

**Abstract N°: 72****A retrospective descriptive study of the histopathological spectrum of Erythema Nodosum Leprosum**

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**Introduction:** Reactions in leprosy are acute exacerbations that interrupt the chronic course of disease. These reactions are classified into Type 1 and Type 2 reaction (erythema nodosum leprosum). In our routine practice leukocytoclastic vasculitis found only in a minority of cases with a variable degree of acid-fast bacilli positivity. There are no recent studies with a primary focus on the histopathology of erythema nodosum leprosum.

**Aims:** To describe the histopathological spectrum of erythema nodosum leprosum in our cases.

**Materials & Methods:** A two-year retrospective descriptive study was conducted at the Department of Dermatology and Venereology and Pathology at All India Institute of Medical Sciences, New Delhi, between January 2021 and November 2022. The study, approved by the institutional ethics committee, involved searching digital records of the pathology department. 167 cases were identified, of which 120 slides were included in the study after excluding cases that did not meet the ENL criteria as evaluated by two dermatopathologists. A pre-designed proforma was used to record the histopathologic findings of each slide, and statistical analysis was carried out using descriptive statistics.

**Results:** Leprosy pattern was seen in 45.8% of biopsies, along with loose leprosy patterns, pan-dermal patterns, predominant subcutis involvement, and top-heavy, and bottom-heavy leprosy patterns. More than half (55%) of the biopsies had dense inflammatory infiltrates, and moderate and mild inflammatory infiltrates were seen in 40.8% and 4.2% of biopsies respectively. Neutrophilic infiltrates of varying severity were seen in the majority of the slides. There was predominant involvement of the subcutis in 6 cases while in 72 cases, the subcutis was involved by the dermal infiltrate extending into it. Fifty-seven (55.4%) biopsies had lobular involvement, while septal and septo-lobular involvement was seen in 3 (2.9%) and 14 (14.6%) of biopsies respectively. We noted medium and small vessel vasculitis in a total of 11 (9.2%) cases. AFB positivity was seen in 25% of cases and was mainly fragmented and granular.

**Conclusion:** In conclusion, we found dermal infiltrates in variable patterns and of varying density. Neutrophils were prominent in the infiltrate. Vasculitis was uncommon and panniculitis occurred largely as an extension of the dermal inflammatory process.



**Abstract N°: 538****Aneurysmal Fibrous Histiocytomas: Case Series and Review of the Literature**Lauren Passby\*<sup>1</sup>, Taherah Khan<sup>2</sup>, Danning LI<sup>2</sup><sup>1</sup>University Hospitals Birmingham NHS Foundation Trust, Dermatology, Solihull, Birmingham, <sup>2</sup>Sandwell and West Birmingham Hospitals NHS Trust, Birmingham, United Kingdom**Introduction & Objectives:**

Aneurysmal fibrous histiocytoma (AFH) is a rare but distinct variant of benign fibrous histiocytoma, representing 1.7% of all histiocytomas (1). As with benign fibrous histiocytomas, AFHs typically present as solitary nodules on the limbs of middle-aged adults, with no predilection regarding sex or race (1). AFHs arise from blood vessel proliferation and haemorrhage into a fibrous histiocytoma, resulting in polychromatic lesions that may have a red/purple/blue/black/yellow hue (1,2). Unlike benign fibrous histiocytomas, AFHs are typically larger lesions, with diameters ranging from 5 – 40mm (1). AFHs may be associated with pain or an episode of rapid growth due to spontaneous haemorrhage. We reviewed all cases of histologically-confirmed AFHs presenting to two UK dermatology centres over a 10 year period to assess patient characteristics, lesion characteristics, diagnostic methods, and treatment options.

**Materials & Methods:**

Through search of departmental histopathology databases, patients were identified who had a confirmed histological diagnosis of aneurysmal fibrous histiocytoma over a 10-year period. Case notes and images were reviewed retrospectively. Written consent was gained for use of medical photography. 13 cases were identified across two UK dermatology centres, all of whom were referred via the NHS Two Week Wait skin cancer referral pathway as solitary lesions. We reviewed patient demographics, clinical features, differential diagnoses, histological findings, and management plans.

**Results:**

The mean age at presentation was 56, with a large range of ages (16-91; median 52), and a slight female predilection (54%). All cases presented as solitary lesions: 10 on limbs, 2 on the head and neck, and 1 on the trunk. The lesions varied in clinical size from 3mm – 20mm in the widest diameter. Trauma (e.g. insect bites, previous excision/intervention, bleeding) at the site of the lesion were documented in 7 cases and there was 1 case of recurrence of the histiocytoma. Many differential diagnoses were considered for the cases including: melanoma, angiosarcoma, dermatofibrosarcoma protuberans (DFSP), blue rubber bleb naevus, glomus tumour, and benign vascular neoplasms. Due to various malignant differential diagnoses, all lesions were excised with a 2mm surgical excision margin. Histopathological examination, and immunohistochemistry analysis of Vimentin, Factor XIIIa, CD31, CD34, and HHV-8 status was conducted. Negativity for HMB-45, MelanA, MART-1, and S100 was commented upon in select cases, helping to exclude melanoma.

**Conclusion:**

Due to the suspicious clinical behaviour of rapid growth and morphological change, AFHs can mimic malignant lesions. Common clinical differential diagnoses include malignant melanoma, angiosarcoma, Kaposi's sarcoma, and adnexal tumours (3). In all cases discussed in this series the diagnosis was made histologically, and immunohistochemistry played an important role in ruling out sinister differentials. Given the potential malignant differential diagnoses, surgical excision remains the mainstay of diagnosis and treatment. Histopathological

diagnosis is critical in these circumstances to exclude the differentials discussed above. Clinicians should consider AFH as a possible differential diagnosis for new or changing vascular-looking lesions, particularly in the demographics and anatomical sites discussed above, however histological diagnosis should be sought.

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**Abstract N°: 1042****Melanocytic Tumour of Uncertain Malignant Potential (MelTUMP) - A Diagnostic Dilemma in Children**

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**Introduction:** The histopathological diagnosis and classification of melanocytic skin tumors is a practical challenge in paediatric dermatopathology. Sometimes, the major difficulty is distinguishing between benign and malignant microscopical patterns of a skin lesion. These borderline entities are defined as melanocytic tumours of uncertain malignant potential (MelTUMPs).

**Case report:** We are reporting the case of a 12-year-old female patient who was referred to a local hospital for a pigmented skin lesion which recently increased in size. The infracentimetric lesion was located on the posterior trunk. Due to dermoscopy findings, the lesion was excised, and the initial histopathological diagnosis was nodular melanoma. Reevaluation of the lesion performed in an oncology center raised the suspicion of a MelTUMP. The microscopy described a proliferation of large rather fusiform melanocytes arranged in confluent junctional nests in a hyperplastic epidermis with lymphocytic infiltrate. Minimal cell atypia and no mitotic activity were identified. HMB45 was positive and Ki67 index was 20%.

Due to the diagnosis difference, other opinions were requested from experts. Two more versions were obtained: compound blue nevus (CBN) and pigmented epithelioid melanocytoma (PEM) with loss of PRKAR1A expression. As ultrasonography (US) revealed bilateral axillary polyadenopathies, sentinel lymph node biopsy (SNB) was performed, showing subcapsular metastases with the same histopathological pattern. PET scan revealed no FDG up-take after the excision. The patient underwent 3 mg/kg Nivolumab immunotherapy every 4 weeks for 1 year. The follow-up including blood tests, US check and CT scan once in 6 months did not show the recurrence. The patient has been free of disease for 2 years.

**Discussion:** The studies show that MelTUMPs are not a well-defined histological entity, as it reunites a set of borderline melanocytic skin conditions. The diagnosis is made by IHC, but the markers used are frequently non-specific. In a minority of cases, these lesions have the ability to metastasize locally with rare systemic metastases. Local excision should be performed in all cases, but some studies show that positive SNB has low importance in diagnosis and prognosis. The WHO 2018 guideline recognizes PEM as an individual lesion and rules it out of MelTUMP category by PRKCA fusion, but inactivation of PRKAR1A is also highly correlated with the diagnosis of PEM. The lesion in our case is diagnosed as PEM, therefore recurrence and mortality rates are low. This condition should be treated as a high grade melanocytoma, according to WHO.

**Conclusion:** Here we report a case of a rare paediatric skin tumour. Owing their frequency and non-specific diagnostic methods, individual entities of MelTUMPs are challenging to be differentiated from one another. The positive diagnosis is often made after being examined by experienced dermatopathologists, as was the case of our patient.



**Abstract N°: 1371****Histopathological Features of Facial Melanoma - What to Expect from an “Ugly Duck” Located on the Face**Luana-Andreea Bosoteanu<sup>\*1, 2</sup>, Mariana Așchie<sup>3, 4, 5</sup>, Mădălina Boșoteanu<sup>3, 4</sup>

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**Introduction & Objectives:**

Cutaneous melanoma of the face is a scarce malignant dermatological entity that implies poorer prognosis respective to primary melanomas of other anatomic sites. This analysis aims to identify the distinctive histopathological features of facial melanomas.

**Materials & Methods:**

We conducted a descriptive, retrospective study on facial melanomas diagnosed in South-Eastern Romania between 2018-2022, extracted from a cohort of 100 melanoma patients. Clinical information such as sex, age and anatomic site was collected. The histopathological subtype, Breslow index, mitotic rate, presence of vascular and perineural invasion were re-evaluated by an expert pathologist, using the collected tissue samples.

