphocytic infiltrate with some neutrophils and eosinophils. There was a rapid improvement in patients' lesions upon initiation of prednisolone, which was used as a short-term treatment.

## **Conclusion:**

This study will empower dermatologists with tools for rapid evaluation of patients suspected of cutaneous vasculitis according to clinical presentations and specific histopathological manifestations specific to COVID-19, which emerged recently.



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## Abstract N°: 1846

### Clinico- histopathological co-relation of erythroderma

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**Introduction & Objectives:** Erythroderma is a severe dermatologic manifestation of a variety of diseases and is characterized by an extreme state of skin dysmetabolism affecting more than 90% of the body surface. Incidence was found to be 35/100,000 in an Indian study. It is a diagnostic challenge as it is multifactorial. Histopathology plays an important role in establishing cause, allowing initiation of appropriate treatment. Objective was to study morphological presentation and histopathological findings in erythroderma due to all causes, in all ages and both sexes and to study clinico -pathological co-relation.

**Materials & Methods:** Retrospective and prospective data of patients with clinical diagnosis of erythroderma were recorded. Epidemiological data, clinical and histopathological features were studied. Clinic- histopathological co-relation was studied.

**Results:** Total of 18 patients were included, mean age was 52 years. Females (66.6%) outnumbered males (33.4%). Mean duration of illness was >20 days in 61.1%.\*\* Maximum patients (>50%) had gradual onset. Commonest complaint was scaling (88.8%). Fever was present in 16.7%. Pedal edema was seen in 22.23%. Nail, hair and eye changes were seen in 22.2%, 16.7%, 5.55% respectively. Most common precipitating factor was drug (50.0%) and pre-existing dermatosis, which were psoriasis, dermatophytosis, eczema seen in 27.8%, 5.55%, 5.55% respectively. Past history of erythroderma was present in 4 patients (22.23%). Scalp and body hair loss was seen in 5.55% and 16.7%. Nail changes like ebonisation, Beau's lines, thickening, dystrophy, nail colour changes, clubbing were seen in 5 patients (27.78%). Palmoplantar involvement was seen in 4 patients (22.2%). Clinical diagnosis was erythroderma secondary to drug (44.44%), psoriasis (27.77%), eczema (11.11%), dermatophytosis (5.55%), and others (11.11%). Drugs implicated to cause erythroderma were AKT (3 cases), carbamazepine (1 cases), methotrexate (1 case) and unknown drug (3cases). Histopathological findings included hyperkeratosis (44.44%), parakeratosis (44.44%), irregular psoriasiform hyperplasia (72.2%), regular psoriasiform hyperplasia (11.11%), hypogranulosis (44.44%), hypergranulosis (11.11%), necrotic keratinocytes (38.89%), spongiosis (44.44%), vacuolar degeneration (27.78%) and perivascular infiltrate (55.56%). Histopathological diagnosis was erythroderma secondary to drugs in 8 (44.44%), psoriasis in 7 (38.89%), idiopathic in 2 (11.11%), subacute LE in 1(5.55%).Out of 18 cases, clinico-histopathologic co-relation was obtained in 66.67% cases of erythroderma secondary to drug and psoriasis.

**Conclusion:** A skin biopsy is the only relevant investigation as histopathological features of underlying disorders are recognizable in more than half of the cases. A comprehensive clinicopathological correlation is of substantial importance to render diagnosis of the cause.



## basaloid follicular hamartoma

Rasha Genedy\*1

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# Introduction & Objectives:

Multiple generalized papules in a child could be distressing to the parents. Here I present

A four-year-old boy, born to non-consanguineous parents, presented with a four-month history of numerous brown papules resembling nevi or skin tags. Lesions initially appeared over the trunk and gradually spread to involve the extremities, genitalia and face. Lesions were not pruritic nor painful. He had no family history of similar lesions.

## Materials & Methods:

Clinical examination revealed numerous discrete non-tender tan to brown 1-2 cm papules (Figure1). Palmoplantar pitting was also noted (Figure 2). Hair, nails, and teeth appeared normal. Excisional biopsy was performed on two papules.

## **Results:**

Histopathology of both lesions showed similar findings, which were bland basaloid proliferation arranged in anastomosing columns and cords with cellular stroma containing spindle shaped cells, and peripheral palisading without clefting. Melanin deposition was noted within the basaloid proliferation, but no mitosis. (Figure 3). Routine investigations were normal. MRI brain and ultrasound abdomen were done to exclude other associations and they were unremarkable.

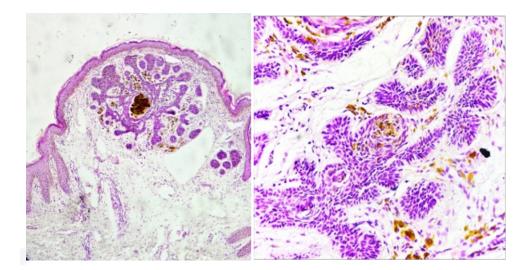
Histopathological features are consistent with basaloid follicular hamartoma (BFH). Given the number of the lesions, palmoplantar pitting and age of the patient, a diagnosis of generalized basaloid follicular hamartoma syndrome was established.

Generalized basaloid follicular hamartoma syndrome (BFHS), a rarely reported autosomal-dominant disorder, manifests at birth or in childhood with numerous skin-colored to brown papules. Other clinical features include palmoplantar pitting, hypotrichosis, alopecia, and hypohidrosis. Development of BFHS is associated with a patch (PTCH1) gene mutation on chromosome 9q23.

# **Conclusion:**

Multiple BFHs can also be part of nevoid basal cell carcinoma syndrome (NBCCS) rendering the distinction between the two syndromes challenging.8, 15, 19 Given the shared genetic profile, both BFHS and NBCCS are believed to exist as a disease spectrum.18 The diagnosis of NBCCS relies on a set of diagnostic criteria that includes bone and neurological manifestations, not present in BFHS.

Lesions of BFH are of substantial importance because of their clinical and histopathological similarity to infundibulocystic BCC. Correct diagnosis is crucial to avoid unneeded surgical procedures.20 Patients with BHFS should be monitored for malignant transformation into BCC as they share the same gene mutation of NBCCS.7, 15





# Strange growths in even stranger places: A rare case of Eccrine Poroma located on the scalp of a 64-yearold male

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# Introduction & Objectives:

Poroma is a rare benign adnexal tumor of epithelial cells, with an incidence of only 0.001 to 0.008% in the general population. This tumor generally occurs in middle-aged individuals and is mostly found on the sole or lateral surfaces of the foot, hands, and hair-bearing areas. However, a poroma on the scalp is extremely rare, with only 18 reported cases worldwide as of writing.

# Materials & Methods: N/A

## **Results:**

A 64-year-old male sought consult at the out-patient department of a tertiary hospital due to the progression in size of a solitary irregularly-shaped pedunculated tumor located on the vertex of the scalp, Histologic findings were consistent with Poroma. The condition was addressed by excision with clear margins.

# **Conclusion:**

Because of the rarity of Poromas, it is indeed challenging to diagnose based on clinical presentation alone, as these lesion are often mistaken for other benign neoplasms especially if the lesions present in an uncommon location. Considering the lack of established specific dermoscopic criteria, and that 18% of poromas show malignant transformation into porocarcinoma15, it is imperative that other diagnostic methods such as dermoscopy and histopathological correlation be performed in order to confirm the diagnosis



# "Clinico-dermoscopic and pathological spectrum of lichen planus cheilitis from a tertiary care hospital of eastern India- A cross-sectional study"

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# Introduction & Objectives:

Lip lesions are a transitional condition between the oral and cutaneous variants, requiring their own categorization. Even though cheilitis is a commonly encountered clinical entity, it is still challenging to diagnose and frequently misinterpreted due to the morphological resemblance between various types of cheilitis. Lichen planus (LP) cheilitis is a relatively rare and less explored condition.

# Objective:

To describe the clinico-histopathological and dermoscopic spectrum of LP cheilitis from a tertiary care hospital of Eastern India.

# Materials & Methods:

A cross-sectional, observational study was conducted. 42 patients with LP cheilitis, irrespective of age and gender, were enrolled. A detailed history and clinical photos were taken. 3-4mm punch biopsy for histopathological examination was done. Clinical and dermoscopic images were captured using a cannon SX 620 HS camera and Heine Delta 30.

## **Results:**

**Among patients attending dermatology OPD, 42 had cheilitis secondary to LP out of 384 cheilitis cases.** The age varied from 13 to 72 years (mean 40 years) and male to female ratio of 1:1. The duration varied from 20 days to 12 years (mean 6 months).

The majority of patients complained of burning 64%, ulceration 12%, and the rest were asymptomatic.

The involvement of the isolated lower lip (71.4%) was relatively more frequent than both lips, among which vermilion was solely affected in 33 (78.5%). Plaque, annular, annular atrophic, ulcerative plaque, erosive and papule subtype were the morphologies observed in decreasing order of frequency. Colour of the lesions varied from reddish white and reddish grey. Vermilion border was involved in 26.1% patients, while none had obliteration. Clinically appreciable wickam striae were further subcategorized as radial and interlacing white lines.

Upon dermoscopy, the brown-grey dots and globules were arranged in unspecific, diffuse, linear, periphery and clustered patterns whereas white dots and globules in the unspecified pattern was seen in 5 cases. Perifollicular pigmentation was seen in 11.9%. Wickham's striae was noted in 90.4%. Apart from the classically described reticulated white lines, we noted the leaf-like (30,71.4%), leaf venation (14,33.3%) and interlacing pattern (9,21.4%). Starburst pattern was seen in 3 cases. Comedo-like openings were found in 11.9%.

Histopathology showed predominant lichenoid reaction pattern (21,84%) and less commonly vacuolar reaction pattern (4,16%) with hyperkeratosis, wedge-shaped hypergranulosis, basal vacuolar degeneration, dyskeratotic kertainocytes, and band-like subepidermal lichenoid infiltrate. Melanin incontinence was also observed.

# **Conclusion:**

Diagnosing LP cheilitis is very challenging in skin of colour as most of the mucosal dermatoses heal with slaty grey to blue-grey pigmentation. This is the largest study focusing on LP cheilitis and characterizing its' clinicodermoscopic and pathological features. Plaque and erosive variant are the most common. The additional clinical clues are the radial white and pigment lines, isolated lower lip involvement, and non-blurring/obliteration of the vermilion border. Large comparison research, including people with skin of colour and fair skin in the future, will provide more hints for detecting LP cheilitis and differentiating it from its clinical imitators.



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# Abstract N°: 2135

## Title: Transillumination test in the diagnosis of a lobulated swelling on the left axilla

Sonikaa Garg , Shreya Kempegowda , Biswanath Behera , Pavitra Aryan , Tara Prasad

# Title: Transillumination test in the diagnosis of a lobulated swelling on the left axilla

## **Introduction & Objectives:**

Diagnosing adnexal tumors is challenging due to the sharing of features clinically and histologically. In addition, the benign one can progress to its malignant counterpart. Furthermore, most of them are located in the head and neck area. An accurate diagnosis requires recognition of the cell of origin, then looking for evidence of malignancy, and finally evaluating whether the lesion is primary or metastatic. Primary cutaneous mucinous carcinoma (PCMC) is a rare cutaneous malignancy of sweat gland origin. PCMC is due to the proliferation of atypical apocrine or eccrine ductal cells and mucin overproduction, leading to islands of tumor cells floating in a sea of mucin.

# Materials & Methods:

A 54-year-old otherwise healthy male presented with an asymptomatic swelling on the left axilla for five years. It had an insidious onset, was slowly progressive, and was not associated with any constitutional symptoms. His family history was unremarkable. The patient complained of occasional mild bleeding from the nodule. Examination revealed a reddish-grey polylobulated mass of size 6cm X 3cm with areas of superficial erosion on the left axilla, giving the appearance of a custard apple.

On palpation, it was a minimally indurated, non-tender, and firm nodule that did not bleed on touch and was not fixed to underlying structures. No lymphadenopathy was noted. The possibilities of apocrine carcinoma, hidradenocarcinoma, and metastasis from carcinoma breast were considered

## **Results:**

Before doing a biopsy, out of curiosity, a transillumination test was performed, which yielded a positive result. Histopathology revealed epidermal acanthosis, and the dermis showed basaloid lobules floating in a pool of mucin. The tumor cells were round to oval with moderate eosinophilic cytoplasm, oval nuclei with coarse nuclear chromatin, and inconspicuous nucleoli suggestive of mucinous carcinoma. Immunohistochemistry (IHC) demonstrated cytokeratin 5/6-, 7+, 20-, ER+, PR+, Her2/neu-, AR+, and GCDFP+. Contrast-enhanced computed tomography of the head, neck, chest, abdomen, and pelvis was considered in view of the poor uptake of mucinous tumors for fluorodeoxyglucose and did not reveal any abnormality. Mammogram, upper gastrointestinal endoscopy, and colonoscopy yield negative results. A final diagnosis of primary cutaneous mucinous carcinoma was made. Considering the potential risk of recurrence, the patient was planned for neoadjuvant chemotherapy with bleomycin, etoposide, and cisplatin, followed by wide local excision.

## **Conclusion:**

To conclude, we report this very rare case of PCMC to highlight its peculiar clinical morphology, besides stressing the transillumination test that aids in suspecting the diagnosis clinically.



## Tattoo-associated lymphomas: myth or reality?

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#### **Case presentation:**

A 67-year-old woman consulted for the appearance of a raised plaque over a red ink tattoo. The tattoo was located on the outside of the right arm and had been made 7 years ago, without skin alterations or discomfort until one year ago. A skin biopsy was performed for microbiological (negative for bacteria, fungi and mycobacteria) and histological study. A dense monomorphous band-like inflammatory infiltrate was observed leaving a Grenz zone, composed of small-medium sized lymphocytes with presence of macrophages and red pigmented histiocytes. Immunohistochemistry showed abundant T-cellularity (CD3, CD8, CD5, CD7 and CD4 positive) with nodular areas corresponding to B lymphocytes (CD20 and CD79 positive). All nodules were positive for BCL2 and negative for BCL6 and CD10, CD5 and CD43. The Ki67 proliferative index was between 5 and 10%. This lesion was therefore compatible with a primary cutaneous T-cell-rich marginal zone B-cell lymphoma. A complete excision of the lesion (free margins) was performed, together with an extension study with abdominopelvic CT scan (no data of extracutaneous involvement), blood tests (no elevation of  $\beta$ 2-microblobulin or other significant alterations) and epicutaneous patch tests (Spanish standard battery and specific ink study, both negative).