**Results:**

12 patients were included, with a 2:1 female-male ratio and mean age at diagnosis of  $67.16 \pm 21.51$ . The most frequent location was the left cheek (50.00%), followed by an equal distribution between the right cheek and right temporal region (16.67% each). The predominant subtype was superficial spreading melanoma (SSM, 58.33%), backed by nodular melanoma (41.67%). The majority had a Breslow index superior to 4.00 mm (66.67%), with a mean thickness of  $9.16 \pm 7.79$  mm. Ulceration was observed in 83.33% of lesions (especially in SSMs) and the average mitotic rate was  $4.16 \pm 3.86/\text{mm}^2$ . Vascular and perineural invasion were seen in 41.67% and 16.67% of cases, respectively.

**Conclusion:**

Our study showed that SSM is the most frequent subtype among facial melanomas diagnosed in South-Eastern Romania. In conclusion, the expected histopathological model of a melanocytic lesion of the face displaying malignant clinical and dermoscopic features is that of a thick, ulcerated melanoma, with a mitotic rate superior to  $1/\text{mm}^2$ , without vascular, nor perineural invasion. Further investigation is required in order to attain a consensus regarding management particularities of this special melanoma location.

**Abstract N°: 1543**

### **Atypical presentation of Adult-onset Still's disease**

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

We present a female case of Still's disease with onset at an advanced age, with an atypical skin rash, associated with odynophagia and pericarditis. We highlight the importance of recognizing atypical presentations in order not to delay the diagnosis and institute the right treatment and support.

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

All information was collected from assessments during hospitalization and subsequent outpatient visits. Manuscripts were reviewed by all cited authors.

#### **Results:**

We present a woman, 66 years old, being followed up due to breast cancer, hospitalized with a maculopapular rash on the trunk, pruritic and persistent, worsening during episodes of high fever, associated with odynophagia and pericarditis.

After complementary evaluation, she also presents anemia, ferritin > 7500, in addition to increase of liver enzymes and altered kidney function. We perform a biopsy of cutaneous lesion and histopathological analysis showed an epidermis with acanthosis, apoptotic keratinocytes and a moderate lymphohistioneutrophilic infiltrate, perivascular and interstitial located in the upper and middle dermis.

The patient were treated with prednisone 60 mg orally and methotrexate 15 mg/week, with improvement in few weeks without new skin lesions.

#### **Conclusion:**

Still's disease is a diagnosis of exclusion based on the Yamaguchi criteria with high sensitivity and specificity. Fever is the most important sign, found in more than 90% of patients, occurring in daily peaks, usually at the same times. Others common signs and symptoms are skin rash, arthralgia and arthritis. Also, it can occur non-suppurative pharyngotonsillitis preceding or occurring together with the fever.

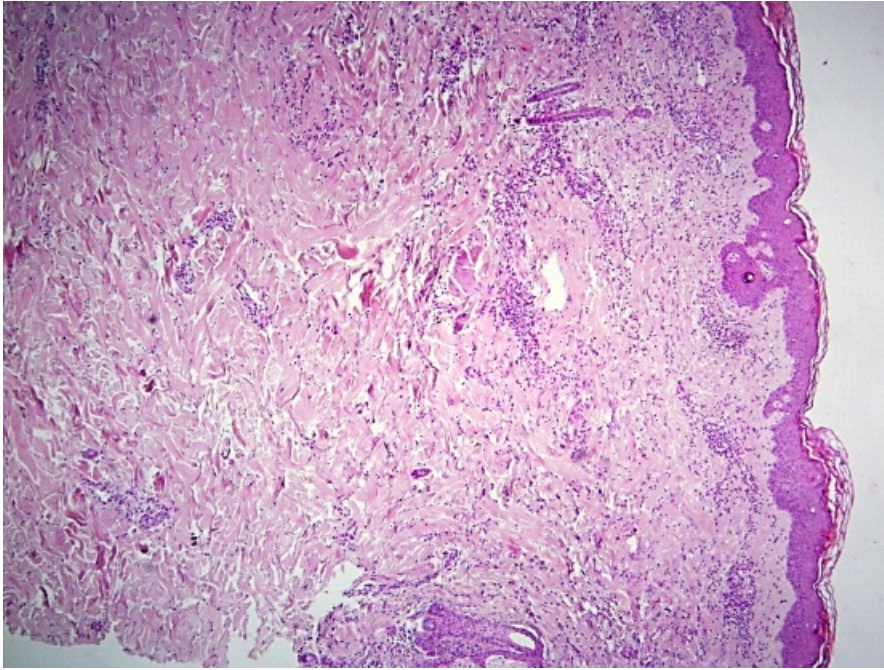
Cutaneous manifestations are found in 60 to 80% of patients, with a characteristic non-pruritic, transient and salmon-pink maculopapular rash, following fever peaks and mainly affecting the thorax and upper limbs. Atypical dermatological manifestations are more common findings in older patients, including persistent and pruritic rash.

As well as the cutaneous manifestations, the histopathological findings can also differ. In case of typical rash, we find histological features of nonspecific lesions composed of a superficial and dispersed perivascular lymphocytic infiltrate of neutrophils in the upper dermis. When persistent itchy papules and plaques are present, there is usually a distinctive histological pattern characterized by isolated or aggregated dyskeratotic/necrotic keratinocytes in the upper layers of the epidermis in association with an infiltrate perivascular and sometimes interstitial inflammation, in the upper and middle dermis, without vasculitis. Our case shows the latter pattern.

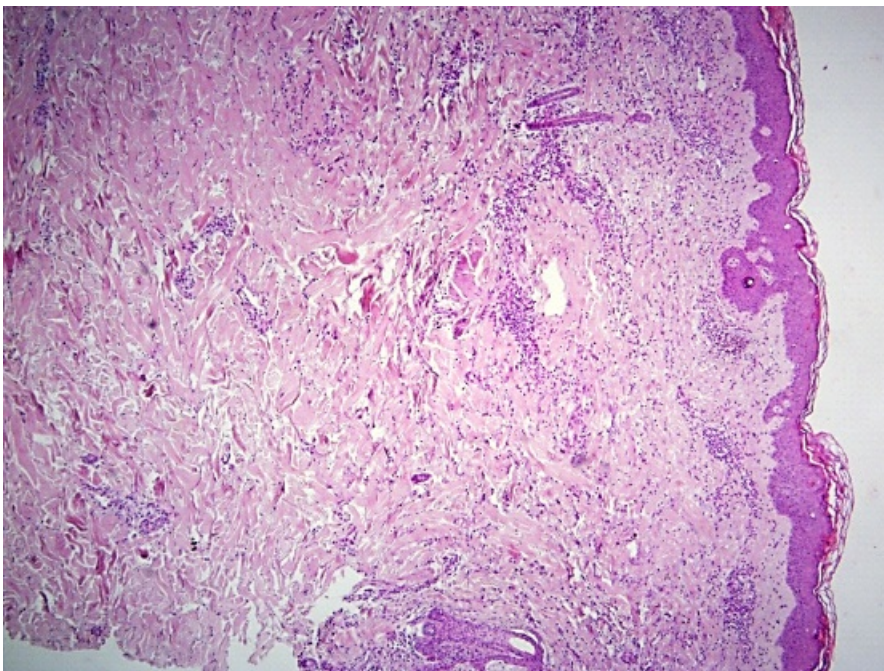
We presented a case of Elderly-onset Still's disease, with atypical clinical manifestation as well as histopathological



records, whose records are scarce in the literature. Despite recent studies demonstrating the possibility of worse evolution in patients in this age group, there was a good and quick response to the established therapy. We've still followed-up the patient for two years and she still in remission of the condition so far.



Epidermis with acanthosis. Upper and middle dermis with moderate lymphohistioneutrophilic, perivascular and interstitial infiltrate. (Hematoxylin & eosin, 40x)



Epidermis with mild spongiosis and dyskeratotic keratinocytes. Edematous upper dermis with neutrophils (Hematoxylin & eosin, 400x).





**Abstract N°: 2013****p16 and CDKN2A Correlation in Familial and Multiple Primary Melanoma: Preliminary Results of an Eastern-European Cohort Analysis**

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**Introduction & Objectives:**

Often referred to as p16 or INK4a/ARF, CDKN2A mutations represent a less studied circumstance in the framework of melanoma, compared to BRAF alterations. CDKN2A, a tumor suppressor gene, is inactivated by homozygous deletions located in the 9p21 chromosomal region, that result in cellular proliferation and dysregulation of pro-apoptotic pathways. This study aims to identify the correlation between clinically and pathologically detected familial and multiple primary melanomas, p16 immunoexpression and CDKN2A genetic testing results.

**Materials & Methods:**

A 5-year retrospective cross-sectional study was conducted on 23 patients with cutaneous familial or multiple primary melanomas. Tissue samples were re-evaluated by expert pathologists and the FISH method was applied to certified melanoma specimens. Nuclear and/or cytoplasmic distribution of p16 expression on 4 μm-thick sections was evaluated, with focus on those with immunohistochemical negativity. Moreover, dual-colour FISH for CDKN2A and chromosome 9 centromere was performed to establish percentage values for p16/CDKN2A deletion on formalin-fixed paraffin-embedded material. In a normal interphase nucleus, two orange and two green signals were expected. Specimens containing  $\geq 15$  nuclei that lacked both signals for CDKN2A (no green signal) and contained at least 1 signal for chromosome 9 centromere (at least one orange signal) were considered positive for homozygous deletion.