#### **Discussion:**

The possibility of the development of pseudolymphomas or cutaneous lymphoid hyperplasia is a well-known complication of tattooing, especially related to the use of red ink. Instances of progression from pseudolymphoma to lymphoma, as well as the emergence of cutaneous lymphomas due to prolonged exposure to external antigens, have been documented.1. Therefore, the development of a cutaneous lymphoma secondary to the components of a tattoo seems plausible. However, documented occurrences of cutaneous lymphomas specifically on tattoos are limited in the literature, with only two reported cases: a primary cutaneous centrofollicular lymphoma2 and a cutaneous large B-cell lymphoma, which appeared after transformation of a pseudolymphoma3. Other previously reported cases were eventually classified as pseudolymphomas. Moreover, a population-based case-control study found no significant association between the development of non-Hodgkin's lymphoma and a history of tattooing.

## **Conclusion:**

The emergence of a cutaneous lymphoma over a tattooed area might be linked to the tattoo itself, resulting from chronic lymphoid stimulation and the potential formation of a clonal population. Nevertheless, the limited number of reported cases raises the possibility that its occurrence in this specific area could be coincidental, unrelated to the substances used in the tattooing process.



**AMSTERDAM** 25-28 SEPTEMBER 2024 EUROPEAN ACADEMY OF DERMATOLOGY & VENEREOLOGY

# Abstract N°: 2200

## Therapeutic and diagnostic challenge in a rare Immunoglobulin A type of Epidermolysis Bullosa Acquisita

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## Therapeutic and diagnostic challenge in a rare Immunoglobulin A type of Epidermolysis Bullosa Acquisita

### Introduction:

Epidermolysis bullosa acquisita (EBA) is a rare autoimmune subepidermal blistering disorder that classically presents as vesicle and tense bullae over trauma-prone sites. It is broadly categorized as mechanobullous type, bullous pemphigoid type, linear IgA bullous disease (LABD) type, and mucous membrane pemphigoid type. We hereby present a rare subtype of LABD type of EBA type which pose both diagnostic and therapeutic challenges.

## **Case report:**

A 22-year-old female presented with mild pruritic, multiple, erythematous edematous plaques with clear fluidfilled tense blisters which ruptured to form raw areas over the forehead and nape of the neck for the past 3 months. No history of photosensitivity, oral ulcers, recurrent fever, joint pain, or blisters at the site of trauma. These lesions subside with 30mg of prednisolone over three to four weeks. Examination revealed various stages of evolution ranging from multiple grouped tiny clear to hemorrhagic fluid-filled vesicles, tense bullae, and hemorrhagic crusting with areas of healing in the form of hypopigmentation without scarring or milia formation. The rest of the mucosal, nail, and systemic examinations were normal. Nikolsky sign and skin fragility tests were negative. Possibilities of LABD, and bullous systemic lupus erythematosus (BSLE) were considered.

Skin biopsy from vesicle revealed subepidermal cleavage with predominant neutrophilic and few eosinophilic inflammatory infiltrates. Direct immunofluorescence showed a linear, u-serrated pattern of immunofluorescence with IgA (3+), and Ig G (2+) deposit at the basement membrane. Salt split indirect immunofluorescence demonstrated floor pattern of IgA, and IgG. Antinuclear pattern antibody (ANA) and ANA profile were negative. Clinical and laboratory findings led to the diagnosis of IgA EBA. The patient was started initially on dapsone and doxycycline, later she developed anemia and transaminitis, hence the drug was withdrawn. Then she was administered tapering doses of prednisolone, methotrexate 20 mg per week, and apremilast 30 mg twice daily for 4 months, despite this she continued to develop 4 to 5 vesicles per day over face and chest. We administered her two cycles of rituximab 1 gram, 2 weeks apart, and was continued on prednisolone 20 mg and methotrexate 20 mg/week. Extensive search for malignancy yield negative results. Serum IgA and IgG were normal. Partial remission was achieved despite combination therapy.

## **Conclusion:**

To the best of our knowledge, there are 90 reports of this variant of EBA. Therapeutic options include oral and injectable steroids, dapsone, methotrexate, mycophenolate mofetil, rituximab, and IVIG. Most of the patients are therapy-resistant and need complex therapy.7 Extensive evaluation is required to diagnose as higher immunosuppression is required for disease control. Table 1 enumerates the therapy-resistant cases. To conclude our case highlights the challenges while treating IgA EBA, and combination therapy showed a partial remission and represents the unusual location of the disease, absent skin fragility, and rarity of the disease.

| Fluid filled tense<br>vesicles<br>Photosensitivity | Clinical tense blister+milia<br>HPE: Subepidermal split with neutro<br>DIF: u serrated linear IgA(3+),IgG(2+)<br>Salt split: floor pattern<br>ANA (IFA): Negative |                   | New lesions over face,<br>trunk and chest while |  |
|--|---|-------------------|---|--|
| Possibility of LABD<br>Bullous SLE                 | Diagnosis of LABD type of EBA   | tapered to 10mg   |   |  |
|  | 1   |                   | 1   | 1  |
| Aug 2023   | Sept 2023   | Jan 2023          |   | Feb 2023                                 |
|  | Prednisolone 40mg tapering<br>10mg every 4 weekly   | Prednisolone 20mg |   | Prednisolone 10mg                        |
| Dapsone 100mg HS                                   | Dapsone withdrawn i/v/o<br>hemolysis, transaminitis   |                   |   |  |
| Doxycycline 100mg OD                               | Doxycycline stopped   |                   |   |  |
|  | Methotrexate 20mg/week  | Meth              | otrexate 20mg/week                              | Methotrexate 20mg/week                   |
|  | Apremilast 30mg BD  | Aprer             | milast 30mg BD                                  | Apremilast 30mg withdrawn                |
|  |   | Inj Rit           | tuximab 1g IV 1st dose                          | Inj Rituximab 1g IV 2 <sup>nd</sup> dose |

Summary of therapeutic options considered in the patient



## Amyloidosis cutis dyschromica - A rare variant of primary cutaneous amyloidosis.

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# Introduction & Objectives:

Considering very few examples documented in the literature, amyloidosis cutis dyschromica is an uncommon type of primary cutaneous amyloidosis. In 2018, it was thought to be a familial disorder caused by a mutation in GPNMB (Glycoprotein non-metastatic melanoma protein B), which results in autosomal recessive amyloidosis. Most frequently observed in the people of the Southeast Asian region. UVB-induced defective DNA repair processes are thought to be the etiology, and they cause keratinocyte destruction leading to apoptosis. Clinically, it manifests as reticular hyperpigmentation without symptoms, with hypopigmented macules found across the body, most typically in the neck, trunk, and limbs. Treatment options include acitretin, topical corticosteroids, CO2 laser, keratolytics, dimethyl sulfoxide, and sun protection. Acetretin, however, is regarded as the drug of choice.

# Materials & Methods: (Case details)

A 32 year old female patient presented to our OPD with multiple, asymptomatic, round to oval, white lesions over bilateral shin and trunk since 1 year. She noticed multiple, asymptomatic generalized pin-head sized white lesions over which started over bilateral legs and later on progressed to involve the trunk. Gradually increased in size and number over the past 6 months. Multiple hypopigmented macules of varying size (5-10 mm), with irregular borders present over bilateral shin and trunk in a symmetric pattern. Diffuse, band-like reticular-rippled hyperpigmentation with intervening normal skin present over the upper back. Similar lesions present over bilateral forearms. A 4 mm punch biopsy was taken from left leg and histopathology was advised.

## **Results:**

The histopathology report from skin suggested epidermal shows mild hyperkeratosis. Basal cell layer showed decrease in melanocytes. Intracytoplasmic melanin is seen only in very few cells. Superficial papillary dermis shows focal collection of acellular eosinophilic homogenous material which was positive for Congo res stain. Hence, the biopsy was confirmatory for late onset amyloidosis cutis dyschromica.

## **Conclusion:**

According to a recent article, very few cases have been documented worldwide. Since it manifests as hypopigmented macules spread widely over the body, especially over the trunk, along with reticular hyperpigmentation, it is challenging to diagnose amyloidosis cutis dyschromica and differentiate from other asymptomatic hypopigmentation disorders like idiopathic guttate hypomelanosis, progressive macular hypomelanosis, and follicular vitiligo. As a result, it's critical to identify it as a differential diagnosis of diffuse hyperpigmentation and macular hypopigmented lesions. A skin biopsy should be taken into consideration in order to confirm the diagnosis and determine the best course of treatment for the patient.



# Integrated Single-cell Transcriptomic Analysis Discloses Homogenous Fibroblast Landscape between Human Scars

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# Title: Integrated Single-cell Transcriptomic Analysis Discloses Homogenous Fibroblast Landscape between Human Scars

# **Introduction & Objectives:**

Human skin wound heals by forming scars, among which hypertrophic scars (HTSs) and keloids are the two most common types with both similarities and differences. However, whether HTSs and keloids are two separate disorders remains controversial. Here, we aim to elucidate the pathogenesis of HTSs and keloids, with a special focus on the fibroblast (Fb) landscape, by integrating the results of scRNA-seq datasets of human scars.

## Materials & Methods:

Twenty samples (including 3 HTSs, 7 keloids, 3 normal scar, 4 keloid adjacent normal skin and 3 normal skin) were included in an integrated single-cell analysis to develop a comprehensive fibroblast (Fb) landscape. Comparative analysis were conducted to decipher the differences of the functional activation and the evolution of Fb population between HTSs and keloids. The pathogenic Fb and the discriminative transcription factor were validated by fluorescent immunohistochemistry. *In vitro* experiments were conducted to evaluate the functional change of fibroblasts upon the activation of key transcription factors.

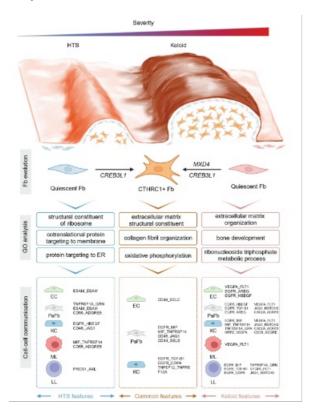
## **Results:**

Comparative single-cell profiling revealed that HTSs and keloid share a similar fibroblast landscape regarding the common pathogenic matrix-producing Fb, increased extracellular matrix production, subtype transition towards mesenchymal CTHRC1+αSMA+ Fb, and activation of *CREB3L1* as a key transcription factor during pathogenic Fb evolution. However, keloids differ from HTSs by exhibiting a higher level of functional activation, pro-inflammatory phenotype transition, and more comprehensive cell-cell communication. Notably, *MXD4* was identified as a discriminative transcription factor between HTSs and keloids. Exogenous over-expression of *CREB3L1* and *MXD4* both activates extracellular matrix production and induces myofibroblast differentiation *in vitro*. Interestingly, fibroblasts transfected with *MXD4* exhibits a higher functional activation level compared to those with *CREB3L1*.

# **Conclusion:**

These findings provide single-cell evidence that HTSs and keloids are the same type of skin fibroproliferative disease with different severities, benefiting clinical translation in terms of the differential diagnosis and the development of precision therapies.

**Graphical abstract:** 





**AMSTERDAM** 25-28 SEPTEMBER 2024 EUROPEAN ACADEMY OF DERMATOLOGY & VENEREOLOGY

# Abstract N°: 2503

# Identifying Cutaneous Collagenous Vasculopathy: A Diagnostic Challenge

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# Identifying Cutaneous Collagenous Vasculopathy: A Diagnostic Challenge

## Introduction:

Cutaneous collagenous vasculopathy (CCV) is a rare, idiopathic microangiopathy affecting the superficial dermal blood vessels. It is clinically characterized by the development of fine, arborizing telangiectasias, typically beginning on the lower limbs and gradually progressing upward. This case aims to highlight the principal characteristics of this lesser-known condition.

## **Case Report:**

A 61-year-old woman with a history of fibromyalgia, iron deficiency anemia, type 2 diabetes mellitus, and nonalcoholic fatty liver presented with a 5-year history of persistent macular lesions. These lesions began as small, non-pruritic, erythematous macules on both external forearms, which she noted would darken after summer but remain unchanged otherwise. The rash did not spread to other body parts, nor were there any signs of involvement on nail beds or mucosal surfaces. Her current medications included semaglutide, nortriptyline, omeprazole, tramadol, and paracetamol as needed. Examination revealed non blanchable telangiectatic patches on both extensor forearms. Routine blood tests for coagulation and connective tissue diseases were unremarkable. However, a punch biopsy indicated telangiectasia with the presence of PAS-positive hyalinized material within the vessel walls, a finding suggestive of cutaneous collagenous vasculopathy.

## **Discussion:**

Cutaneous collagenous vasculopathy (CCV) is a rare microangiopathy of dermal blood vessels, often underrecognized and predominantly reported in middle-aged and elderly individuals, though recent cases indicate a broader demographic may be affected. The disease typically presents with asymptomatic, telangiectatic macules that begin on the lower extremities and may progress to cover more extensive areas without involving mucosal surfaces. Despite its benign nature, the diagnostic challenge posed by CCV can lead to patient distress due to its persistent and spreading nature.

CCV is clinically and histopathologically distinct from other vascular disorders such as generalized essential telangiectasia and hereditary hemorrhagic telangiectasia, mainly through its unique histological findings of dilated vessels with collagen deposits. These findings are crucial for diagnosis, particularly in the absence of systemic symptoms or significant lab abnormalities. The pathogenesis is thought to involve endothelial damage followed by abnormal collagen deposition, although the exact mechanisms remain unclear.