**Results:**

Among the analysed cases, 8 displayed homozygous deletion of CDKN2A and 10 samples presented loss of p16 immunoexpression. All CDKN2A-mutated specimens associated absent p16 immunoreaction, and – on the other hand – only 2 p16-negative melanomas were correlated with CDKN2A monosomy. In the genetically altered group, 62.5% of cases (n=5) encompassed histopathological characteristics of superficial spreading melanomas, backed by 25% nodular melanomas and 12.5% acral melanomas. The average Breslow index was  $6.65 \pm 3.61$  mm and 75% of them (n=6) were ulcerated. Moreover, the average mitotic rate afferent to this group was  $4.875 \pm 3.58$ /mm<sup>2</sup>. 50% of the CDKN2A-mut patients also displayed BRAF positivity. Furthermore, 87.5% of the cases with certified CDKN2A mutations presented with loco-regional and/or visceral metastases at the time of diagnosis, most commonly in the liver.

**Conclusion:**

This analysis highlights the quasi-perfect association between CDKN2A homozygous deletions and p16 absence at immunohistochemical evaluations, therefore establishing the value of the latter as a cost-effective test, useful to predict the aggressive biological behaviour and unfavourable prognosis in familial and/or multiple primary melanomas. These discoveries are believed to initiate tailored diagnostic and therapeutic algorithms for the Eastern-European population with CDKN2A-mutated melanomas.

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**Abstract N°: 2059**
**comparison of the Union for International Cancer Control (UICC) 8th edition of the TNM Classification of Malignant Tumours (pTNM) and the Brigham and Women's Hospital (BWH) staging system for staging high risk squamous cell carcinomas; a one year review**

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**Introduction & Objectives:**

Squamous cell carcinoma's (SCC) of the skin can be staged using two classification systems, the eighth edition of the Union for International Cancer Control (UICC) 8th edition of the TNM Classification of Malignant Tumours (pTNM) and Brigham and Women's Hospital (BWH) staging system. Some studies have shown the BWH to be superior to the pTNM in subclassifying high risk tumours, thereby minimizing the number of patients recommended for radiologic evaluation, close surveillance, and possible adjuvant therapy. Our goal was to evaluate high risk SCC cases at our institution and compare in greater detail the subclassification criteria used both the pTNM and BWH systems.

**Materials & Methods:**

All cases discussed at the high risk SCC multidisciplinary team meetings in 2022 were analysed. For each patient, the BWH and pTNM dataset criteria were recorded. Patient sex and age and site at diagnosis were recorded. All statistical analysis was performed using Microsoft excel.

**Results:**

In total, 86 surgical specimens were analysed, incorporating 81 patients. 63% (n=51) of patients were male and 37% (n=30) were female. The average age was 79.6 years.

68.6% (n=59) of specimens were from the head and neck, 27.9 % (n=24) were from the upper or lower limbs and 3.5% (n=3) were from the chest or back.

The BWH and pTNM staging systems were compared (Table 1).

Table 1

<b>TNM</b>	<b>% (n=)</b>	<b>BWH</b>	<b>% (n=)</b>
T1	29 (25)	T1	32.5 (28)
T2	24.4 (21)	T2A	33.7 (29)
T3	46.5 (40)	T2B	27.9 (24)
		T3	5.8 (5)

The BWH system appears to distribute high risk cases in a more standardised fashion when compared to the pTNM system.

For BWH T2A (n=29), the most common criteria used for this classification was size (75.8%, n=22).

For BWH T2B (n=24), the most commonly fulfilled criteria were size (87.5%, n=21) and poor differentiation (79.1%, n=19). 87.5% (n=21%) of these cases were diagnosed fulfilling 2 criteria, 16.6% (n=4) were diagnosed fulfilling 3 criteria.

For pTNM pT3 (n=40), 32.5% (n=13) were diagnosed by depth alone, with size of less than 4cm and no invasion beyond the subcutaneous fat. Only 45% (n=18) fulfilled the size criteria of pT3 and 20% (n=8) fulfilled the criteria of invasion beyond subcutaneous tissue.

Of the 40 pTNM pT3 cases, 12.5% (n=5) remained T3, 50% (n=20) were downgraded to 2B, 27.5% (n=11) were downgraded to 2A and 10% (n=4) were downgraded to 2A on the BWH system.

**Conclusion:**

The BWH classification system appears to distribute\*\* high risk SCC specimens in a more standardised fashion than the TNM system. This may allow for improved identification of patients in this overall category who would better benefit from radiological close follow up or adjuvant therapy.



**Abstract N°: 2215**

**Post-transplant lymphoproliferative disorder, monomorphic type, in a kidney transplant patient following administration of anti-influenza vaccine**

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Post-transplant lymphoproliferative disorder, monomorphic type, in a kidney transplant patient following administration of anti-influenza vaccine

**Introduction & Objectives:**

Post-transplant lymphoproliferative disorders occur in 1-12% of transplant recipients. Post-transplant lymphoproliferative disorders (PTLD) are iatrogenic immunodeficiency-associated lymphoid proliferations. They comprise a heterogeneous spectrum of diseases ranging from polyclonal hyperplasia to aggressive lymphomas. Their pathogenesis is largely unknown and closely related to low immunity, which allows uncontrolled lymphocytic proliferation. Most of these disorders arise from B cell lineage, however, a small subset involves T cell proliferations. In all PTLD subgroups, primary cutaneous involvement is rare.

**Materials & Methods:**

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**Results:**

We present the case of a 49-year-old female two kidney transplants recipient who developed painful nodules on the trunk, neck and axillae that appeared one day after receiving the influenza vaccine. On physical examination, tender and skin-colored lesions were found. Histopathology showed a diffuse infiltration of sheet-like collections of large cells, some of them binucleated, with marked nuclear atypia and several mitotic figures. Neoplastic cells reacted positively for CD2, CD3, CD30 and perforin and negative for EBV and ALK

The combination of these findings led to the diagnosis of anaplastic monomorphic T-cell PTLD.

**Conclusion:**

Despite being a rare complication, PTLD should be considered in the differential diagnosis of a primary cutaneous lymphoproliferative disorder in the setting of a kidney transplant recipient.

Although transplant patients receive annual influenza vaccination as a preventive protocol, to our knowledge we report the first case of PTLD coinciding with influenza vaccine. While the reaction might just be a coincidental occurrence of unrelated causes, more studies are still necessary to elucidate the molecular mechanism underlying a possible causal association.



**Abstract N°: 2255**

**TRIM 11 Fusion Melanocytoma**

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**Introduction & Objectives:**

A 41-year-old Asian female had a lesion for 3 years on the left arm. Her background included Raynauds. There was no preceding trauma. Examination demonstrated a 3cm warm and tender nodule with overlying hyperpigmentation. There was no lymphadenopathy.

**Materials & Methods:**

We present a rare case of TRIM 11 fusion melanocytoma presenting as a warm tender brown nodule.

**Results:**

An incisional biopsy showed fairly uniform spindle cells with focally prominent nucleoli and nests of polygonal cells with more clear cytoplasm. Melanocytic differentiation was suggested by immunohistochemistry; focal positivity of S100, HMB45, SOX10 and Melan-A. Negative for SMA, desmin, CD34, CK, SYN, CG, HHV8 and TFE3. There was minimal mitotic activity and no necrosis, proliferation index was low (Ki67); suggesting melanoma or metastasis unlikely. Reverse transcription-polymerase chain reaction was negative for common clear cell sarcoma gene changes. Full excision showed a subcutaneous nodule of 13mm depth extending into the dermis and subcutis. Radial margin 5mm and deep margin 0.4mm. Lobules of large polygonal cells with vesicular nuclei and prominent nucleoli were noted. Nuclear positivity to antibody directed against TRIM11 as well as a heterogeneous positivity of NTRK1 were demonstrated. Fluorescence in situ hybridization confirmed the presence of a TRIM11 fusion.

**Conclusion:**

A diagnosis of cutaneous melanocytoma with TRIM11 rearrangement was made. Our patient completed 2-year follow-up with no signs of recurrence or metastasis. The longterm behaviour of these tumours is uncertain. There are 13 cases in the literature, one of which was shown to recur and metastasize after 13 years (1).

**References:**

1. Parra O, Linos K. Cutaneous Melanocytic Tumor with CRTCL1:TRIM11 Fusion: Review of the Literature of a Potentially Novel Entity. *Biology (Basel)*. 2021 Dec 7;10(12):1286. doi: 10.3390/biology10121286. PMID: 34943200; PMCID:



**Abstract N°: 2269**

**Spindle cell predominant trichodiscoma (Neurofollicular hamartoma)**

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**Introduction & Objectives:**

Multiple asymptomatic facial papules are relatively common complaint. Importance of reaching a precise diagnosis of these lesions is relevant for proper management of the patients.