## **Conclusion:**

This case report underscores the critical importance of recognizing cutaneous collagenous vasculopathy (CCV) as a potential diagnosis in patients presenting with chronic, unexplained telangiectasias. By enhancing awareness of CCV's distinctive features and improving its differentiation from similar vascular conditions, clinicians can achieve more accurate diagnoses, ultimately facilitating better patient management and outcomes.



## Rare Case of Multiples Epidermolytic Acanthomas in the Scrotal Region After Unilateral Orchiectomy

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**Introduction & Objectives:** Epidemolithic acanthoma (EA) is a rare disorder that can affect different areas of the body, with cases of isolated lesions such as scrotum, anus, eyes, genius and leg. For diagnosis, clinical correlation is required and the presence of multiple AEs in the genital and peri-genital region represents an individualized variant with well-defined characteristics. Because it is an entity with few publications in the literature, our goal is to report a case of multiple epidermolytic acanthomas in the scrotum after unilateral radical orchiectomy, which may often not be diagnosed by lack of knowledge of this disease and similarity with other genital disorders.

**Materials & Methods:** Patient, male, white, Brazilian, 50 years old, complaining of nodules in the scrotum that appeared 6 months ago, after performing surgery of right unilateral radical orchiectomy by a testicular adenomatoid tumor for 6 months. He states that since then he has emerged erythematous papules that scratch in the scrotum without modifications. The lesions were widespread in the scrotum, totaling 7. They were oval erythematous to normochromic papules with diameter between 1 and 5mm. The surface was keratotic. It was opted for excisional biopsy of one lesion with a 3 mm punch. The anatomopathological showed hyperplastic epidermis characterized by proliferation of spinked layer cells, with elongated and wide interpapillary ridges, hypergranulose and hyperkeratosis, as well as paraqueratosis, with granular and vacuolar degeneration of the spinous layer and layer granulosa, being the diagnosis suggestive of epidermolytic acanthomas.

**Results:** Epidermolytic acanthomas are benign and rare tumors of the skin, appear with erythematous to brownish papules smaller than 1cm and appearing in middle aged individuals. They usually occur as solitary lesions in intertriginous areas, such as armpits and groin. However, the presence of multiple epidermolytic acanthomas in the scrotal region is an unusual and poorly documented occurrence in medical literature. Because it is clinically prone to diagnostic errors in other lesions, additional examinations such as dermatoscopy and skin biopsy are required for confirmation. In dermatoscopy, "pearly white areas" are the striking feature of AE, which would correspond to compact hyperceratosis above hypergranulose and acanthosis. Possible etiological factors include ultraviolet irradiation, immunosuppression, trauma, viral infection and mutations involving the genes of keratin 1 and 10.

**Conclusion:** EA is a benign, no contagious disease, subject to cryotherapy, CO2 laser, or topical drugs such as imiquimod, calcipotriol and tacrolimus. It should be different from contagious diseases such as condyloma that need treatment, since treatment is not essential for EA. Thus, more researches needed to better understand the etiology, clinical course and management of multiples epidermolytic acanthomas in the scrotal region.



# "Efficacy of Topical Tacrolimus in Cutaneous Rosai Dorfman Disease: A Rare Case Report with Possible COVID-19 Association"

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**Introduction & Objectives:** Rosai Dorfman disease (RDD) is a rare non-Langerhans cell histiocytosis with an unknown etiology. Commonly presenting as painless bilateral cervical lymphadenopathy and fever, RDD may also involve extranodal sites, including the skin, occurring in over 40% of cases, with the skin being the sole affected organ in 3% of cases

**Materials & Methods:** Here, we present the case of a 58-year-old woman with a 4-month history of a rash characterized by multiple reddish-orange nodules on her left breast. Notably, the patient had a recent history of COVID-19 infection two months prior to the rash onset. Dermoscopic examination revealed a homogeneous reddish-orange area with branched vessels at the periphery. Ultrasound-guided biopsy confirmed the diagnosis, with immunohistochemistry showing positivity for S100, CD68, and CD163, while CD1a was negative. PET-CT imaging showed no evidence of systemic involvement, and the patient's complete blood count was within the normal range, that confirming cutaneous RDD.

**Results:** Treatment with topical 0.1% tacrolimus twice daily resulted in complete resolution of lesions over a 4-month period

**Conclusion:** Our findings suggest that topical tacrolimus may be an effective treatment option for cutaneous RDD, offering a promising therapeutic approach for this rare form of RDD. Additionally, the potential association between RDD and COVID-19 warrants further investigation.



# Nuclear Morphometric Analysis in Basal Cell Carcinoma Diagnosis and Subtype Classification: A Scoping Review

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**Introduction & Objectives:** Nuclear morphology results from the complex interplay of genetics, cell cycle, cytoskeletal dynamics, and tumor environment. Stemming from the dogma that form is shaped by function, morphology analysis has historically been used in microscopy to infer information about tumor biology. Computerized nuclear morphometric analysis (NMA) is a pioneering technique that provides a means of addressing the subjective nature of histopathologic cancer diagnosis. This scoping review evaluates the application of NMA in diagnosing, classifying, and assessing the risk profile of basal cell carcinoma (BCC).

**Materials & Methods:** Adhering to the Joanna Briggs Institute Reviewer's Manual (2020 version), a comprehensive search strategy was executed across multiple electronic databases including Web of Science, Scopus, Google Scholar, EMBASE, and MEDLINE on December 24, 2023. The methodology was detailed in a priori published protocol. Data extraction was performed using a standardized tool, followed by a narrative synthesis to summarize and interpret the findings.

**Results:** The scoping review of the literature identified thirteen studies that met the inclusion criteria. Although the evaluation of these studies revealed substantial variation in research designs, all studies addressing the utility of NMA in the diagnosis of BCC consistently affirmed the efficacy of NMA in distinguishing BCC from other lesions or non-lesional epithelium. Regarding risk-profile assessment and subtyping, the evidence indicates that aggressive BCCs are characterized by larger nuclear areas, perimeters, maximum diameters, ferets, and marked anisonucleosis. In terms of nuclear chromatin analysis, aggressive BCCs demonstrate darker nuclear gray levels with greater variance and higher fractal dimensions. Evidence suggests that changes in nuclear morphology may arise secondarily during tumor progression, as NMA failed to distinguish between low-risk primary tumors and early recurrent lesions. This observation has implications for the detection of subclinical extensions and contributes to a deeper understanding of tumor biology.\*\*

**Conclusion:** The current literature on the application of NMA in diagnosing and classifying BCC is limited, yet promising. A consistent conclusion has emerged regarding the potential of NMA as a reliable tool for BCC diagnosis, risk profile assessment, and subtype classification. The technique's ability to predict the aggressiveness of BCC tumors underscores its wide-reaching implications. These extend beyond standard dermatopathology, potentially benefiting cytopathology, automated histopathology workflows, machine learning, and more. With the advent of information technology, the accessibility of image analysis software has been significantly democratized, which may serve as a source of motivation for some innovative research to come.



# Vellus hair cyst a diagnostic challenge

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# Introduction & Objectives:

Vellus hair cyst, first noted by Esterly in 1977, is a rare benign condition affecting adolescents or young adults, with no gender or race preference. It can occur sporadically or be inherited in an autosomal dominant pattern, associated with keratin 17 gene mutations. Typically, it presents as multiple asymptomatic papules on the trunk and limbs, resembling milia, keratosis pilaris, or childhood eruptive histiocytoma. Healthcare providers should be familiar with this condition to aid diagnosis, assist pathologists, and reassure patients of its benign nature.

# Materials & Methods:

A 23-year-old female, previously healthy, presented with sporadic (micro)papular lesions on the ventral region of her forearms, describing a slight sensation of "pricking." She had experienced two intermittent episodes over the past two years, with spontaneous onset and disappearance. Physical examination revealed discrete hyperchromic/grayish papules in the affected areas. An incisional biopsy confirmed the diagnosis of eruptive vellus hair cysts upon histopathological review.

## **Results:**

Eruptive vellus hair cysts are uncommon in dermatological practice, lacking a distinct appearance for immediate diagnosis. Case reports contribute valuable insights, noting their usual presentation as persistent, papular, and hyperpigmented lesions. Spontaneous resolution occurs in up to 25% of cases, while symptoms may include itching or altered sensitivity. Squeezing can release viscous material containing fragmented hairs. Dermatoscopy may reveal circular or oval lesions with distinctive features, while high-frequency ultrasound and optical microscopy with 10% potassium hydroxide offer alternative diagnostic tools. These cysts can resemble acneiform eruptions on the back and buttocks, resistant to standard therapy, with their true etiology confirmed as vellus hair cysts..

The cause of vellus hair cysts remains elusive, occurring sporadically or in an autosomal dominant inheritance pattern. Mutations in the keratin 17 gene have been implicated. These cysts, often found in children, may have smooth or rough surfaces with central umbilication. Rupture can lead to inflammatory reactions resembling foreign body granulomas. Perforating dermatoses, milia, keratosis pilaris, adnexal tumors, acne, elaioconiosis, and childhood eruptive histiocytoma should be considered differentials. Treatment options are limited and include topical tretinoin, tazarotene, calcipotriol, oral isotretinoin, exfoliation, dermabrasion, CO2 laser, curettage, and surgical incision.

# **Conclusion:**

Eruptive vellus hair cyst can mimic other skin conditions. Physician awareness aids diagnosis, with histopathology and other tools helpful. Knowing the diagnosis reassures patients; treatment is challenging, but the condition is benign with potential for regression.





## A Clinico-pathological Study of Facial Granulomatous Dermatoses

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# Introduction & Objectives:

Granulomatous dermatoses arise from various aetiological factors, often exhibiting significant convergence in both clinical manifestations and histopathological features. Multiple causes can yield same histological pattern, and conversely, a single cause may provoke spectrum of histologic patterns, thereby posing diagnostic complexities for clinicians.

Objective: To diagnose aetiology of facial granulomatous dermatoses by determining histopathological profile, correlating it with the clinical features and determining the concordance rates

# Materials & Methods:

A total 94 patients of facial granulomatous dermatoses attending tertiary care hospital from January 2021 to December 2023 were included. All patients were subjected to routine clinical and laboratory examinations including a lesional skin biopsy for the histopathological study. Few patients were subjected to special stain. A clinico-histopathological correlation was done and concordance rates were calculated.

## **Results:**

Out of total 968 skin biopsies performed during the study, 177 were from face. Facial granulomatous dermatoses was clinically suspected in 94 and histologically proven in 89. The mean age of patients was 38.76 ±12.3 years (age range 7-71 years) with male to female ratio of 1.69:1. Majority of cases (n=79,84.04%) were of infectious origin, commonest being leprosy (68.09%), followed by cutaneous tuberculosis(7.45%), leishmaniasis(2.13%), phaeohyphomycosis(1.06%) and actinomycosis(1.06%). The non-infectious causes included sarcoidosis(6.38%), granulomatous rosacea(3.19%), granulomatous cheilitis(2.13%), necrobiotic xantho-granuloma(1.06%) and tattoo granuloma(1.06%).

Histologically, epithelioid cell granuloma was found in 69.15%, followed by histiocytic in 11.70%, sarcoidal in 7.45%, mixed inflammatory in 4.26%, palisading and foreign body granuloma in 1.06% each.

An overall concordance rate of 84.78% was observed in our study, considering the subclassification of leprosy while for leprosy cases, the concordance rate was 96.97%.

## **Conclusion:**

The major cause of facial granulomatous dermatoses in developing countries is still infection, leprosy being the commonest of all. The integration of clinico-histopathological features is very crucial for timely diagnosis and institution of correct treatment.



**AMSTERDAM** 25-28 SEPTEMBER 2024 EUROPEAN ACADEMY OF DERMATOLOGY & VENEREOLOGY

## Abstract N°: 2900

# Clinicopathological Survey of 204 Rosacea Patients Regarding Rosacea Subgroups and Severity

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<sup>1</sup>Department of Dermatology, Razi Hospital, Tehran University of Medical Sciences, Tehran, Iran,<sup>2</sup>Department of Pathology, Razi Hospital, Tehran University of Medical Sciences, Tehran, Iran, <sup>3</sup>Department of Plastic Surgery, Razi Hospital, Tehran University of Medical Sciences, Tehran, Iran, <sup>4</sup>Autoimmune Bullous Diseases Research Center, Tehran University of Medical Sciences, Tehran, Iran

# Introduction & Objectives:

Few studies are available regarding the clinical and histopathological findings in rosacea. Previous studies demonstrated that although the intensity of inflammation especially perifollicular lymphohistiocytic infiltration was higher in PPR than ETR, there is no specific histological pattern that can be diagnostic for any rosacea subtype. The goal of this study was to determine and compare the histopathological findings of rosacea subtypes.

# Materials & Methods:

The histopathological findings of 204 rosacea patients were analyzed retrospectively and the histopathological findings were compared among clinical subtypes. Rosacea diagnosis was made according to the National Rosacea Society (NRS) Expert Committee. The four rosacea subtypes were erythematotelangiectatic (ETR), papulopustular (PPR), phymatous rosacea (PHYR), and ocular rosacea (OR) defined by a modified NRS classification system. The severity was defined based on established criteria. We asked one dermatopathologist to determine both epidermal and dermal changes. The specimens were evaluated also in terms of Demodex mite presence.

## **Results:**

Thirty-Two Percent of patients were male and 68% were female. Seventy-three patients had ETR and 110 had PPR, 12 were ETR + PPR, 4 ocular, 2 phymatous, and 3 had Morbihan's edema. Perivascular and perifollicular lymphohistiocytic infiltration, perifollicular exocytosis, follicular spongiosis, and ectatic vessels were almost found in all subtypes. Solar elastosis was higher in ETR. Spongiosis, exocytosis of inflammatory cells into epidermis, acanthosis, and granulomatous reaction were higher in PPR. Inflammatory cells exocytosis was more in PPR and phymatous. Demodex folliculorum was identified in 27% of ETR, 33.6% of PPR, 50% of phymatous, one ocular patient, and none of Morbihan edema. Demodex brevis were found in 5% of ETR, 3% of PPR, and 50% of phymatous. Demodex brevis not folliculorum was more in phymatous. Spongiosis was the most common finding in ocular rosacea.

# **Conclusion:**

Our data did not show any histopathological finding specific for clinical rosacea subtypes. In support of previous studies, we suggested that perifollicular lymphohistiocytic infiltration is a nonspecific finding that is found in all rosacea subtypes. Based on our study, the most frequent epidermal changes in both ETR and PPR subtypes were spongiosis, exocytosis of inflammatory cells, acanthosis, and hyperkeratosis. Histopathological markers of sun exposure in rosacea including ectatic vessels in the superficial dermis, solar elastosis, dermal edema, and perivascular inflammation were all common in both ETR and PPR with no significant difference. Demodex mites were found in a high portion of rosacea patients. It was believed that Demodex mite would play a role in rosacea pathogenesis by inducing perifollicular inflammation. In our study, the frequency of Demodex mite was 31.5% in ETR and 37.2% in PPR. Interestingly our study showed that follicular spongiosis is significantly higher in mite

positive patients and follicular exocytosis was significantly lower in mite negative patients suggesting that mite may be associated with follicular spongiosis, rather than follicular exocytosis. However, considering current literature, mites are not the only etiologic factor of follicular inflammation and other factors including dysregulated innate immune system and neurogenic dysregulation contribute to the pathogenesis of rosacea.



## Histiocytoid Sweet Syndrome: Review of a Rare Entity, its Clinical Presentation, Diagnosis, and Treatment

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**Introduction & Objectives:** Histiocytoid Sweet Syndrome (HSS) is a rare variant of Sweet Syndrome, characterized by an infiltrate of histiocytoid or monocyte-like cells rather than the typical neutrophilic infiltration. This distinct presentation often leads to diagnostic challenges, as it can mimic other dermatological conditions including Leukemia Cutis. This review aims to summarize the different histopathological findings, differentials, associated diseases, and therapeutic strategies of HSS.

**Materials & Methods:** A comprehensive review of the literature was conducted using PubMed. The search strategy employed the following keyword: "Histiocytoid Sweet Syndrome". Additionally, all results were uploaded to Rayyan where articles were selected based on predefined inclusion and exclusion criteria to ensure the selection of appropriate studies for this literature review.

**Results:** Histiocytoid Sweet Syndrome is a rarely diagnosed variant of SS that was first described in 2005, often presenting as erythematous papules, nodules, or plaques in the face, neck, and superior trunk. Its diagnosis usually requires histopathological examination, revealing a dense dermal infiltrate predominantly composed of histiocytoid cells, which are monocyte-like with an oval or indented nucleus, resembling histiocytes, rather than the typical neutrophils seen in classical Sweet Syndrome. Such cases are often mistaken with leukemia cutis, with multiple shared characteristics including positivity for CD68, CD43 and MPO, hence posing a challenge for pathologists. The lack of expression of CD34 and CD117 excludes leukemic or highly undifferentiated cells, aiding in the differentiation between both diseases. HSS has been associated with underlying systemic conditions such as malignancies, myelodysplastic syndrome, and autoimmune diseases. A drug-induced etiology was also described in some cases, including antibiotics, bortezomib and vaccines. When it comes to management, HSS lesions often respond to corticosteroids. However, recurrences can be observed and associations with other diseases could complicate the therapeutic approach, requiring the prescription of alternative medications, such as etanercept, dapsone, and colchicine.\*\*

**Conclusion:** Histiocytoid Sweet Syndrome is a rare skin tumor with a challenging diagnosis and management. Final diagnosis often depends on histopathological evaluation of the skin lesions as HSS is an important mimicker of leukemia cutis. It is associated with diverse conditions and can be induced by pharmaceutical agents. Similarly to SS, its management relies on systemic corticosteroids. Alternative therapies may be necessary in particular cases, along with a multi-disciplinary approach.

Table 1: Diagnostic criteria for Sweet Syndrome

| Major criteria | Abrupt onset painful erythematous papules and nodules   |
|----------------|---|
|                | Histopathological evidence of neutrophilic infiltration without evidence of vasculitis.   |
| Minor Criteria | Fever over 380 C  |
|                | Association with an underlying hematologic or<br>visceral malignancy, pregnancy, or a preceding upper<br>respiratory infection, gastrointestinal infection, or<br>vaccination |
|                | Excellent response to treatment with systemic corticosteroid or potassium iodine  |
|                | Abnormal laboratory values upon presentation for at<br>least three of the following four tests: ESR II20 mm/h,<br>positive CRP, leukocyte II8,000 or neutrophils II70%        |



#### Features of skin melanoma accompanied by metastasis to the sentinel lymph node

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**Introduction & Objectives:** cutaneous melanoma (CM) is a malignant skin tumour originating from altered melanocytes and metastasizing quickly to the sentinel lymph node (SLN). The incidence of CM is increasing every year by 2%. The biggest influence on development of CM has sun exposure and genetic predisposition. 5-year survival rate of CM reaches up to 93% and depends on several factors: CM morphological type, Breslow depth, number of mitoses, ulceration, as well as metastases to the SLN and other organs. The exact localization of metastases in the SLN also plays important role in the prognosis. According to research, the best prognosis is when metastases are in the peripheral sinuses of the SLN. Therefore, this study aims to evaluate CM metastases localization in the SLN, correlation with the primary tumour and characteristics of skin melanoma, in the presence of the metastases in the SLN.

**Materials & Methods:** 154 patients diagnosed with localized cutaneous melanoma and regional sentinel lymph metastases between 2002 and 2021 were retrospectively reviewed. The patients were categorized into different groups based on several factors: age ( $\leq$ 44, 45-64, 65-79, <sup>3</sup>80; mean age 61), CM localization (head-neck, upper extremities, abdomen-chest, back-trunk, lower extremities), Breslow thickness (<1mm, 1-2mm, 2-4mm, >4mm) and morphological types (superficial spreading, nodular, acral, *lentigo maligna*, rare). The statistical program IBM SPSS was used for the statistical analysis of the data. Chi-square test and descriptive statistics was used during statistical analysis. Correlation was considered statistically significant when p<0.05.

**Results:** this research indicates that metastases in SLN are more frequent in nodular CM (53.7%), in CM of the lower extremities (37%), Breslow depth >4 mm (51.3%). Additionally, a strong association between CM with SLN metastases Breslow depth and CM morphological type and ulceration was found. Higher Breslow thickness was notably prevalent in nodular and acral CM (68.7% and 66.7% respectively) compared to superficial spreading CM (17%) (x2 =61.01; p<0.001). Presence of ulceration is more likely when CM with SLN metastases Breslow depth is >4mm (61.1%) compared to Breslow thickness <1mm (2.1%), 1-2mm (10.5%), 2-4mm (25.3%) (x2 =11.79; p=0.008).\*\*

**Conclusion:** in this study it was observed that when cutaneous melanoma (CM) is located on the lower extremity and has a Breslow thickness exceeding 4mm, it tends to metastasize to the sentinel lymph node (SLN).



#### clinicopathological features of blue nevus

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#### Introduction & Objectives:

Blue nevi are benign tumors derived from dermal melanocytes. Generally, dermal melanocytes regress during the latter stages of gestation; however, residual melanin-producing cells may persist in the dermis of specific anatomical regions such as the scalp, sacral region, and distal extremities. These regions are most commonly associated with the occurrence of blue nevi. Various types of blue nevi have been identified, including Common Blue Nevus, Cellular Blue Nevus, Epithelioid Blue Nevus, Pigmented Epithelioid Melanocytoma, and Malignant Blue Nevus.

#### Materials & Methods:

This study employed a cross-sectional design, where data pertaining to patients with blue nevi were extracted from archived files within the specified time period. Efforts were made to address data deficiencies through telephonic communication with the patients. Subsequently, the collected data was analyzed using appropriate statistical tests.

#### **Results:**

A total of 332 patients were included in this study. Among them, 87.95% presented with Common Blue Nevus, 10.24% with Cellular Blue Nevus, 0.90% with Epithelioid Blue Nevus, and no cases of Pigmented Epithelioid Melanocytoma or Malignant Blue Nevus were observed. Of the patients, 62.65% were female and 37.35% were male. The average age of the patients was 38.12±2.20 years. There was no significant difference in the average age between male and female patients. However, the average age of Common Blue Nevus patients was significantly higher than that of Cellular Blue Nevus patients. The average age at lesion onset was 19.67±3.32 years, with Common Blue Nevus patients exhibiting a significantly higher average age at onset compared to Cellular Blue Nevus patients. The average time interval between lesion appearance and biopsy was 165.34±27.98 months. Regarding lesion location, the head and neck region were most frequently affected, followed by the upper limbs, lower limbs, buttocks and sacrococcygeal region, trunk, genitals, and oral cavity. The difference in lesion location between the two common subtypes of blue nevi was significant. Only 13 patients presented with multiple lesions. Among the patients, 49.6% experienced an increase in lesion size, 22.9% reported a change in lesion color, 6.25% had a history of lesion bleeding, and 8.8% reported a history of lesion-related pain. Additionally, 14.5% had a family history of Blue Nevus, while 6% had a family history of melanoma. The average size of excised lesions was 11.85±1.54 mm, with Common Blue Nevus patients exhibiting a significantly lower average lesion size compared to Cellular Blue Nevus patients. No recurrences were observed among patients who were followed up through telephone calls. The average follow-up period was 87.55 months.

#### **Conclusion:**

We anticipate that this study will enhance the understanding of local disease characteristics among physicians and researchers, thereby facilitating more accurate diagnosis and appropriate treatment for patients.

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#### Abstract N°: 4029

#### clinical and dermoscopic aspects of extensive tuberous xanthoma

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## Introduction & Objectives:

Xanthomas are defined as papular or nodular lesions of brown-yellowish color; they contain foamy macrophagic histiocytes, loaded with lipids. Xanthomas can be classified into several types: including tuberous xanthomas, tendinous xanthomas, palmar crease xanthomas, diffuse plane xanthomas, eruptive xanthomas, and xanthelasma.

Their existence always reveals an underlying disease, their large size and the existence of at least two types proves to be a rarity: as is the case presented.

#### Materials & Methods:

We report a case of one patient treated in the dermatology department at Tangier University Hospital.

A 17 -year old girl with a family history of xanthomas in both her brothers and her uncle.

#### **Results:**

it's about a 17-year-old female patient presented with numerous large masses located in her elbows; knees; ankles, buttocks ; feet, and hands. The measures of the largest one was 20 cm × 10cm in size; she also had a gerontoxon . Dermoscopy has shown structreless yellow areas surrounded by an erythematous halo. Upon blood tests ; the patient exhibited elevated levels of low-density lipoprotein cholesterol (LDL: 604 mg/l)and was subsequently diagnosed with Type IIa familial hypercholesterolemia and multiple large co-existing tuberous and tendinous xanthomas which is a rare clinical presentation.Trans-thoracic cardiography didn't show any abnormalities.She was put on statins associated to dietetic measures and then reffered for surgical excision to remove the massive xanthomas from the elbows, knees, and feet.

#### **Conclusion:**

Extensive tuberous xanthoma can occur, requiring a multidisciplinary approach involving a dermatologist, endocrinologist, plastic surgeon, and geneticist.

Even though it's a rare condition, looking for metabolic disorders is a must ,and cardiac evaluation must be performed in order to eliminate any atherosclerotic plaques.

Finally, for a perfect management in this type of affections, a genetic study should be conducted.



#### Unveiling hookworm Folliculitis: understanding parasite interaction with follicular structures

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#### **Introduction & Objectives:**

Cutaneous larva migrans (CLM) is an ectoparasitic skin infestation typically caused by nematodes of the genus *Ancylostoma sp*, primarily in tropical and subtropical regions. The nematode larvae penetrate the skin through contact with soil contaminated by animal feces. Although they lack the enzymes necessary to degrade the basement membrane, preventing completion of their biological cycle in humans, they can still migrate through intraepidermal tunnels, resulting in erythematous and pruritic skin lesions characterized by a serpiginous tract appearance. Adnexal involvement in CLM, manifesting as folliculitis, is a rare clinical presentation, with limited histological descriptions available.

#### **Materials & Methods:**

We report two cases of CLM presenting with follicular involvement in patients with pruritic lesions following travel to subtropical areas. The application of skin ultrasound to guide the biopsy facilitated the visualization and identification of the parasite within the follicle in the second case.

#### **Results:**

A 32-year-old woman developed an abdominal skin lesion following a trip to Thailand, while a 28-year-old woman exhibited a knee lesion after a volunteer mission to Senegal. In the second case, a skin ultrasound revealed follicular dilatation at the distal end of the erythematous tract, culminating in a dilated area. Within this dilation, a small bilaminar hyperechogenic structure was identified. In both cases, an incisional biopsy of the lesions was performed. The larva extended through the external root sheath in both cases. In case 1, it made contact with the vitreous layer, creating a tunnel that extended to the proximity of the hair bulb. Interestingly, in the upper areas, a cavity with eosinophilic material was identified in the epidermis (case 1) and in the hair epithelium (case 2), corresponding to the larva's track. In both cases, the perifollicular dermis showed a sparse mixed inflammatory infiltrate, consisting of lymphocytes and scattered eosinophils. Treatment based on a single dose of weight-adjusted oral ivermectin, as an alternative to albendazole, has been successfully employed in the two presented cases.

**Conclusion:** This study highlights not only the rarity of follicular involvement in CLM but also offers new insights into its anatomopathological presentation and spread through the hair follicle.



## Epidemiological and Clinical Profile of Pilomatricoma: 65 cases

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## Introduction & Objectives:

Pilomatricoma is a benign cutaneous adnexal tumor arising from the hair matrix. It is most commonly encountered during the first two decades of life and is predominantly localized in the cervicofacial region.