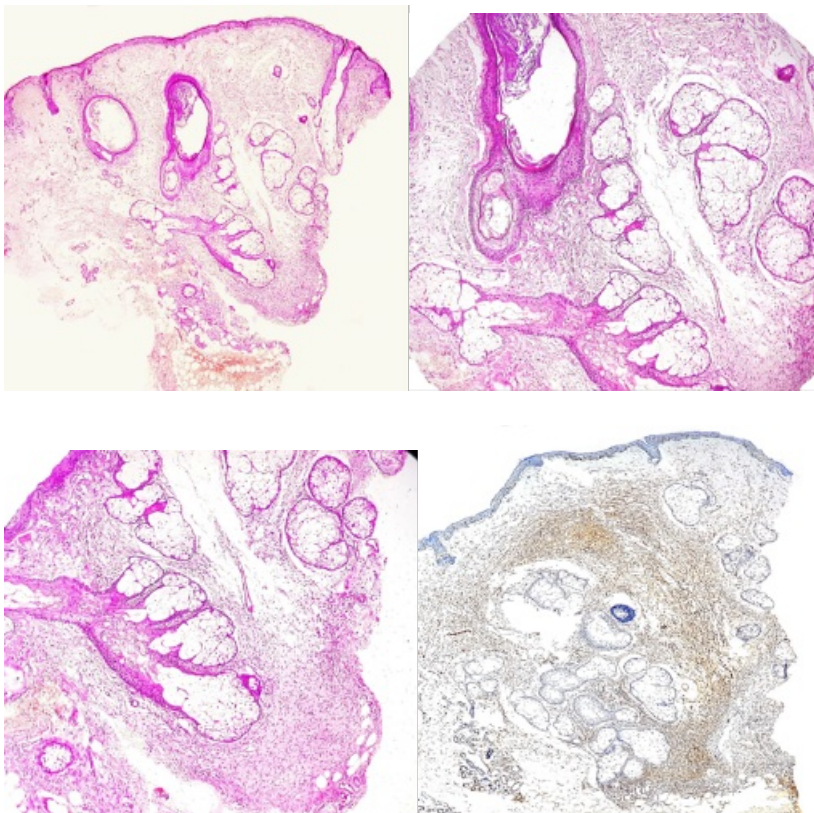
**Materials & Methods:**

A 52 year old female patient presented by 8 year history of multiple asymptomatic facial papules. Lesions are skin coloured dome shaped papules located predominantly on lower face.

Biopsy was taken from one of the papules and examined histopathologically.

**Results:**

Examination of the biopsy revealed a distorted pilosebaceous unit with enlarged sebaceous glands surrounded by spindle cell proliferation. Immunohistochemical staining with S100 revealed positive staining of the spindle shaped cell. Fig 1



The diagnosis of Spindle cell predominant trichodiscoma (Neurofollicular hamartoma) was confirmed.

Patient was instructed to undergo imaging for chest and kidney. The investigations revealed presence of multiple lung cysts. This leads to diagnosis of Birt Hogg Dube syndrome.

**Conclusion:**

Neurofollicular hamartomas, trichodiscomas and fibrofolliculomas lie in the same hamartomatous spectrum. Birt Hogg Dube syndrome is an autosomal dominant disease. Mutations in the FLCN gene making a protein called folliculin. Symptoms of BHD generally do not appear until adulthood.

Association of with multiple non-cancerous (benign) skin tumors, particularly on the face, neck, and upper chest. These growths typically first appear in a person's twenties or thirties . Lung cysts, spontaneous pneumothorax, emphysema and kidney tumors (benign and malignant).

Patients diagnosed with Birt Hogg Dube syndrome need screening with :

1- Renal MRI

2- Chest CT

Also patients are instructed to avoid Cigarette smoking to minimize possibility of developing lung lesions.



**Abstract N°: 2472**

**KSD: a manually curated database for regulation of keloid sensitivity regulation factors**

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**Introduction & Objectives:**

**Background:** Keloids is a fibroproliferative dermal tumor, of which the etiology is unknown. Early and effective diagnosis of keloid using diagnostic biomarkers can be helpful in the development of new personalized treatments. Therefore, a thorough and comprehensive understanding of the diagnostic biomarkers in keloid is of great importance. **Objectives:** Build a database that catalogues information on the sensitivity regulation factors associated with keloid.

**Materials & Methods:**

We collected all the literature related to keloid in PubMed before 10 September 2022. A totally 3621 papers were downloaded. By manually proofread the abstract, multiple sensitivity regulation factors were collected. To ensure the unification of factors curated from literature, some other databases and tools were used for annotation. Then all the information is integrated into the database KSD. By manually curating thousands of published literatures, multiple sensitivity regulation factors [134 drugs, 275 proteins, 92 RNAs, 32 genes, 27 cells, 13 pathways and other records] were recorded with appropriated annotation. To illustrate the value of the data we collected, data mining was performed including functional annotation and network analysis.

**Results:**

Multiple sensitivity regulation factors (134 drugs, 275 proteins, 92 RNAs, 32 genes, 27 cells, 13 pathways and other records) were identified from the papers. To illustrate the value of the data we collected, data mining was performed including functional annotation and network analysis. The most enriched pathway is "Pathways in cancer". All the data can be searched and downloaded on KSD (<http://ksd2023.cn/>).

**Conclusion:**

In summary, KSD provides a practical and user-friendly web-based tool that may serve as a specialized platform for clinicians and researchers to better understand the detailed mechanisms of keloid sensitivity.



**Abstract N°: 2769****Correlation between clinical and histological features of dermatopathic lymphadenopathy: Analysis of 39 lymph nodes**Damla Demir<sup>1</sup>, Aysenur Ihvan<sup>2</sup>, Ezgi Aktaş\*<sup>3</sup>, Ebru Zemheri<sup>2</sup>, Ilkin Zindanci<sup>1</sup><sup>1</sup>Umraniye Training and Research Hospital, Dermatology and Venereology, Türkiye, <sup>2</sup>Umraniye Training and Research Hospital, Pathology, Türkiye, <sup>3</sup>Prof Dr Cemil Taşçıoğlu City Hospital, Dermatology and Venereology, Türkiye**Introduction & Objectives:**

Dermatopathic lymphadenopathy is a well-defined histologic entity with an underestimated prevalence in the general population. It is commonly associated with chronic skin diseases, presented as enlarged lymph nodes with paracortical hyperplasia, however in some cases the lymph nodes may remain at a normal size and may not be associated with other system disorders rather than skin diseases. The present study was conducted to analyse the characteristics of histologically diagnosed cases of dermatopathic lymphadenopathy at our unit. We also aimed to investigate any association between the clinical features of the patients and the histological findings.

**Materials & Methods:**

A total of 39 lymph node samples from 39 patients diagnosed with dermatopathic lymphadenopathy were included in this prospective cohort study.

**Results:**

Thirty-four (87%) patients had a dermatologic disorder. The presence of paracortical eosinophils were significantly higher in the patients with dermatologic disorders ( $p = 0.001$ ) while the presence of dilated sinuses was significantly more common in the patients without a dermatologic disorder ( $p = 0.035$ ). The presence of dilated sinuses and medullary histiocytes were significantly more common in the patients with lower body surface area involvement of the disease compared to the ones with a higher body surface area ( $p = 0.003$ ,  $p = 0.034$ ; respectively).

**Conclusion:**

Most of the patients included in the study had one of a broad spectrum of undiagnosed dermatologic disorders. The clinical significance of the relation between histologic and clinical findings in dermatopathic lymphadenopathy remains to be explained. Dermatopathic lymphadenopathy should always be considered in differential diagnoses of patients with persistent lymph node enlargement even when absolute dermatologic disorders are not present. Since various skin disorders may be the cause of lymphadenopathy, performing a full-body examination before lymph node excision might prevent unnecessary procedures.





**Abstract N°: 2842****Presence of amorphous masses is more common in dermatoscopically classical actinic keratosis than more advanced lesions**

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**Introduction & Objectives:**

Actinic keratosis (AK) is a common lesion on sun exposed sites of light skinned individuals and is a hallmark of cumulative sun damage. Meanwhile a histologic hallmark of cumulative UV damage is solar elastosis that when most prominent is seen as amorphous masses of degraded elastotic material.

**Materials & Methods:**

To compare the presence of clinical and dermatoscopic signs of AK and intraepithelial carcinoma (IEC) with the presence of amorphous masses in histopathology we included 29 patients with the mean age of 78 years. Clinically, we counted presence of subjective symptoms, induration (papular AK), diameter larger than 1cm, rapid enlargement, bleeding, erythema, and ulceration. Dermatoscopically, we assessed presence of white starburst pattern, white areas, milky areas, and vascular structures as glomerular, hairpin, linear etc. Histopathologically presence of amorphous masses was assessed with the light microscope and routine haematoxylin and eosin stain. Statistical analysis was performed with SPSS.  $P < 0.05$  was considered statistically significant.

**Results:**

According to histopathology, our sample consisted of atrophic AK (N=3), intermediate AK (N=19), Bowenoid AK (N=3) and SCC in situ (N=4). Amorphous masses were present in 72.4% (N=21) of cases. Patient age was not significantly higher in the group with amorphous masses than without this histologic feature (79.0 vs 75.5 years respectively,  $p=0.15$ , t-test for equality of means). We identified no clinical signs that were more common in cases with amorphous masses. Nevertheless, in our sample all except one dermatoscopically classical AK had amorphous masses (92.3%) in comparison with nine (56.2%) lesions in AK/SCC group that had dermatoscopically suspicious features ( $p=0.044$ , Fisher's Exact test). Three of seven Bowenoid AK or SCC in situ lesions (42.9%) did not have amorphous masses.

**Conclusion:**

This study shows an interesting finding that the presence of amorphous masses is more common in dermatoscopically classical AK in comparison to more advanced lesions. This supports the association of cumulative sun damage and AK and leaves space for other additional mechanisms needed for progression after certain threshold of UV damage is acquired.

**Abstract N°: 2929****Cutaneous Schwannomas: Clinical Manifestations, Diagnosis and Management: A Narrative Review**Elio Assaad Saad<sup>1</sup>, Myriam Bouer<sup>2</sup>, Leah Nasr<sup>2</sup>, Ala'a Fawaz<sup>3</sup>, Nancy Emmanuel<sup>4</sup>

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**Introduction & Objectives:** Cutaneous Schwannomas are the most common benign peripheral nerve sheath tumour; they usually present as asymptomatic skin-coloured nodules, are mostly common on the head and neck area but can present in other areas as well. They are rarely diagnosed on ultrasound, and diagnosis is usually confirmed through histopathology and immunochemistry. The treatment is by surgical excision, particularly if symptomatic. This is, to the best of our knowledge, the first in-depth review of the literature on cutaneous schwannomas.