#### Materials & Methods:

We report a retrospective descriptive series of all histologically confirmed cases of pilomatricoma over a 12-year period (2011-2023) to define its main characteristics

#### **Results:**

We collected 65 cases. The mean age at diagnosis was 38.4 years (range: 5-76 years). Patients under 20 years of age represented only 24.6%. The male-to-female ratio was 0.8. The mean duration of evolution was 29.5 months (range: 1 month-6 years). The diagnosis of pilomatricoma was rarely considered initially. The main differential diagnoses were cysts followed by calcinosis and dermatofibroma. The skin overlying the lesion was normal in 69.2% of cases. In other cases, it was erythematous (9.2%), pigmented (4.6%), bluish (6.12%), whitish (1.52%), anetodermic (1.52%), or perforated/ulcerated (3.05%), mimicking a keratoacanthoma (KA). The main locations, in descending order of frequency, were the upper limbs (44.6%), cervicofacial region (35.35%), lower limbs (15.38%), pubis (1 case), back (1 case), and buttocks (1 case). Tumor size ranged from 0.4 to 4 cm. Lesions were giant (> 3 cm) in 6.15% of cases. Pain was noted in 16.92% of patients. Lesions were always solitary. No associated morbid clinical conditions were found. Onset at the site of vaccination in the arm was reported in one case. Five patients underwent ultrasound, which did not show specific signs. Surgical excision was performed for all patients. Histological examination showed the presence of basaloid cells and mummified cells with calcifications (40%) and a giant cell inflammatory reaction (67.7%) in all cases. After surgery, only one recurrence was observed after 3 months, and one patient developed another pilomatricoma in a different location.

#### **Conclusion:**

The diagnosis of pilomatricoma is histological. Consistent with literature data, there is a female predominance in our series and the common form presents as a subcutaneous, solitary, hard nodule, sometimes painful, and smaller than 3 cm. Perforating, erythematous, pigmented, bluish, anetodermic, and giant forms are described and present in 30% of our patients. Healing without recurrence is the rule after surgical excision. The originality of our study lies in the predominance in adults, predominance of involvement of the upper limbs, presence of perforating forms, and occurrence of recurrence.



#### Histopathological surprise from simple looking keloidal lesions

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#### Introduction:

Kaposi's sarcoma has some histological variants bieng the keloidal variant is an exceptionally rare phenomenon, with the first documented 3 cases in a 1994 report by Schwartz et al. The lesions exhibited a firm, rubber-like texture resembling such of keloid. We present a case of Egyptian female complians of firm cutanoeus lesions on her upper limbs and chest resembling small rounded keloidal lesions. Upon primary histological examination there was dermal expansion by extensive keloidal collagen with few perivascular infilterate so, a diagnosis of keloid was suggested by pathologist. The presence of such keloidal collagen could blurr the specific spindle shaped cellular infilterate. Further work up utilizing Immunohistochemistry revealed strong CD 31/34 staining and unviels various shaped vascular spaces confirming the diagnosis of Kaposi sarcoma.

Through this lecture we discuss the existing insufficient literature and describe the clinical and pathological challanges for such rare entity since early diagnosis may help with effective treatment.



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## Abstract N°: 4619

## A rare constellation of cutaneous hemophagocytosis in a Sweet syndrome

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## Introduction & Objectives:

Cutaneous hemophagocytosis and Sweet syndrome are both rare and independent of each other. Clinically inevident cutaneous hemophagocytosis in the setting of a Sweet syndrome is extremely rare, with only one other case documented in the literature.

#### **Materials & Methods:**

Here, we present a case of a 38-year-old female patient with a painful skin rash and a right swollen knee joint that occurred two weeks after onset of streptococcal angina treated with amoxicillin and salicylic acid. Medical history was unremarkable.

#### **Results:**

Skin lesions presented as succulent livid red plaques from the forehead to the extensor sides of the upper arms. Clinically, a classical Sweet syndrome presented by post-infectious onset, distribution, morphology of typical skin lesions, and abnormal laboratory values including neutrophilic leukocytosis. Surprisingly, skin biopsy revealed not only a neutrophilic, Sweet-typical inflammation with interstitial neutrophils, eosinophils, superficial band-like neutrophils and a marked papillary edema, but also a striking phagocytosis of neutrophils by macrophages. In addition, immunohistochemistry marked immature neutrophils as histiocytoid cells (MPO- and CD68-positive). There was a rapid regression of the complaints and skin lesions under systemic high-dose prednisone therapy.

#### **Conclusion:**

Cutaneous hemophagocytosis can be a manifestation of primary hemophagocytic lymphohistiocytosis (HLH), which typically presents with fever, hepatosplenomegaly, cytopenia and coagulopathy. An ineffective pathogen elimination has been postulated which is compensated by excessive macrophage activation. Cutaneous hemophagocytosis restricted to the dermis is considered a localized, abortive form of HLH, here triggered by a previous respiratory streptococcal infection and possible drug cofactors. Extensive clarification is recommended only if indicators for a primary HLH are present.



## Lichenoid Dermatitis: A Clinicopathological Study Of 111 Patients

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#### **Introduction & Objectives:**

A number of clinically diverse inflammatory skin disorders are linked by a set of histopathological features that have been referred as lichenoid tissue reaction, the prototype of this category is lichen planus.

#### **Materials & Methods:**

Clinical and demographic evaluation of all patients diagnosed with lichenoid dermatitis on histopathology (focal vacuolar degeneration, lichenoid infiltrate at upper dermis and perifollicular location with pigment incontinence) were studied.

#### **Results:**

Total 111 patients were included, majority were in the age group 21-30 years (26.4%), followed by 41-50 years (19.6%)

There was no sex predilection. Most common diagnoses were lichen planus pigmentosus (23%), followed by classical lichen planus (19%), lichen planopilaris (16%) drug induced lichenoid eruption (8.8%), pityriasis lichenoides chronica (6.8%), hypertrophic lichen planus (5.8%), oral lichen planus (4%) others were lichen planus pemphigoides, atrophic Lichen planus, discoid lupus erythematosus, lupus vulgaris, lichen scrofulosorum, pityriasis lichenoides et varioliformis acuta, lupus erythematosus, linear lichen planus, nail lichen planus.

Diagnosis was established on histopathology in 85% cases and clinicopathological correlation was required in 14% cases.

Most common histopathological findings seen were lichenoid infiltrate in the dermis (96%), focal vacuolar degeneration of basal layer (93%) pigment incontinence (85%), hyperkeratosis (75%), acanthosis (66.6%).

On correlation of histopathological findings with the clinical findings, lichen planus pigmentosus was the most common diagnosis seen on trunk, face and upper extremities.

#### Table 1 showing the frequencies of common lichenoid dermatoses in the study

| Disease                            | Number of cases | Percentage (%) |
|------------------------------------|-----------------|----------------|
| lichen planus pigmentosus          | 26              | 23%            |
| classical lichen planus            | 21              | 19%            |
| lichen planopilaris                | 18              | 16%            |
| drug induced lichenoid<br>eruption | 10              | 8.8%           |
| pityriasis lichenoides chronica    | 7               | 6.8%           |
| hypertrophic lichen planus         | 6               | 5.8%           |
| oral lichen planus                 | 5               | 4%             |

## Table 2 showing comparison of frequencies of diagnoses with other studies

| Disease                               | Reeta Dhar et al<br>(N= 104)<br>** | Muralidhar,<br>Aparna (N=150)<br>** | Kumar et al<br>(N=107)<br>** | Current study<br>(N= 111) |
|---------------------------------------|------------------------------------|-------------------------------------|------------------------------|---------------------------|
| lichen planus<br>pigmentosus          | 72.4%                              | 38.67%                              | 6.66%                        | 23%                       |
| classical lichen<br>planus            | 67.4 %                             | 28.57%                              | 26.6%                        | 19%                       |
| lichen planopilaris                   | 5.8 %                              | 8.6%                                | -                            | 6%                        |
| drug induced<br>lichenoid eruption    | 1.1 %                              | 9%                                  | 3.33%                        | 8.8%                      |
| pityriasis<br>lichenoides<br>chronica | 5.8 %                              | -                                   | 5.55%                        | 6.8%                      |
| hypertrophic<br>lichen planus         | -                                  | 10.6%                               | 2.22%                        | 5.8%                      |
| oral lichen planus                    | 1.16%                              | 2.6%                                | -                            | 4%                        |

 Table 3 showing comparison of common histological features with other studies

| Features   | Reeta Dhar et al<br>(N= 104)<br>** | Muralidhar,<br>Aparna et<br>al (N=150)<br>** | Kumar et al<br>(N=107)<br>** | Current study<br>(N= 111) |
|--|------------------------------------|--|------------------------------|---------------------------|
| lichenoid infiltrate<br>in the dermis            | 94.1%                              | 58%  | 93.3%                        | 96%                       |
| focal vacuolar<br>degeneration of<br>basal layer | 94.1%                              | 95.2%  | 96.6%                        | 93%                       |
| pigment<br>incontinence                          | 83.7%                              | 90.32%                                       | 93.3%                        | 85%                       |
| hyperkeratosis                                   | 86.6%                              | 80.6%  | 93.3%                        | 75%                       |
| acanthosis                                       | 73.2%                              | 46.66%                                       | 83.3%                        | 66.6%                     |

**Conclusion:** It is important to understand that lichenoid tissue reaction has several dermatoses under its domain. Hence it is crucial to record precise clinical as well as histopathological findings for appropriate diagnosis and management.



## A retrospective histopathological study of patients with various granulomatous dermatoses attending a tertiary care hospital.

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**Introduction & Objectives:** Granulomatous dermatoses can be seen in infectious as well noninfectious conditions. Clinical features of these diseases are overlapping; thus, a confirmatory histopathology remains gold standard for accurate diagnosis.

## Materials & Methods:

This is a retrospective study with inclusion of both indoor and outdoor patients from June 2021 to march2024. Skin biopsy obtained from patients clinically diagnosed as various cutaneous granulomatous diseases was analyzed. Record was maintained in the form of demographic characteristics, morphology of lesions, duration of disease, clinical and histopathological diagnosis.

## **Results:**

126 cases were included in the study (84 males and 42 females), with a male to female ratio of 2:1. Most common age group affected was 21-30 (n=29;25.8%). Infectious granulomatous dermatoses (n=120;90%) were more common than the non-infectious ones (n=6;5.3%). Among the infectious granulomatous dermatoses, leprosy (n=80; 63.75%) remained the major entity followed by lupus vulgaris (n=11; 9.8%). Amongst leprosy cases borderline tuberculoid leprosy (n=40;35.7%) was the most common entity followed by borderline lepromatous leprosy (n=14;12.5%) and least common was mid borderline leprosy (n=2 1.78%). Most common form of lesion was erythematous plaque (n=60;53.5%) followed by hypo pigmented patch.

Among the noninfectious granulomatous disorders granuloma annulare (n=3;2.6%) was more common followed by sarcoidosis, Crohn's disease and granulomatous cheilitis (n=1, 0.8%) each.

#### **Conclusion:**

Histopathology along with proper history and clinical examination are gold standards for diagnosis and subclassification of cutaneous granulomatous lesions.





## Localized primary cutaneous amyloidosis: A series of 21 Cases

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## Introduction & Objectives:

Localized Primary Cutaneous Amyloidosis (LPCA) is a rare condition characterized by the deposition of amyloid substance in the dermis, without involvement of internal organs. It includes macular amyloidosis (MA), lichen amyloidosis (LA), biphasic amyloidosis (BA), and nodular amyloidosis (NA).

#### Materials & Methods:

All histologically confirmed cases of LPCA from a span of 27 years (2007 - 2023) were included in our study.

#### **Results:**

Twenty-one cases of LPCA were diagnosed, including 15 cases of LA, 4 cases of MA, and 2 cases of BA. No cases of NA were recorded. The average age of the patients was 46.4 years (ranging from 32 to 82 years), with 14 females (9 LA, 3 MA, 2 BA) and 7 males (6 LA, 1 MA), resulting in a male-to-female ratio of 1/2. A family history was found in 2 sisters.

Pruritus was the main reason for consultation, with an average duration of 8 years (ranging from 3 months to 20 years). Cutaneous examination revealed pruritic hyperkeratotic and pigmented papules for LA, hyperpigmented, gray-brown reticulated macules sometimes confluent in MA, and hyperpigmented maculopapules in both cases of BA.

In patients with LA, lesions predominated in the pre-tibial area (93%), forearms (28%), trunk (34%), posterior aspects of the legs (14%), and buttocks (14%). Localization was solitary (6.6%), bilateral (40%), and multiple (53.3%).

In patients with MA, lesions predominated on the forearms (50%), abdomen (25%), and upper back and chest (25%). For BA, lesions were located on the legs (100%), and forearms and trunk (50%). Three patients had a generalized form of LPCA.

Dermoscopy was performed on 5 patients with LA, revealing a central whitish hub surrounded by gray-brownish granules. Histological examination confirmed amyloid deposits in the dermal papillae in all cases of LPCA.

For LA, dermocorticosteroids and keratolytics led to improvement in pruritus (84%) and disappearance of keratosis (38%). For MA and BA, acitretin resulted in partial improvement in three patients.

## **Conclusion:**

LPCA, rare in individuals with fair skin types, is characterized by amyloid deposition limited to the superficial dermis. LA and MA are the most common forms. Middle-aged women are primarily affected. Preferred localization varies, with a tendency towards the interscapular region for MA and the pre-tibial area for LA. Our series showed a predominant localization of MA on the forearms. Pruritus is the most common symptom, but its absence is

observed in 10 to 40% of cases. Generalized forms of LPCA, observed in 3 of our patients, are rare with only a few isolated cases reported in the literature. The definitive diagnosis remains histopathological; however, dermoscopy can be highly beneficial in showing the appearance of pigmented papules around a "central hub".

Histopathology typically shows deposition of amorphous, eosinophilic, and Congo red-positive material in the papillary dermis, melanophages, as well as increased pigmentation of the basal layer. The amyloid deposit is usually located higher up in the dermal papillae in MA.

Treatment primarily relies on topical corticosteroids. Other therapeutic alternatives include calcipotriol, topical tacrolimus, ablative lasers, phototherapy, colchicine, topical cyclosporine, capsaicin, menthol, and methotrexate. Acitretin may present an interesting treatment option, as seen in our patients.



#### Tinea corporis bullosa misdiagnosed as bullous diseases

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## Introduction & Objectives:

We reported a patient with recurrent misdiagnosis of tinea corporis bullosa, providing reference and basis for the diagnosis of the disease.

## Materials & Methods:

A 67-year-old man with a 7-month history of erythematous pruritic papules and vesicles visits to our department. He was diagnosed with linear IgA bullous dermatosis and pustular psoriasis in other hospitals, and dispensed with retinoids, total glycerides of peony, and topical glucocorticoid treatment 2 months before. However, the patient's skin lesions and itching symptoms did not improve significantly, even exacerbated.

#### **Results:**

Upon physical dermatological examination, there were diffuse annular erythematous papulovesicular lesions on the extremity and trunk. Fungal hyphae were detected through direct microscopy. Skin biopsy obtained from the patient's back showed obvious hyphae, a basket like structure of the stratum corneum, bulla within the epidermis where a large number of neutrophils gathered and eosinophils infiltrated, sponge edema. IgG, IgM, IgA, C3 were negative by direct immunofluorescence, confirming a diagnosis of tinea corporis bullosa. After receiving oral itraconazole, the patient had a good prognosis in the timing of follow-up 4 weeks visits.

## **Conclusion:**

Our case report provides clinical doctors with the importance of distinguishing and correctly diagnosing tinea corporis bullosa.



#### Tuberous sclerosis of Bourneville: about 8 cases

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#### **Introduction & Objectives:**

Bourneville tuberous sclerosis complex (TSC), or systemic tuberous sclerosis (STS) or also called EPILOIA (Epilepsy - low intelligence - adenoma sebaceum), is a rare congenital condition that highlights the multiplicity of affected organs. Its prevalence is estimated at an average at 1/10,000 live births.

Although the clinical picture of TSC is dominated by cutaneous and neurological involvement, it remains underestimated in our Moroccan context due to the lack of awareness of the first signs of the disease.

We report eight cases of TSC collected at the dermatology department at the Mohammed VI University Hospital Center in Marrakech.

#### **Materials & Methods:**

Single-center retrospective study of 8 cases of tuberous sclerosis of Bourneville whose objective is to describe the epidemiologic profile; clinical and paraclinical, evolving and to compare our results with those of the literature, emphasizing diagnosis and early treatment.

#### **Results:**

Our series is characterized by a median age varying between 1 year and 32 years, with a clear female predominance (75%)

Dermatologic involvement dominated by achromic macules and angiofibromas was the mode of revelation in the other three patients. Multiple hypomelanotic patches were noted in all our patients. Koenen's tumors and the skin-like appearance were noted in one patient in the series.

Epilepsy was the mode of revelation and the reason for consultation in five children, two of whom had West syndrome before the age twelve months young. The skin involvement was discovered during the dermatological examination.

Our patient (case 5) presented with asymptomatic cardiac involvement, where we discovered incidentally during echocardiography.

The course of the disease varies depending on the different manifestations found in the patient.

#### **Conclusion:**

Tuberous sclerosis of Bourneville is characterized by multisystem involvement. Brain and kidney damage have a particularly serious clinical aspect, because they cause serious complications and can be life-threatening.

To conclude, it is important to remember, the importance of requesting a complete imaging evaluation, including cerebral, cardiac, renal and ophthalmological in any patient with TSC.



## Comparative analysis of the ABCB5 gene expression in perilesional vitiligo and normal skin biopsies

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## Introduction & Objectives:

Vitiligo is the most common skin pigmentary condition in humans. Its etiology is attributed to an autoimmune attack on the melanocytes which results in achromic lesions. There is a deregulated autoimmune attack against melanocytes, activation of free radicals and proinflammatory cytokines with a preference for certain skin areas. The principal ABCB5 isoform is a transmembrane transporter located in cytoplasmic membrane melanocytes that eliminates intracellular toxic metabolites, it also protects against chemotherapeutic agents in normal melanocytes and melanoma cells. It has been seen that ABCB5 is expressed in the mesenchymal stem cells of the dermis, in addition to having immunoregulatory properties co-expressed with the programmed death ligand PD-1, and as a regulator of pro-inflammatory chemokines produced by macrophages, neutrophil overstimulation, and promoter of Treg lymphocyte activity.

This work suggests that decreasing or altering the immunological regulation of mesenchymal cells can facilitate the immunological attack of CD8+ T lymphocytes in the epidermis, affecting the viability of melanocytes, resulting in skin depigmentation.

#### Materials & Methods:

This research aims to include 15 biopsy samples and 5 control samples. At present, we have gathered 5 biopsy samples from patients with non-segmental progressive vitiligo diagnosis through punch technique of 4mm from a public dermatology center, and 2 control samples that were donated from discarded materials at a private practice. Two probes were designed to detect messenger RNAs of the tyrosinase and ABCB5 genes in skin biopsies of patients and healthy controls, one for the tyrosinase messenger RNA (labeled with fluorescein) and other for ABCB5 messenger RNA (labeled with Cy5). Confocal microscopy was used to evaluate the emissions from these fluorophores. It's planned to perform immunohistochemistry with

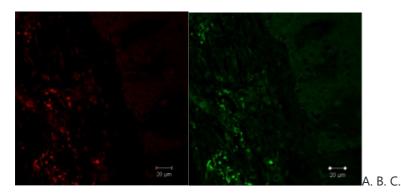
antiABCB5 monoclonal antibodies to enhance the gene detection to afford quantitative and comparative results between the transition zone and controls.

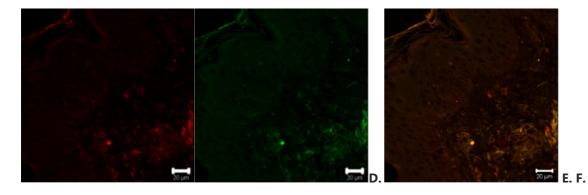
#### **Results:**

A, B. Confocal microscopy view of normal skin sample, image of melanocytes displaying green fluorescence indicative of tyrosine (Tyr) presence within in the basal epidermis. C. The presence of red (Cy5) in the transitional zone of epidermis, some parts of the dermis and blood vessels, but it is absent in the intracellular space and melanocytes. D, E. Vitiligo-affected basal epidermis exhibits reduced fluorescence as compared to controls, F. suggesting diminished tyrosinase activity and the presence of Cy5 red fluorochrome in a few dermis cells. Additionally, fewer differences were observed between vitiligo perilesional skin and healthy skin expression of ABCB5, which warrants further quantification and comparison with the rest of the sample.

#### **Conclusion:**

Previous studies conducted in melanoma skin report an overexpression of the protein related to the cell capacity of detoxification and tumor differentiation, normal skin present much less expression compared to melanoma samples and at the moment, our observations allow us to think that ABCB5 expression in vitiligo skin is lightly minor than in normal skin, what can be related to its immunoregulatory role in mesenchymal cells and the depletion of the regulatory response of local inflammatory environment.







**AMSTERDAM** 25-28 SEPTEMBER 2024 EUROPEAN ACADEMY OF DERMATOLOGY & VENEREOLOGY

## Abstract N°: 5804

## Post-Contraceptive Sweet Syndrome: An Atypical Case.

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#### Introduction:

Neutrophilic dermatosis of the sweet type is a rare skin condition characterized by acute inflammatory lesions, often painful, associated with neutrophil infiltration in the skin. It can be triggered by various medications, but progestin contraception remains a rare cause. Here, we present a case of post-medication Sweet syndrome on the face.

## **Observation:**

A 30-year-old female, with no significant medical history, presented to the dermatological emergency department with a facial rash evolving for 24 hours. During the medical history, the patient reported receiving an intramuscular injection of a progestin contraceptive a week prior. On clinical examination, erythematous, infiltrated, edematous, and painful plaques, round and sometimes confluent, were observed on the face. Blood tests showed normal complete blood count results, but slightly elevated C-reactive protein and erythrocyte sedimentation rate levels. Skin biopsy confirmed dermal neutrophil infiltration. The diagnosis of post-progestin contraception neutrophilic Sweet dermatosis was made. The patient was treated with indomethacin and vitamin C, along with rest and sun protection, resulting in significant improvement.

#### **Discussion:**

Neutrophilic Sweet dermatosis, although rare, can manifest as a drug reaction, especially after taking oral contraceptives or other medications. In our case, the onset of skin lesions on the patient's face shortly after the injection of a progestin contraceptive suggests a potential link between the two events. Histopathological confirmation of dermal neutrophil infiltration further supports this diagnosis.

Treatment of post-medication neutrophilic Sweet dermatosis often relies on non-steroidal anti-inflammatory drugs such as indomethacin, as well as immunomodulatory agents. In our case, the administration of indomethacin and vitamin C led to significant improvement in the patient's skin symptoms.

It is important to promptly recognize and treat this skin condition, as it can cause significant discomfort for the patient. Close monitoring and regular follow-up are also necessary to assess the disease's progression and adjust treatment if necessary. By raising awareness among healthcare professionals about this rare cutaneous manifestation potentially linked to the use of certain medications, we can improve the management of patients with post-medication neutrophilic Sweet dermatosis.



## "Sebaceous Trichofolliculoma: A Rare Follicular Hamartoma Masquerading as Basal Cell Carcinoma"

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#### **Introduction & Objectives:**

Sebaceous trichofolliculoma, an uncommon benign tumor arising from the hair follicle, is characterized by the presence of sebaceous glands and hair follicle structures. İt tends to be more prevalent among middle-aged adults. Clinically and dermoscopically, sebaceous trichofolliculoma can mimic basal cell carcinoma (BCC). However, simple surgical excision is sufficient for treatment. Therefore, it is crucial to distinguish this rare benign tumor from lesions that require aggressive surgery. Herein, we present a case of sebaceous trichofolliculoma exhibiting clinical and dermoscopic features closely resembling those of BCC.

## Materials & Methods:

A 75-year-old male presented with a 3-year history of a slowly growing papule located in the right zygomatic region. Upon clinical examination, a smooth-surfaced, soft, skin colour papule with telangiectasias was observed. On dermatoscopic examination, thick branching vessels, blue-gray ovoid globules and yellowish-orange areas were noted at the periphery of the papule. A biopsy was performed on the lesion, initially diagnosed as BCC. Histopathological examination of the biopsy specimen demonstrated features consistent with sebaceous trichofolliculoma, including hair follicle structures and sebaceous differentiation. Following the diagnosis of sebaceous trichofolliculoma, the patient underwent surgical excision of the lesion with clear margins. Postoperative recovery was uneventful, and there was no evidence of recurrence during the follow-up period.

#### **Results:**

Sebaceous trichofolliculomas are rare, benign hamartomatous cutaneous adnexal tumors originating from the hair follicle. On examination, central opening, hair follicle structures and sebaceous differentiation helpful in the diagnosis. If the hair is plucked as in our case, trichofolliculoma can be clinically misdiagnosed as basal cell carcinoma, keratoacanthoma, trichoepithelioma or sebaceous hyperplasia. Also dermoscopic features of sebaceous trichofolliculoma can closely resemble those of basal cell carcinoma, leading to diagnostic challenges. Arborizing vessels, focal ulceration, and blue-gray ovoid globules typically associated with basal cell carcinoma, may also be observed in sebaceous trichofolliculoma .While these dermoscopic findings may overlap between BCC and sebaceous trichofolliculoma, histopathological examination remains the gold standard for definitive diagnosis.Formun Üstü Histopathologically, sebaceous trichofolliculoma is characterized by multiple hair follicles embedded within a fibrous stroma, often accompanied by sebaceous glands. Our histopathological findings were consistent with it.

## **Conclusion:**

Accurate diagnosis of sebaceous trichofolliculoma is paramount to avoid unnecessary aggressive surgery. While dermoscopy provides valuable insights, histopathological examination remains the gold standard for definitive diagnosis. Clinicians should exercise caution in interpreting dermoscopic findings and consider histopathology to guide appropriate management strategies. Further research is warranted to elucidate optimal diagnostic approaches for this rare benign tumor.



## A study on epidermal aging: New insights from change of skin surface lipidomics profile and phenotype in different age groups

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**Introduction & Objectives:** \*\* The stratum corneum (SC), the uppermost layer of the skin, play an important role in the skin barrier function and appearance. It is a lipid-rich region that provides structural support and limits entry of chemicals. The integrity of SC can impact skin pigmentation, the presence of fine lines, and overall skin texture. Over the years, several investigations have shown an association between the alteration of skin's lipid composition and diseased skin, including psoriasis and atopic dermatitis. Nevertheless, the relationship between change in the specific composition of skin lipids as age range and their phenotype has not been elucidated. This study aimed to uncover the relationship between common visible alterations and lipid composition changes in aging skin by analyzing the lipid profile of the SC.

#### Materials & Methods:

To perform skin surface lipid profiling of the stratum corneum, skin tape stripes (STS) were obtained using D-squame discs from 101 Korean women in various age (age range= 20–24, 30-39, 43-46, 50-59, 67-69 years; sampling sites: cheek). At the same time, the skin's features, including skin roughness, transparency, color, gloss, and oil content were evaluated. The skin lipidomic analysis of STS samples were performed by liquid chromatography-tandem mass spectrometry (LC-MS/MS). The effect of the specific lipids associated with aging was evaluated by using PCR analysis on keratinocytes and melanocytes.

#### **Results:**

Among the skin parameters, there were significant changes in skin color and roughness by age. Evident reductions in skin color (brightness) were observed on individuals in their 40s and 60s. Within the stratum corneum, there is a significant association between aging and decrease in the content of long chain fatty acid (C>20), ceramide EOS, and cholesteryl metabolite. There was a high correlation between the skin darkness and decline long chain fatty acid with aging. Furthermore, treatment with long chain fatty acid (behenic acid) inhibited melanin synthesis on melanocytes. Decreased lipids in aging skin, including long chain fatty acid, ceramide EOS, and cholesteryl metabolite, treatment increased the expressions of keratinocyte differentiation markers.

#### **Conclusion:**

The findings indicate that alterations in skin texture and color resulting from the aging process are associated with modifications in the specific lipid composition of the SC. Therefore, recovery of specific lipid species that are characteristic of the young groups could be a promising approach to improving age-related skin changes.