**Materials & Methods:** A literature review was conducted by looking up “Cutaneous Schwannoma” on PubMed. This search resulted in 18 articles, of which 14 were found to be relevant and met the inclusion criteria and thus were reviewed extensively for the synthesis of this study. We also screened the references of selected articles and 15 additional relevant articles were included for discussion purposes.

**Results:** Of the 21 reported patients with cutaneous schwannomas in the literature, 11 were males and 10 were females. In terms of age, 33% of the cases occurred in patients younger than 25 years of age, 29% of the cases occurred in patients who are between 25 years and 50 years of age and 38% of the cases were of patients above 50 years old. The cutaneous schwannomas were mostly nodular in shape, with a firm consistency, and they varied in size, ranging from 0,5 cm to 14 cm in their largest diameters. In terms of location, 62,5% were in the head and neck area, 12,5% were located on lower limbs, 4,2% on upper limbs, and 20,8% on the trunk. Diagnosis was confirmed by histopathology in all cases, with mainly 2 histologically different tissue types described: Antoni type A and Antoni type B tissues. Antoni type A tissues are highly cellular, and the cells tend to align forming palisades of nuclei piling up, separated by Verocay bodies, whereas Antoni type B tissues represent myxoid edematous areas that alternate with Antoni type A tissues. Immunohistochemically, strong S-100 protein expression and intensity are relevant findings in the diagnosis of cutaneous schwannomas. Treatment was mostly through surgical excision when indicated, however one of the reported cases refused surgery and instead was treated by tumescent suction technique. There was no mention of recurrence in any of the cases.

**Conclusion:** Cutaneous schwannomas are a rare type of cutaneous tumor with only 21 reported cases in the literature, they seem to occur equally in both genders and can present on various locations of the body but are most common in the head and neck area; they are usually encapsulated, exhibit Antoni type A and B tissue patterns and are S-100 positive. Using different modalities, including ultrasound, histological and immunochemical studies, cutaneous schwannomas can be diagnosed and differentiated from other tumors with a similar presentation. Treatment, when indicated, is by surgical excision, with no apparent risk of recurrence.

**Abstract N°: 3200****A scoping review evaluating the effect of the provision of clinical information on the histopathologic diagnosis of cutaneous melanoma**

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**Introduction & Objectives:** The Cancer Council Australia guidelines on "What clinical information should the clinician give the pathologist to aid diagnosis of melanoma?" advise that additional clinical information such as specimen type, clinical diagnosis and dermoscopic images could usefully inform the diagnosis of melanoma. This scoping review aimed to update the available evidence on the impact of the provision of clinical information on the histopathology diagnosis of cutaneous melanoma.

**Materials and Methods:** We screened all references that were retrieved in the Cancer Council Australia guidelines' 2018 evidence review, and searched PubMed, Embase and Cochrane Library databases for new records up to September 2022. We also undertook forward and backward citation searches of 14 studies included in the Cancer Council's evidence summary. Eligible studies needed to have histopathology diagnosis of melanocytic skin lesions and evaluate clinical information that was routinely available to the requesting clinician. A qualitative synthesis of results was performed.

**Results:** From 1875 records screened, 163 references underwent full-text review, and seven articles were included. Four articles were diagnostic agreement studies, two were a retrospective cohort studies, and one was a case-control study. Five studies demonstrated that the provision of clinical information led to changes in diagnosis. One study showed that in 68/408 interpretations, the introduction of dermoscopic images yielded an upgrade in diagnosis from low grade to high grade atypia or from high grade atypia to melanoma, compared to 8/136 interpretations resulting in a downgrade of atypia. All four diagnostic agreement studies found that dermoscopic images increased interobserver agreement, whereby one study demonstrated that the overall Cohen's K for cases studied without dermoscopy was 0.813, however improved to 0.882 with the use of dermoscopy. Similarly, another study found that the Fleiss' kappa amongst all three dermopathologists increased from 0.447 without dermoscopy to 0.497 with dermoscopy. One study found that when pathologists increasingly had more access to clinical information in five step procedures from 1- no information; 2- age/sex/location of the lesion; 3- clinical diagnosis; 4- clinical image and 5- dermoscopic image, there was a linear increase in the level of diagnostic confidence (p for linear trend  $p < 0.001$ ). One case-control study showed that reporting the anatomical site, suspected clinical diagnosis and laterality of the lesions significantly decreased the odds of a false-negative diagnosis by 0.16, 0.46 and 0.41 respectively. In addition to the above clinical factors, also reporting the specimen type, history of melanoma, history of lesional trauma or treatment on the pathology request form significantly reduced the odds a false-positive diagnosis.

**Conclusion:** Provision of relevant clinical information may influence pathologists' diagnoses of melanocytic lesions.

32<sup>ND</sup> EADV Congress 2023  
11 OCTOBER - 14 OCTOBER 2023  
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**Abstract N°: 3215****Clinical and histopathological features of inverted follicular keratosis**

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**Introduction & Objectives:**

Inverted follicular keratosis is a rare benign tumor of follicular infundibulum usually present as a solitary, flat topped or verrucose, pigmented or skin color, asymptomatic papule less than 1 cm in diameter. It almost always occurs on hairy skin. Histologically characterized by invaginated proliferation of mature squamous cells and presence of squamous eddies.

Four growth patterns have been described; a wart-like, Keratoacanthoma-like, Solid nodular and cystic variants. Most of the lesions are treated with surgical excision. In this cross-sectional study all the patients with diagnosis of inverted follicular keratosis who were referred to our center in a period of 4 years were evaluated clinically and histologically.

**Materials & Methods:**

The medical records of patients with histologically confirmed inverted follicular keratosis were gathered to determine demographic data, clinical information, pathological characteristics and the type of treatment received by the patients.

**Results:**

Fifty-six patients were included in this study. Mean age of patients was  $50.71 \pm 19.76$  with minimum of 10 and maximum of 82 years old. It is mostly found in age of over 50 years old and prevalence increased with increase in age especially in lower extremity and upper extremity. Most patients were male (73.2%). Size of lesions varied between 0.3 to 3 cm with mean size of 1.01 cm. Most lesions were in a papular form (41.1%) and located on face (57.1%). Verrucose and ulcerated lesions were less common. Involvement of lower extremity was more prevalent than upper extremity and trunk and latter involvement is only seen in male patients. In only one patient neck was involved. Multiple lesions were observed in 3 patients. Duration of lesion was below 3 months in majority of patients.

For patients younger than 30 years old, most of their lesions (60%) lasted for 6-12 months Also, patients over 50 years had most of their lesions for less than 3 months. (71.4%) ( $p < 0.05$ ).

Squamous eddies (80%), Exo-endophytic growth (50%), endophytic predominance (37.5%), papillomatosis (35.7%), parakeratosis (79%), hypergranulosis (34%), nuclear atypia (29%), keratin plug (25%), dyskeratotic cell (62.5%), coarse keratohyalin granules (33.9%), cystic structure (5%) and melanin pigment (23.2%) was observed. Approximately 86% did not have acantholysis, 98.2% did not have marginal buttress formation, 72% did not show mitosis and 26.8% of patients did not have any dermal inflammation. The most common type of cell found in dermal inflammation was lymphocytes.

**Conclusion:**

Inverted follicular keratosis is a rare benign tumor that can have unusual clinical and histological features. Biopsy



is warranted to confirm the diagnosis and plan the best treatment.

**32<sup>ND</sup> EADV Congress 2023**  
**11 OCTOBER - 14 OCTOBER 2023**  
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**Abstract N°: 3552**

**Voluminous scrotal mixed cystic lymphatic malformation mimicking a sarcoma: An uncommon presentation**

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**Introduction :**

Cystic lymphatic malformations are caused by lymphatic developmental abnormalities, resulting in abnormal vessels and cystic dilatations. These malformations can be classified as macrocystic, microcystic, or mixed and may be congenital or acquired. We report a case of a patient with a mixed lymphatic malformation that was superinfected with Koch's bacillus.

**Case report:**

We present a case of a 21-year-old patient with an asymptomatic bilateral inguinoscrotal swelling that had progressively increased in size since the age of 8. The patient reported weight loss and anorexia for 2 months. Upon clinical examination, a voluminous, hard subcutaneous inguinoscrotal mass was identified on the right side, extending towards the contralateral side, measuring approximately 15 cm in size, and presenting with multiple translucent vesicles. Dermoscopy revealed lacunae with a reddish hypopyon appearance associated with pinkish-white areas. Thoracic-abdominal-pelvic scan revealed a tumor process covering the entire pelvis and active lesions of pulmonary tuberculosis. Surgical biopsy revealed a caseo-follicular tuberculosis with cystic lymphangioma lesions. After completing anti-tuberculosis treatment, the mass became soft and mobile. The patient underwent sclerotherapy with bleomycin and was a candidate for treatment with sirolimus.

**Discussion:**

Scrotal localization of cystic lymphatic malformations is uncommon, making differential diagnosis with condyloma difficult. These lesions typically appear at birth or in early childhood but can also be acquired and develop later in life due to trauma, surgery, radiotherapy, or certain infections. Although a few case reports have described the presence of acquired scrotal microcystic malformations of idiopathic origin, histology is essential for orienting the diagnosis and guiding management. To our knowledge, this is the first case of a mixed lymphatic malformation presenting at an advanced age and complicated by a tuberculosis infection. This infection resulted in significant inflammation of the lymphatic vessels, with the development of secondary fibrosis, which mimicked a sarcoma on imaging. Surgical biopsy allowed for diagnosis and appropriate management. The management of mixed lymphatic malformations is challenging due to the presence of a double component, especially in a location as particular as that of our patient. Sclerotherapy can induce regression of the macrocystic component, while m-TOR inhibitors have immunosuppressive, antiproliferative, anti-angiogenic, and lymphangiogenic properties and have been shown to provide satisfactory responses to diffuse lymphatic malformations in the literature.