#### Diagnostic dilemma of cutaneous histiocytic sarcoma - Histopathology to the aid : A rare case report

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#### **Introduction & Objectives:**

Histiocytic sarcoma (HS) is a rare hematological neoplasm originating from histocytes or dendritic cells. Lesions can be nodal or extra nodal, and are commonly seen in gastrointestinal sites in fifth decade of life. Skin is affected in almost 7% of the cases, and lesions can be maculopapular or soft in consistency or subcutaneous nodules with central necrosis and ulcers. Diagnosis is confirmed by immunohistochemistry (IHC). The prognosis is poor due to late presentation and delay in diagnosis.

#### **Case report**

A 51-year old male patient with chronic liver disease secondary to hepatitis B infection presented with multiple discrete, shiny, erythematous, tender nodules of varying sizes (ranging from 1cm to 2 cm in diameter) over trunk and proximal extremities for one month. A diagnosis of histioid leprosy was considered however, neurological examination was unremarkable with no loss of sensations and slit skin smear was negative for acid fast bacilli. Skin punch biopsy was then performed from a nodule over trunk, which was suggestive of lymphoproliferative disorder and IHC analysis was advised. Fite stain was negative. Complete blood count was normal and liver function test was mildly deranged. At next visit after one month, the size and number of lesions had drastically increased. A repeat punch biopsy was performed and IHC analysis was done. Histopathology showed transdermal infiltrate of tumour cells, dense localization in the reticular dermis, with periadnexal localization in the deeper dermis and infiltration in the form of sheets in between the collagen bundles. Grenz zone was noted below the epidermis. Tumor cells were positive for CD 45, CD68 and CD 4, diagnostic of Histiocytic Sarcoma. Hot spot Ki-67 index was 48% which indicates a high proliferative activity. Patient received chemotherapy at local hospital and is in clinical remission.

#### **Discussion:**

The World Health Organization defines histiocytic sarcoma as a malignant neoplasm with morphologic and immunophenotypic features of histiocytes. HS is a less discussed entity in dermatology due to paucity of cases that have exclusive cutaneous manifestation. In this case the resemblance of lesions to histioid leprosy was interesting and misleading. The morphology is very similar to that of other large cell neoplasms such as diffuse large B-cell lymphoma or anaplastic large cell lymphoma. Despite morphologic similarities, there are histomorphologic, immunohistochemical, and molecular features that can provide distinction between histiocytic sarcoma and its mimics.

## **Conclusion:**

The diagnosis of histiocytic sarcoma can be very difficult due to its rarity and histologic overlap with various mimics. It is essential to recognise morphologic cues and use immunohistochemical markers in order to verify histiocytic differentiation and rule out mimics.

25 SEPTEMBER - 28 SEPTEMBER 2024 POWERED BY M-ANAGE.COM



# YAP1-NUTM1 fusions are infrequent in porocarcinomas arising in a cohort of immunocompetent and immunosuppressed patients from two tertiary U.K. centres.

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## Introduction & Objectives:

Eccrine porocarcinomas are a rare tumour with a significant risk of metastasis. Genetic fusion events between the *YAP1* and *NUTM1* gene were reported in 64% of 11 cases in a landmark study in patients from Japan. Recently, *YAP1 - MAML2* fusion events have been shown to be present in a significant proportion (43%) of a cohort of 14 immunocompetent French patients. We sought to establish the frequency of this fusion and other fusion events in a broader ancestral background, as well as in the context of immunosuppression.

## Materials & Methods:

We have:1) Reviewed the existing literature and summarised the known gene fusions which have been found in association with porocarcinomas. 2)Carried out a RNA sequencing based fusion detection assay on 21 samples of porocarcinomas in patients who were immunocompetent and immunosuppressed from two tertiary dermatological centres.

#### **Results:**

To date 40 gene fusion positive porocarcinoma cases have been identified from our literature review. 72.5% were *YAP1-NUTM1*, 22.5% *YAP1-MAML2*, 2.5% *WWTR1-NUTM1*, and 2.5% *CTNND1/PAK1*. In our cohort of 21 patients, 43% were female, had a median age of 70 years and predominantly were of white European ancestry. Six of the patients were immunosuppressed; two out of the six had received renal transplants. The most frequent site of porocarcinoma was the lower leg. Of previously reported fusions, we detected a *YAP1-NUTM1* fusion in one case (4.7%) and a *YAP1-MAML2* fusion in one other (4.7%). We detected a fusion involving *CTNNA1-PAK2* in 2 samples from one case, and *PAK2-CTNNA1* and *CTNNA1-PAK2* in an additional case. Additionally, we found a fusion involving *TTC6-MIPOL1* which has been reported previously in other cancers. Eleven cases did not have any fusion detected. A further five cases had changes involving additional genes undergoing validation.

## **Conclusion:**

We found a low rate of *YAP1-NUTM1* fusions in our cohort, suggesting that the frequency of this fusion may vary between populations. This has diagnostic implications for the use of either a fusion assay for genetic diagnosis or immunohistochemistry for NUT1. Additionally, we identified potential driver fusions involving PAK2, some which have been recently identified in poroma. The fusions we found were different to previous groups and this may be due to the patient population studied, the inclusion of immunosuppressed patients and the different assay technologies used. Our work adds to the existing literature on the range of fusions in eccrine porocarcinoma.



## Clinico-Pathological Insights in Dermatologic Conditions: An Analysis of 1133 Skin Biopsies from a Tertiary Care Facility

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<sup>1</sup>Seth V.C.Gandhi & M.A Vora Municipal General Hospital., dvl

# Title- Clinico-Pathological Insights in Dermatologic Conditions: An Analysis of 1133 Skin Biopsies from a Tertiary Care Facility

## Introduction & Objectives:

Skin biopsies are a pivotal tool in dermatology, frequently used by dermatologists to diagnose a variety of skin conditions. However, the interpretation and diagnostic yield of these biopsies can be influenced by various factors. In our study, we set out to investigate the clinico-pathological correlations and disparities in different dermatological conditions. Our objective was not only to identify these correlations but also to understand the underlying factors that impact them. This analysis aims to provide a deeper understanding, improving diagnostic precision and patient management.

#### Materials & Methods:

A retrospective analysis of 1133 biopsies carried out in the outpatient and inpatient department of Dermatology, Venereology and Leprology was conducted over a period of 2 years and looked for clinico-pathological correlation.

#### **Results:**

Out of 1133 cases the maximum and minimum age biopsied was 72 years and 4 years, respectively, with a mean age of  $39.81 \pm 18.43$  years. The most common age group biopsied was 30-39 years(n=540;47.66%). There were a total of 644 males and 489 females giving a male to female ratio of 1.3:1. The most common site biopsied was the lower limb (n = 348) and the least common was the face (n = 88) These cases were classified into 4 major categories: inflammatory/infectious (65.56%;n=743), neoplastic (21%;n=238), other diseases (10.06%;n=114), and non-diagnostic/non-concordant (3.35%;n=38). Inflammatory diseases were subcategorised into granulomatous diseases accounting for 5.33%(n=47), psoriasiform diseases 25.43%(n=189), lichenoid diseases 17.90%(n=133), vasculopathic diseases 6.73%(n=51), spongiotic diseases 28.46% (n=211), and vesiculobullous diseases 15.61% (n=116). Most common infectious condition was hansens diasease(n=74;9.95%). Amongst neoplastic cases keratinocytic neoplasms accounted for 78.99%(n=188), melanocytic neoplasms 8.82%(n=21), and other cell-derived neoplasms 12.18%(n=29). Most common benign tumour reported was pyogenic granuloma . Basal cell carcinoma was the most common malignant neoplasm reported. The overall concordance rate observed in the present study was 97.63% (1106 out of 1133).

#### **Conclusion:**

The study underscored a robust clinico-pathological correlation, affirming the indispensable role of skin biopsy in dermatological diagnostics. While the results were encouraging, there's always room for improvement to refine diagnostic accuracy and elevate the standard of patient care. To achieve this, a multifaceted approach is essential. Regular and systematic departmental evaluations can provide valuable insights into diagnostic practices. Collaborative discussions between pathologists and clinicians can foster interdisciplinary learning and problem-

solving. Additionally, re-evaluating cases with discrepancies through repeat biopsies can help pinpoint diagnostic challenges and improve accuracy.



**AMSTERDAM** 25-28 SEPTEMBER 2024 EUROPEAN ACADEMY OF DERMATOLOGY & VENEREOLOGY

#### Abstract N°: 6849

#### Diagnostic confusion: two cases of gestational pemphigoid mimicking scabies

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#### Introduction & Objectives:

Gestational pemphigoid (PG) is a rare obstetric autoimmune blistering disease, usually occurring during the second or third trimesters of pregnancy. Its rarity makes it unfamiliar to clinicians, often misdiagnosed and consequently improperly managed. Herein, we report two cases of PG mistakenly treated as scabies

## **Clinical cases:**

We report the cases of two pregnant patients, aged 36 and 26 years respectively, at 20 and 33 weeks of gestation, who presented to our facility with symptoms evolving for about ten days before admission, characterized by persistent generalized itching associated with postbullous erosions covered with honey-colored crusts. Both patients were initially treated for scabies with benzyl benzoate without improvement.

Histopathological examination of skin biopsy revealed a pattern of eosinophil margination along the junction with linear deposits of C3 and C1q along the basement membrane in both cases.

The diagnosis of gestational pemphigoid, thus established, led to the initiation of treatment with oral corticosteroids at a dose of 0.5 mg/kg/day, resulting in significant improvement of itching and cutaneous lesions in both patients.

#### **Discussion:**

The diagnosis of PG should be considered based on the often characteristic clinical presentation and confirmed by histologic and direct immunofluorescence studies of skin biopsy. Treatment primarily relies on corticosteroid therapy, with a risk of relapse during subsequent pregnancies. This obstetric dermatosis, described as rare in the literature, is likely underdiagnosed, often leading to diagnostic delays resulting in persistent symptoms,

significant psychological impact, and considerable maternal-fetal risk.

## **Conclusion:**

The two cases reported here highlights the importance of considering the diagnosis of gestational pemphigoid in any generalized pruritus during pregnancy, especially in the presence of characteristic lesions, without resorting to easily made diagnoses, to ensure optimal management of parturients.



#### Rare case of hidradenoma papilliferum on a background of inflammatory linear verrucous epidermal nevus

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## Introduction & Objectives:

Inflammatory linear verrucous epidermal nevus (ILVEN) is rare hamartoma derived from embryonic ectoderm. It usually appears at birth or within first five years of life, an adult onset has been described too. Typically presenting with pruritic unilateral lesions, following lines of blaschko.

Hidradenoma papilliferum (HAP) is a rare benign cystic, papillary tumour earlier thought to arise from apocrine glands, is now thought to be of mammary-like gland origin.

Here, we present an interesting case where a female in her 30's presented to us with few raised lesions in right inguinal area which started at birth as single papule. The lesions increased in their size and number during puberty. Along with this, three nodular swellings were palpated, which were present for 4 years.

#### Materials & Methods:

An otherwise healthy, female, in early 30s presented to us perturbed by appearance of multiple, raised, intensely itchy, recalcitrant lesions around right inguinal area since puberty. At birth, her mother had noticed a single hyperpigmented papule which after remaining static for years, commenced to increase in size, number and elevation around puberty. Examination of right upper thigh and right inguinal area revealed two types of lesions; multiple hyperkeratotic, hyperpigmented papules and three, well circumscribed, firm, non-tender nodules of approximately 1 cm diameter, one showing ulceration. On enquiry, she revealed that she noticed these nodules about 4 years ago. Grossly, the nodules appeared tan-to-pink, fleshy, and well circumscribed. Examination of other areas was unremarkable.

A 4mm punch biopsy from a papule and an excisional biopsy of one of the nodules was sent for histopathological examination.

#### **Results:**

Punch biopsy from papule revealed epidermal hyperkeratosis, focal parakeratosis, papillomatosis and perivascular sparse lympho-histiocytic infiltrate, corroborating our impression of ILVEN and excisional biopsy of nodular swelling revealed findings characteristic of HAP namely, tubular, cystic spaces containing papillary structures, their lumina lined by inner layer of single row of columnar cells and outer layer of cuboidal myoepithelial cells.

The verrucous lesions of ILVEN were abalated with 2940 nm Er: YAG laser monthly sittings over a period of 3 months. The remaining two nodular swellings were surgically excised in-toto and sutured under aseptic precautions. Itching reduced significantly and the verrucous and papular lesions flattened. Excision site of the nodules had epithelized well without any recurrence till the last follow up 5 months post procedure

#### **Conclusion:**

Here we present a case of ectopic HAP over an unusual site - inguinal area on a background of lesions of ILVEN.

Our diagnosis was mainly clinical and was supported by a compatible histological picture. Although the occurrence of HAP with ILVEN is very rare, the possibility of coexistence cannot be excluded.



#### Perforating folicular pustular granuloma annulare

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#### Perforating folicular pustular granuloma annulare

## **Introduction & Objectives:**

Granuloma annulare is a skin disorder characterized by ring-shaped, asymptomatic, or pruritic plaques. Classic forms include localized, generalized, and subcutaneous granuloma annulare. Although it has been associated with certain conditions such as diabetes, dyslipidemia, or HIV, its etiology remains largely unknown. Several other infrequent atypical variants of the disease, such as perforating, palmo-plantar, blaschko-linear, pustular, and visceral granuloma annulare, have been described. Among these forms, perforating granuloma annulare is the most commonly reported. We report a case of granuloma annulare with uncommon clinical and histological features.

#### Results (clinical case):

A 69-year-old patient presented with a minimally pruritic cutaneous eruption of 12 years duration. Clinical examination revealed erythematous papules and scattered, confluent pustules located on the trunk and lower limbs. The patient had been treated with topical corticosteroids and PUVA without an adequate clinical response. Histological study showed subcorneal pustulosis and dermal necrobiosis surrounded by a radial infiltrate consisting of histiocytes and neutrophils, predominantly peri and intra-follicular, along with perforation and necrosis of the follicle wall. The clinical and histological data allowed the diagnosis of perforating follicular pustular granuloma annulare. The patient is being treated with isotretinoin with clinical improvement of the lesions.

#### **Discussion:**

Perforating granuloma annulare can present either localized to the extremities or generalized. It produces asymptomatic or pruritic red, crusty, scaly, umbilicated, or pustule-like papules, annular plaques, or scars. In addition to granulomatous inflammation, the perforating variant exhibits distinct histological findings such as transepithelial elimination of altered collagen. This extrusion of collagen may clinically resemble pustules, but they are not genuinely created by neutrophilic infiltration. However, true pustules can also infrequently develop when numerous neutrophils leak out along with the transepidermally extruded necrobiotic materials. Some cases of granuloma annulare with genuine pustules but without transepithelial extrusion of collagen have also been reported, making it impossible to classify them as a perforating variant. Nevertheless, the clinical presentation as a generalized pustular eruption is rare, with fewer than five cases reported in the literature.

#### **Conclusion:**

An atypical variant of granuloma annulare is presented. Clinical-histological correlation is very important to assess the correct diagnosis and treatment of these cutaneous entities.





#### Clinical and histopathological study of cutaneous hypopigmented lesions

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#### **Introduction & Objectives:**

Hypopigmentary disorders comprise a significant group of disorders that affect Indians and Asians. The pigment disturbance in darker skin individuals can be very distressing to the patient and the family.

These disorders cover a wide array of pathologies including Infections, Autoimmune processes, Lymphoproliferative disorders, and Sclerosing diseases. Histological diagnosis is particularly important because treatments for these diseases are varied and specific

The intention of this study is to correlate the histopathological findings with clinical findings of hypopigmented disorders of skin to arrive at an accurate diagnosis.

#### Materials & Methods:

Skin biopsies from patients with hypopigmented skin lesions were collected from patients in the Department of Dermatology and collected skin biopsies were processed and reported in Department of Pathology, Medicover Hospitals, Hyderabad. Descriptive statistical analysis and hypothesis will be conducted using appropriate statistical software packages.

#### **Results:**

The different types of clinical diagnosis observed in this study were pityriasis versicolor, Hansen's disease, vitiligo, LSA, Lichen striatus, IGH, PLC, parapsoriasis, Woronoff ring of psoriasis, DLE and PMLE. Clinicopathological correlation was observed in 83% of cases. Among them pityriasis versicolor showed 75%, Lichen striatus showed 45%, PLC showed 65% correlation and parapsoriasis showed 31% correlation.

#### **Conclusion:**

Systematic approach of clinical, histopathological examination and Immunohistochemistry will provide an accurate diagnosis of hypopigmented disorders and thereby reducing the patient distress.



## Pathogenesis and clinical manifestations of acanthosis nigricans

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## Introduction & Objectives:

Acanthosis nigricans (AN) is a dermatosis characterized by thickened, hyperpigmented plaques and papular hypertrophy typically symmetrically distributed on the intertriginous surfaces and neck. Early recognition of acanthosis nigricans is very important because it can be a cutaneous manifestation of a variety of systemic disorders, primarily with endocrinologic disorders and obesity when found in adolescents. Severe AN is rare and may be associated with malignancy.

## Materials & Methods:

17 patients ( 6 male and 11 female) with AN were included in the study. The aim was to study the epidemiology, clinical features and histopathological characteristics of patients with AN.

Diagnosis of associated disorders was established by history, physical examination, body mass index (BMI), measurement of hormones: thyroidnfunction tests, free testosterone, 17 (OH) progesterone, dehydroepiandrosterone sulfate (DHEAS), cortisol, gonadotropins, prolactin, immunoreactive insulin, and C-peptide levels.

#### **Results:**

In our study, the flexural involvement (groins, knees and elbows) was seen in 16 patients, dorsal involvement was seen in 2 patients. Increased serum testosterone levels were seen in 3 patients and increased DHEAS levels - in 5 patients. Regarding the types of AN, obesity was seen 13 patients, syndromic AN was seen in 3 patients. The commonest histopathological feature of patients with AN was hyperkeratosis, seen in all cases, papillomatosis was seen in 10 patients, dermal infiltrate of lymphocytes and plasma cells was seen in 11% patients and irregular acanthosis was seen in 2 patients.

## **Conclusion:**

Acantosis nigricans is a manageable condition, however, the complete reversion of the lesions is difficult to achieve. Treatment of the lesions is only for cosmetic reasons, but frequently it is the patient's primary concern. Usually AN improves with treatment of its underlying conditions. Correction of hyperinsulinemia often reduces the burden of hyperkeratotic lesions. Likewise, weight reduction in obesity-associated acanthosis nigricans may result in resolution of dermatosis.



#### Not Just in Children: Adult-Onset Xanthogranuloma - A Entity to Remember

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**Introduction & Objectives:** Juvenil xanthogranuloma, a rare variant of non-Langerhans cell histiocytosis, typically manifests as solitary or multiple papules or nodules predominantly affecting children. However, its occurrence in adults, particularly in the absence of systemic involvement, remains exceedingly uncommon.

**Materials & Methods:** Data from medical records and literature review. The keywords included were "xanthogranuloma" and "adult-onset".

**Results:** We present a case of a 25-year-old male, with no significant medical history, including dermatological conditions, and no family history of skin diseases. The patient presented to our dermatology department with a 2-month history of an asymptomatic erythematous papule on the left groin measuring approximately 0.5 cm in diameter. There were no signs of inflammation or ulceration, and the surrounding skin appeared normal. The patient reported that the lesion had appeared spontaneously and had been accidentally traumatized during shaving in the preceding weeks. He denied experiencing any pain, pruritus, or systemic symptoms associated with the lesion. Upon examination, He had no other similar skin lesions. Histological examination of the excised lesion revealed a dermal infiltrate composed of histiocytes with abundant foamy cytoplasm, scattered lymphocytes, and occasional multinucleated giant cells, supporting the diagnosis of adult-onset xanthogranuloma. On one-year follow-up, the patient remained asymptomatic with no signs of systemic diseases or new skin lesions.

**Conclusion:** Xanthogranuloma is a rare histiocytic disorder characterized by the proliferation of histiocytes laden with lipid-laden foamy cytoplasm, often arranged in a granulomatous pattern. While typically observed in pediatric populations, particularly infants and young children, isolated xanthogranuloma can occasionally manifest in adults. The clinical presentation of isolated xanthogranuloma in adults can vary widely, ranging from solitary papules or nodules to multiple lesions resembling eruptive xanthomas. Management of isolated xanthogranuloma typically involves surgical excision of the lesion, as was performed in our case. Recurrence following excision is rare, and systemic involvement is exceedingly uncommon. However, long-term follow-up is recommended to monitor for any signs of recurrence or development of systemic disease.



## Bullous striae distensae: a rare entity

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## **Introduction & Objectives:**

Striae Distensae, or stretch marks, are a common occurrence in dermatological practice. However, Bullous Striae Distensae is an exceptionally rare complication, with limited mentions in medical literature.

The significance of our case report lies in its rarity, aiming to bring attention to this relatively unknown entity.

#### Case report

A 41-year-old woman with a 4-year history of Crohn's disease was admitted to the Gastroenterology department due to anasarca. A dermatological consultation was sought for bullous lesions that had developed over the past two weeks. Dermatological examination revealed tense, edematous, shiny and lobulated fluid-filled lesions arranged linearly along the stretch marks of the abdomen, thighs, and popliteal fossae. The rest of the clinical examination showed tense edema of the lower limbs as well as moderate ascites. Abdominal ultrasound revealed a dysmorphic cirrhotic liver, ascitic fluid analysis showed a transudate, and laboratory tests revealed a serological profile of chronic hepatitis B, altered liver function, a low prothrombin level at 37%, and hypoalbuminemia at 9 g/L. The diagnosis of Bullous Striae Distensae in the context of cirrhosis was confirmed, and the primary focus was on correcting the hypoalbuminemia and liver dysfunction.

#### **Discussion:**

Striae Distensae are common skin lesions characterized by atrophic, linear plaques that often appear parallel to each other. While typically benign and linked to factors like pregnancy or rapid weight gain, they can also arise from various medical conditions. Bullous Striae Distensae (BSD) is an exceptionally rare variant, documented only a few times in medical literature. Its exact cause remains uncertain, though some theories suggest mechanical stress on the dermis leads to rupture of dermal blood vessels and subsequent blister formation. Almashat et al. proposed that BSD may be triggered by conditions like nephrotic syndrome and chronic steroid use, especially in cases of anasarca. In our case and others, patients with hypoalbuminemia due to different pathologies experienced BSD, with our case marking the third reported instance associated with cirrhosis. Ascites in our patient may have heightened mechanical stress on the dermis, potentially increasing susceptibility to BSD. Differential diagnosis for BSD includes autoimmune bullous diseases and bullous infections, and management revolves around treating the underlying cause.

## **Conclusion:**

While BSD is uncommon, it's crucial to recognize it to prevent misdiagnosis, unnecessary biopsies, and inappropriate treatment. Further research is warranted to improve our understanding of BSD's underlying pathogenesis.



## Wells Syndrome: A case report

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#### **Introduction & Objectives:**

Wells syndrome or eosinophilic cellulitis is a rare dermatosis of unknown etiology. It is clinically characterized by an acute dermatitis resembling cellulitis. It is benign but recurrent.

We report a 37-year-old woman with acute erythematous bullous plaque on the right forearm.

## Materials & Methods:

A 37-year-old woman with a history of Widal syndrome presented to our clinic with a limited erythematousbullous plaque with centrifugal annular erythema on the right forearm followed by pruritus and burning at the site.

Laboratory tests revealed an absolute eosinophil count of 1500 cells/mm3. Histopathologic examination of a biopsy taken from the plaque revealed a highly edematous papillary dermis. It is the site of a perivascular and periannexal cellular infiltrate rich in neutrophils and eosinophils, with no obvious signs of vasculitis.

Based on the clinical presentation and histopathology, a diagnosis of Wells syndrome was made. The patient was treated with oral prednisone 35 mg/day for 10 days. The forearm lesion resolved, leaving mild hyperpigmentation with induration. No new lesions appeared during treatment and 1 month follow-up.

#### **Results:**

Wells syndrome is a rare dermatosis of unknown etiology. It can be explained as an inappropriate eosinophilic response to a variety of stimuli due to an abnormal function of eosinophil regulatory systems. Wells syndrome is most commonly observed in adults, but can occur at any age.

The typical clinical presentation of eosinophilic cellulitis is a mildly pruritic, recurrent, cellulite-like plaque, although various clinical presentations such as urticarial, papulonodular, and vesicobullous lesions have been reported, depending on the location of the infiltrate.

The diagnosis of Wells syndrome is based on the clinical features and course of the disease, especially its recurrences, and the histopathologic features of eosinophilic infiltration of the dermis and "flame figures" located in the mid to deep dermis. Flame figures are not unique to Wells syndrome and can be seen in any condition with an eosinophilic infiltrate, such as insect bites, pemphigoid, and Churg-Strauss syndrome.

Treatment of Wells syndrome is not codified. Corticosteroids and dapsone are the two main treatments for Wells syndrome.

## **Conclusion:**

Wells syndrome can easily be confused with acute bacterial cellulitis. Therefore, first-line clinicians should be aware of this dermatosis. The diagnosis of Wells syndrome is mainly based on histologic examination and identification of flame figures, but these findings are not specific to Wells syndrome.



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#### Differentiation between Pigmented Pilomatricoma, Melanocytic Matricoma and Pilomatrical Carcinomas

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**Introduction & Objectives:** in this report we present a case of pigmented pilomatricoma, among with it's clinical features, dermoscopy and histological images, and also perform a literature revision about it.

Our objective is to analyze this rare variant of a common tumour, applying it to the practice of dermatologists, since most articles found about this topic came from Pathology journals and it was verified a lack of information and images turned to the practice of Dermatology. Also, we intend to clarify the differences between clinically or histologically similar neoplasms such as melanocytic matricomas and pilomatrical carcinomas.

**Materials & Methods:** we report the case of a 17 year-old female who presented with a nodular blue-black lesion on the right upper arm. Pathology showed a nodulo-cystic lesion extending from the dermis into the subcutis, with prominent shadow cell formation and melanocytes. In order to deduce the differential diagnosis, a literature review was performed and a table containing the main distinctions between them was made.

**Results:** 

|                            | Pigmented<br>pilomatricoma   | Melanocytic<br>matricomas  | Pilomatrical carcinoma   |
|----------------------------|--|--|--|
| Epidemiology               | Young individuals<br>Head and neck (3)   | Sun-damaged skin<br>Males<br>60-80 years old   | Elderly individuals<br>Male<br>Head and neck (3)   |
| Clinical Features          | Pigmented cyst   | Pigmented nodule-<br>cystic lesion   | Asymptomatic, slowly<br>growing, firm,<br>nontender nodules  |
| Histology                  | Integral cystic structure<br>with large aggregates of<br>shadow cells, presence<br>of calcification and<br>granulomatous<br>response | rounded solid<br>aggregations of matrical<br>cells positioned mostly<br>within the dermis and<br>upper part of the<br>subcutaneous tissue (3)<br>and focal areas of<br>shadow cell formation,<br>lack of epidermal or<br>infundibular connection<br>or calcification of ghost<br>cells | pleomorphic basaloid<br>cells with prominent<br>nucleoli and frequent<br>atypical mitoses<br>accompanied by central<br>areas with keratotic<br>material, shadow cells,<br>and foci of necrosis (3) |
| Melanin deposition         | Infrequent   | Infrequent   | Infrequent   |
| Melanocytes<br>hyperplasia | Rare   | More frequent  | Infrequent   |

## **Conclusion:**

The case reported illustrates the clinical and histological features of Pigmented pilomatricomas, which are similar to melanocytic matricomas and pilomatrical carcinomas. The most important differential diagnosis of these three entities are pigmented basal cell carcinomas and melanomas, therefore is of extreme interest that dermatologists and pathologists learn their characteristics.