**Conclusion:**

Mixed lymphatic malformations can become superinfected, leading to the development of significant fibrosis, which can be responsible for a giant, hard, and fixed mass mimicking a malignant pathology. Surgical biopsy is crucial in orienting the diagnosis and guiding the therapeutic management.

**32<sup>ND</sup> EADV Congress 2023**  
**11 OCTOBER - 14 OCTOBER 2023**  
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**Abstract N°: 3992****Different faces of Prurigo pigmentosa. Report of 3 cases from Ukraine.**

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**Introduction & Objectives:**

Prurigo pigmentosa (PP) is a rare disease of unknown etiology characterized by itchy red rash and reticulate darkening of the skin. It was described as dermatitis that mostly affected Japanese women and was believed to be endemic in Japan. However, PP is underdiagnosed in the other countries due to a lack of awareness among healthcare professionals. The number of reported cases has been increasing, suggesting that the disease's geography is wider than previously believed. The objective of this case report series is to underline different clinical and morphological features and highlight the importance of clinicopathological correlation.

**Materials & Methods:**

We present 3 cases to demonstrate the full range of the clinical and pathological features of PP. 2 patients were females in good health although slightly underweight, and the third was a male who had undergone recent weight loss. All the patients experienced itchy rash that eventually resulted in a net-like pigmentation.

**Results:**

A 23-year-old female presented with a 10-year history of itchy red patches and occasional blisters in her axillae, under her breasts and groin. She noticed flare-ups after wearing synthetic cloth and during pregnancy. On examination, multiple red patches, papules and postinflammatory hyperpigmentation were noted. A biopsy showed a superficial mixed infiltrate, with small intraepidermal pustules, and clusters of necrotic keratinocytes. These findings were consistent with PP. The patient declined doxycycline as a treatment option because she was breastfeeding. She is being closely monitored with regular follow-up.

A 16-year-old female presented with a 3.5-year history of pruritic inflammatory dermatosis. Physical examination revealed a symmetrical rash over the back and posterior neck. It comprised papules and papulovesicles coalescing into reticulated pattern. Biopsy included late-stage and newly developed elements. In the early stages, there was a mixed perivascular infiltrate. The epidermis showed mild spongiosis, and a few basal layer keratinocytes were necrotic. A fully developed lesion contained a dense infiltrate containing neutrophils, eosinophils, and lymphocytes. Lymphocytic exocytosis and conspicuous cytoid bodies in all layers of the epidermis were noted. A late-stage lesion showed a mild perivascular infiltrate with numerous melanophages. Based on these findings, a diagnosis of PP was confirmed. Treatment with dapsone was effective.

A 25-year-old male presents with an intensely itchy rash on the back and sides of the trunk. The rash exhibits a reticular pattern and left behind net-like hyperpigmentation. Over the past month, he intentionally lost weight by following a ketogenic diet. A biopsy revealed vacuolar degeneration of isolated keratinocytes and the presence of cytoid bodies. The dermis showed a moderate perivascular infiltrate with scattered melanophages. Considering the patient's clinical symptoms and medical history, these changes were consistent with PP.

**Conclusion:**

The pathogenesis of the disease remains unclear, but it is often associated with ketosis and a restrictive diet. It may resolve spontaneously or improve with an increase in carbohydrate intake. Our study of 3 patients diagnosed with PP showed a wide range of clinical and morphological features. Due to its rarity and varied clinical presentation, it can easily be underrecognized. Early diagnosis is critical to avoid unnecessary treatment and recurrences.

**32<sup>ND</sup> EADV Congress 2023**  
**11 OCTOBER - 14 OCTOBER 2023**  
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**Abstract N°: 4059****Dermatofibroma and Dermatomyofibroma. Two different myofibroblastic tumors in a patient -A rare case of multiple dermatomyofibroma with dermoscopic findings**

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**Dermatofibroma and Dermatomyofibroma. Two different myofibroblastic tumors in a patient -A rare case of multiple dermatomyofibroma with dermoscopic findings and literature review****Introduction & Objectives:**

Dermatomyofibroma is an rare benign myofibroblastic tumor and typically presents as a single lesion in adult women. However, cases with multiple dermatomyofibromas have been rarely reported. To the best of our knowledge, in the literature, our patient is the fourth case with multiple dermatomyofibromas, in addition, a unique case that presented with two different myofibroblastic tumors (Dermatofibroma and dermatomyofibroma).

**Materials & Methods:**

A 28 year-old female patient presented with multiple overgrowing, nodular lesions on her back and both shoulders since four years. Excisional biopsy was performed for three lesions by Plastic and Reconstructive surgery Department. The first lesion on back was resulted as dermatofibroma with high proliferation index (ki-67 was 7%). Other two lesions that excised from shoulders resulted as hypertrophic scar. Intralesional corticosteroid injection was performed for 3 sessions. After 5 years, she applied with 3 other new lesions (2 on right hemithorax and 1 on posterior cruris). On dermatological examination, there were hyperthrophic scars on excision areas. There were 0,3 to 0,5 cm, pinkish-brown new papules on her back. In addition, there was a new 1cm diameter, brown nodule on her posterior cruris. She had diagnosed with hypothyroidism and there was no other systemic disease or drug history. Dermoscopic examination revealed pink-brown structureless area on papules on her back. On the other hand, we detected central scar-like area on the nodue on her leg which suspected us it could be a dermatofibroma. Excisional biopsy was performed for the 2 lesions on back with leiomyoma and dermatofibroma preeliminary diagnoses and single lesion on leg as dermatofibroma. Two lesions on back diagnosed with hypertrophic scar and the nodule on leg as dermatofibroma.

**Results:**

However, the history and clinical examination of the patient were not consistent with the diagnosis since she had no history of trauma or surgery. In view of many diagnostic hypotheses, such as leiomyoma, dermal fibromatosis, dermatofibrosarcoma protuberans, eg., we requested immunohistochemistry evaluation. Immunohistochemistry revealed focally positive smooth muscle actin and negative desmin, CD34 and S100. According to histopathological and clinical findings, we suggested that our case is compatible with dermatomyofibroma.

**Conclusion:**

Dermatomyofibroma is a myofibroblastic benign tumor that generally located on shoulder, axilla, neck and upper trunk. It is an uncommon condition and multiple form of dermatomyofibroma is extremely rare. In the literature, we found three cases with multiple dermatomyofibromas. Diagnosis of dermatomyofibroma can be challenging for both pathologist and dermatologist. It can be confused with hypertrophic scar, keloid, leiomyoma, as well as

other myofibroblastic originated tumors.

**32<sup>ND</sup> EADV Congress 2023**  
**11 OCTOBER - 14 OCTOBER 2023**  
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**Abstract N°: 4489****Expanding the spectrum of skin neoplasms in Muir-Torre Syndrome: beyond sebaceous tumours**

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**Introduction & Objectives:**

Muir-Torre syndrome (MTS) is a hereditary disorder characterized by internal malignancies and sebaceous tumours. Diagnosis of MTS is based on clinical features, family history and genetic testing. Two cases of MTS-associated actinic keratosis (AK) and cutaneous squamous cell carcinoma (SCC) have been reported to date. We present two patients with genetically confirmed MTS who presented with AK and SCC, which had loss of the same DNA mismatch repair genes seen in the patients' internal malignancies and sebaceous tumours.

**Materials & Methods:**

Case presentation and literature review.

**Results:**

The first patient, a 71-year-old man consulted for a 6-month erythematous, hyperkeratotic plaque on the right hand. He had previously been diagnosed with MTS-related two colorectal adenocarcinomas, a jejunal adenocarcinoma and a sebaceous adenoma. Histological examination showed keratinocytic dysplasia of the lower layers of the epidermis, parakeratosis and solar elastosis, and was diagnosed as an AK.

The second patient, an 80-year-old woman with a history of MTS-related two colorectal adenocarcinomas, an endometrial adenocarcinoma, a sebaceous carcinoma and two keratoacanthomas, presented with a hyperkeratotic nodule on the right forearm. The biopsy showed keratinocytic dysplasia, altered maturation and horn pearls with extension into dermis, and was diagnosed as an invasive SCC. No sebaceous differentiation was found. One margin of the lesion showed AK.

Immunohistochemistry showed loss of expression of MLH1 and PMS2 repair genes in the first patient and loss of PMS2 and MSH6 in the second patient, with normal expression in adnexal structures and normal surrounding skin.

**Conclusion:**

Reduced DNA mismatch repair (MMR) protein expression in keratinocytes have been related to the development of SCC and precancerous skin lesions in MTS-unrelated skin cancers. However, our cases suggest that patients with MTS may be at increased risk of developing AK and SCC. These lesions are not typically associated with MTS and may be overlooked during dermatologic evaluations. Therefore, it may be worthwhile to consider testing skin lesions for MMR gene mutations in patients with a history of cancer or other risk factors for cancer, even if they do not meet the criteria for a specific hereditary cancer syndrome.



**Abstract N°: 4490****Interesting cases of atypical melanocytic lesions at OOOOOO Hospital : Spitz nevus and pigmented epithelioid melanocytoma**Hyungkeon Park\*<sup>1</sup>, Sook Jung Yun<sup>1</sup><sup>1</sup>Chonnam National University Hospital, Dermatology, Gwangju, Korea, Rep. of South**Introduction & Objectives:**

Spitz nevus composed of large epithelioid and spindled cells and pigmented epithelioid melanocytoma (PEM) composed of heavily pigmented epithelioid and dendritic cells are uncommon atypical melanocytic neoplasms. These tumors are infrequent in Korean population. This study aims to analyze the clinicopathological characteristics of Spitz nevus and PEM in Korean from our institution.

**Materials & Methods:**

Thirty-four patients with Spitz nevus and 6 patients with PEM diagnosed by skin biopsy from January 2006 to March 2023 were retrospectively analyzed. Data on age, sex, duration, symptoms, and clinical manifestations were obtained from medical records and photographs. Spitz nevus were classified as Spitz nevus, pigmented Spitz nevus, and other variants of Spitz nevus. Atypia, epidermal hyperplasia, Kamino body, pigmentation, and deep mitosis were evaluated in histopathological slide reviews of Spitz nevus, except pigmented spindle cell nevus.

**Results:**

In Spitz nevus, the mean age of onset was 17.3 years, the male/female ratio was 15:19, and the mean duration was 21.8 months. Most patients were asymptomatic (82.4%, n = 28/34), and the most frequent manifestations were black colored (47.1%, n = 16/34) and papule (67.6%, n = 23/34). The most common site was lower extremities (35.3%, n = 12/34), and the mean size of lesions was 0.9 cm. Histopathologically, they were classified as pigmented Spitz nevus (n = 13/34), Spitz nevus (n = 10/34), and variant Spitz nevus (9 pigmented spindle cell nevus, 1 agminated, and 1 desmoplastic Spitz nevus). In Spitz nevi, except pigmented spindle cell nevus, they were 23 compound melanocytic nevus and 2 intradermal melanocytic nevus, and they showed epidermal hyperplasia (n = 22/25), Kamino body (n = 14/25), pigmentation (n = 13/25), and deep mitosis (n = 1/25). In 9 pigmented spindle cell nevus, the mean age of onset was 26.3 years, the male/female ratio was 3:6, and the mean duration was 22.0 months. Most patients were asymptomatic (88.9%, n = 8/9), and the most frequent manifestations were black colored (88.9%, n = 8/9), macule and papule (each 44.4%, n = 4/9), and the mean size of lesions was 1.1 cm. In addition, among 6 PEM, the mean age of onset was 11.9 years, the male/female ratio was 2:4, and the mean duration was 41.0 months. Most patients were asymptomatic (66.7%, n = 4/6), and the most frequent manifestations were black colored (66.7%, n = 4/6) and nodule (33.3%, n = 2/6), and the mean size of lesions was 1.3 cm.

**Conclusion:**

This results will be helpful in understanding and diagnosing patients with atypical melanocytic neoplasms.

**Abstract N°: 5257**

**Motif arc-en-ciel : une caractéristique dermatoscopique rare du lichen plan**

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**Introduction :**

The “rainbow” dermoscopy pattern was first described in 2009, as a reddish-blue staining associated with different colors of the rainbow in kaposi sarcoma patients.

This pattern was initially linked exclusively to the Kaposi’s disease, then this has been the subject of much discussion over the years. The current literature describes this pattern in various pathologies other than Kaposi’s disease. We report the case of a patient who presented a rainbow pattern in dermoscopy of palmoplantar lichen planus lesions

**Observation:**

A 60-year-old patient consulted for pruritic palmoplantar lesions, which had been evolving for 5 years.

The clinical examination showed an erythematous acquired palmoplantar keratoderma, which was bilateral and symmetrical , mainly on areas of pressure.

The Dermoscopic examination showed the presence of an erythematoviolin background, surmounted by an irregularly distributed whitish scales, as well as rounded yellow areas. We also note the presence of

a rainbow pattern in some areas.

A skin biopsy was realized objectivating a Lichen planus.

**Discussion:**

The “rainbow” pattern has been described in various diseases especially in cases of melanoma, blue nevus angiokeratomas, dermatofibromas, botriomycomas, stasis dermatitis and several other pathologies, and much more rarely in lichen planus lesions.

The “rainbow” pattern is caused by the existence of a double component : a vascular one, made of large lumen and thin wall capillaries associated to a cellular proliferation, whether keratinocytic, melanocytic, fibroblastic or other.

The authors explain this through optical phenomena, by comparing the skin to a diffraction grating. In fact, when light is emitted by the dermoscope crosses this double component, it is going to be divided into different wavelengths of light, which will be responsible for this amalgam of colors.

In the case of the lichen planus, this pattern could be explained by an acanthosis, hyperkeratosis or hypergranulosis associated with a dilatation of the dermal vessels.

The “rainbow” pattern is therefore an optical phenomenon, which is not specific to any etiology, therefore it is necessary to look for and identify the other associated dermoscopic structures in order to orient the diagnosis.

**Conclusion:**

In our case, we have objectified the presence of yellow rounded areas on a purple background, these are two structures that have been described in the literature on the palmoplantar lichen planus lesions, which led us to think first of this diagnosis.

32<sup>ND</sup> EADV Congress 2023  
11 OCTOBER - 14 OCTOBER 2023  
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**Abstract N°: 5561**

**Multinucleated cell angiohistiocytoma: a case series**

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Multinucleated cell angiohistiocytoma: a case series

**Introduction & Objectives:**

Multinucleated cell angiohistiocytoma (MCA) is an uncommon benign vascular and fibrohistiocytic proliferation, which occurs most often in women between 40 and 70 years of age.<sup>1</sup>

**Materials & Methods:**

We report a series of 6 patients. All patients presented with single to multiple asymptomatic erythematous-violaceous lesions with well-defined borders.

**Results:**

The average age of presentation was 60 years (ranging from 51-66 years) and 3 patients (60%) were female. Multiple lesions were present in 3 patients (60%). Regarding the location of the lesions, two (40%) were located on the dorsal aspect of a finger, two (40%) on the anterior aspect of the thighs and 1 (20%) on the dorsal aspect of the hand. The most frequent clinical diagnosis was annular granuloma (considered in 3, 60% of the cases). Due to the benign evolution of the condition, all patients refused treatment.

Histopathologically findings include vascular hyperplasia associated with an increased number of factor XIIIa-positive fibrohistiocytic interstitial cells and multinucleated cells with angular contours located in the dermis. Although not pathognomonic of MCA, the presence of multinucleated giant cells is the most specific histopathological finding.<sup>1</sup> On immunohistochemistry, the multinucleated cells are stained by CD68 and vimentin. Mononuclear dendritic cells are positive for vimentin, factor XIIIa, MAC387, and lysozyme. Endothelial cells, in turn, are positive for vimentin, CD31, CD34, and factor VIII.

**Conclusion:**

Differential diagnosis includes fibrous papules of the nose, Kaposi sarcoma, and atrophic dermatofibroma. Staining for human herpesvirus 8 is negative. <sup>2</sup>

Given the benign nature of this entity, patients often refuse treatment. Described treatments include physical modalities, such as cryotherapy, ablative CO2 laser, pulsed dye laser and Nd:YAG 1064. Rare cases of spontaneous resolution have been reported.

**Abstract N°: 5573****A painful tumor in a woman's forearm: a clinicopathological diagnosis**

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A painful tumor in a woman's forearm: a clinicopathological diagnosis

**Introduction & Objectives:**

Acquired elastotic hemangioma, first described in 2002, is an uncommon benign vascular proliferation that classically presents in older individuals on sun-exposed sites.

**Materials & Methods:**

The authors present the case of a 65-year-old-woman, with a known history of a biliary tract carcinoma, that presented to the dermatology department for a painful erythematous tumor on her left forearm.

**Results:**

Clinical examination disclosed an 8x7mm solitary red plaque, with well-defined borders. With the clinical hypothesis of an aneurysmatic dermatofibroma, excision of the lesion was scheduled. On low magnification view, the biopsy showed a narrow band of the papillary dermis uninvolved by underlying pathology (grenz zone), prominent solar elastosis and a well-defined horizontally oriented proliferation of vascular channels on the dermis. Higher magnification revealed that some blood vessels displayed jagged, slit-like spaces and there was focal protrusion of endothelial cells, without cytological atypia, towards the lumen of the vascular space. Histochemistry was positive for CD31 and ERG, and negative for D2-40. The diagnosis of acquired elastotic hemangioma was established and the patient was reassured.

**Conclusion:**

Acquired elastotic hemangioma should be differentiated from other vascular proliferations, including low grade angiosarcoma (which present nuclear atypia, layering of endothelial cells and positivity to D2-40), Kaposi sarcoma and capillary hemangioma.

**Abstract N°: 5618****Mid-dermal elastolysis: a diagnosis to keep in mind**

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**Introduction & Objectives:**

Mid-dermal elastolysis (MDE) is an acquired condition characterized by a focal and selective loss of elastic fibers in the mid-dermis. Misdiagnosis of this entity is frequent. We herein report a new case.

**Materials & Methods:****Results:**

A 68-year-old woman with a medical history of high blood pressure and hepatitis B presented to our dermatology department with an asymptomatic eruption of the trunk evolving for one week. Physical examination revealed multiple annular erythematous patches with fine scaling affecting the trunk and proximal extremities. Face and distal extremities were spared. Diagnosis of pityriasis rosea was suspected. At one month-follow up, we noted a regression of inflammatory aspect leaving multiple well-circumscribed patches with fine wrinkles. The rest of physical examination was unremarkable. A skin biopsy with histopathological examination using hematoxylin-eosin staining revealed changes in elastin fibers with perivascular inflammatory infiltrate made of lymphocytes. Orcein-stain showed focal and selective loss of elastic fibers in the mid-dermis. Diagnosis of MDE was made. Therapeutic abstention was considered.

**Conclusion:**

Recognition of MDE is important to avoid unnecessary investigations. We describe an original observation of type-1 MDE in an older woman with a particular association with hepatitis B.

Since it was first described in 1977, more than 100 cases of MDE have been reported in the literature. Yet, the entity remains underdiagnosed. The condition usually affects middle-aged Caucasian females at odds with our observation in an older woman. Clinically, MDE often exhibits as well-circumscribed and fine wrinkled patches, as it was observed in our patient (Type 1). Perifollicular protrusions (Type 2) and reticulate erythema (Type 3) are considered as less frequent variants of MDE. In all subtypes, the trunk and proximal extremities are the most affected sites. The hallmark of the disease is the histopathological aspect using elastic stains mainly Orcein. Patchy or band-like loss of elastic fibers in the mid-dermis helps in making diagnosis. Similar to our case, evidence of clinical or histopathological inflammation has been reported in 47% of the patients. Furthermore, the association of MDE and several inflammatory, infectious and autoimmune processes has been described. Importantly, cases of granuloma annulare and pityriasis rosea preceding elastolytic destruction need particular attention. Thus, it remains delicate to determine if the initial inflammatory aspect observed in our patient is a true association with pityriasis rosea or not. On the other hand, association of MDE with hepatitis in B seems to be particular in our case as it hasn't been reported in other patients. Physiopathology of MDE is still not well understood. Overexpression of matrix metalloproteinases and decreased activity of their inhibitors are thought to lead to a selective and focal loss of elastic fibers. The course of the condition remains benign. Several therapeutic options such as dapsone, steroids, colchicine and hydroxychloroquine have been proposed. Results are usually deceiving as the regression of already wrinkled lesions is often not achieved. Therapeutic abstention was considered in our patient giving the age and the absence of aesthetic discomfort.

**32<sup>ND</sup> EADV Congress 2023**  
**11 OCTOBER - 14 OCTOBER 2023**  
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**Abstract N°: 5621****A case of late-onset papular elastorrhesis of the neck**

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**Introduction & Objectives:** Papular elastorrhesis (PE) is a rare acquired skin disorder that affects the elastic tissue. Its etiopathogenesis remain unknown. It generally occurs in women in the first or second decade of life. Clinically, PE is characterized by multiple asymptomatic, nonfollicular, hypopigmented papules commonly distributed on the trunk and extremities. It is a benign skin condition with no extracutaneous involvement.

**Materials & Methods:** We herein report a case of late-onset PE of the neck.

**Results:** A 78-year-old female patient with history of glaucoma presented with asymptomatic papules on the neck that appeared two years ago. There was no history of previous inflammation or trauma nor a family history of similar lesions. Dermatological examination showed multiple, non follicular, monomorphous, flesh-colored, firm, 2- to 5- mm papules located on lateral and posterior sides of the neck. Ophthalmologic examination was normal. A skin biopsy was performed and histopathological exam revealed a normal epidermis and thickened collagen fibres. Orcein staining showed an important loss and fragmentation of elastic tissue in the dermis with no calcification. The diagnosis of PE was made. The patient was reassured about the benignity of this skin condition and thus received no treatment.

**Conclusion:** PE is a rare defect of the dermal elastic fibers, first reported in 1987 by Bordas et al. It is characterized by multiple asymptomatic monomorphous, hypopigmented or skin-colored papules affecting commonly the trunk and extremities. Atypical locations such as the face, the neck, the scalp and the retroauricular region have been reported in the literature. To our knowledge, neck involvement was described in three cases, associated with other locations in two out of the three cases. PE is most likely to occur in young people with predilection for the female gender. Late onset of PE is uncommon. The highest reported age of onset is 62 years. The histopathological assessment of the lesions reveals characteristic rarefaction and important fragmentation of the elastic fibers of the reticular dermis. The collagen is thickened or normal. Differential diagnosis of PE includes perifollicular elastolysis, mid-dermal elastolysis, pseudoxanthoma elasticum, pseudoxanthoma elasticum-like papillary dermal elastolysis and papular acne scars. However, PE is a benign condition and treatment includes topical tretinoin, oral antibiotics, oral isotretinoin and intralesional injection of triamcinolone but the response is variable. Considering the uncertain improvement and the benign outcome, therapeutic abstention should be considered. In summary, our case is distinguished by the uncommon late-onset of PE and the atypical location of the lesions.



**Abstract N°: 5654****Uncommon Breast Dermatoses**

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**Introduction & Objectives:**

Some dermatologic conditions are unique to the breast and nipple, whereas others may incidentally involve these structures. Considering the functional, sexual, and aesthetic importance of this area, the estimation of appropriate diagnosis and prompt treatment are of utmost importance.

**Materials & Methods:**

Case 1: A 39-year-old married lady presented with a painful thick indurated hyperpigmented plaque on left breast for 6 months. Histopathology showed features of morphea. The patient was treated with weekly methotrexate 10 mg per week with significant improvement.

Case 2: A 24-year-old unmarried female presented with itchy hypopigmented lesions on both breasts for 6 months. Cutaneous examination revealed multiple hypopigmented and depigmented patches with follicular plugging involving both breasts. Histopathology features of lichen sclerosus. Patient showed good response to a combination of oral isotretinoin and methotrexate.

Case 3: A 56-year-old post-menopausal female presented with erythematous, indurated, tender plaque involving the right areola and nipple with hemorrhagic discharge associated with a single, firm, tender, palpable mass measuring 4cmx6cm with no axillary lymphadenopathy for six months. Histopathology revealed a diagnosis of Paget's disease of the breast with ductal carcinoma in situ. The patient was managed by surgical excision.

Case 4: A 38-year-old lady with seven months of gestation presented with itchy, eczematous, plaque over the left breast areola with nipple inversion and underlying firm, indurated, non-tender palpable mass with no axillary lymphadenopathy since five months. A core needle biopsy revealed a diagnosis of Paget's disease in pregnancy. The patient underwent tumor excision after delivery.

Case 5: A 21-year-old unmarried female presented with an asymptomatic eczematous lesion on her right nipple for 3 years. She has been diagnosed with nipple eczema and was treated with topical steroids without significant improvement. Histopathology showed features of nipple adenoma. Mammography was normal. The patient denied surgical excision of the nipple.

Case 6: A 52-year-old married lady presented with sudden onset galactorrhea, and breast engorgement for 7 days with a delayed menstrual cycle. She was diagnosed to have pemphigus vulgaris, received a rituximab infusion 3 months ago, and was on cyclophosphamide for 3 months. Her MRI brain and prolactin were normal. She was diagnosed with drug-induced galactorrhoea due to cyclophosphamide. Cyclophosphamide was substituted with azathioprine with improvement in symptoms.

Case 7: A 52 years old male presented with asymptomatic nodular lesions on both breast, right arm and abdomen for 2 years with generalized lymphadenopathy. Hematological investigations showed a markedly raised total leukocyte count with 55% atypical lymphocytes in peripheral smear. FNAC from lesions showed features of low-grade primary B cell lymphoma.

Case 8: 74 years old male presented with painless nodules over both the breast for 2 years with axillary lymphadenopathy. Histopathology and immunohistochemistry revealed features of Primary diffuse large B cell lymphoma.

**Results & Conclusion:** Lesions presenting as dermatitis and mass lesions over breasts need early detection by histopathology and appropriate treatment to maintain the structural and functional aspects.

32<sup>ND</sup> EADV Congress 2023  
11 OCTOBER - 14 OCTOBER 2023  
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**Abstract N°: 5708****unilateral purpura associated to deep vein thrombosis**

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**Introduction :**

Purpura is a clinical sign rarely associated with venous thrombosis; it reflects the extravasation of blood outside the vessels. We report a case of a patient who present a purpura associated to vein thrombosis.

**Case report:**

A 57-year-old man; chronic weaned smoker; followed for pulmonary tuberculosis TPM+ on antibacillary treatment (ERIPK4), a pulmonary-renal syndrome; who presented 2 days after his hemodialysis session, a painful swelling of the right lower limb followed by the development of a skin rash. The examination showed a unilateral petechial and ecchymotic purpura of the right lower limb with bullous lesions with clear content; a unilateral swelling of the same limb extended to the root of the right, and a large right bursa. The rest of the examination was unremarkable.

Blood samples revealed normocytic normochromic anemia at 9g/dl, platelets at 168000/ul, a normal white blood cell count, hepatic cytolysis: ASAT at 66 u/l, ALAT at 57 u/l, GGT at 108, urea at 1.73g/l, creatinine at 85.6 mg/l, CRP at 177 mg/l and procalcitonin at 2.65. Doppler ultrasound of the right lower limb revealed a right deep venous thrombosis (DVT) of the right lower limb involving the external iliac vein, the superficial and deep common femoral vein and the popliteal vein.

The decision was to continue the antibacillary treatment under strict hepatic surveillance, to put the patient on antibiotics based on ceftriaxone 2 grams per day and anticoagulant (bemiparin 5000 ui per day), thereafter the purpura began to regress; and on the dermatological level to carry out a thrombophilia and paraneoplastic assessment and a skin biopsy; the patient refused the treatment and left against medical advice.

**Discussion:**

Purpura has two mechanisms of onset. The first is hematological: coagulation problem allows blood to leave the blood capillaries, as in thrombocytopenia and other thrombopathies. The second mechanism is vascular: the integrity of the blood vessel wall or capillaries is altered, which allows blood to escape into the dermis, in these cases the purpura is bilateral. Other rare causes of asymmetric purpura have been attributed to unilateral Schamberg disease or lichen purpuricus and Covid 19.

While DVT is associated with an acute increase in venous pressure, for unclear reasons it rarely causes purpura, having been described twice previously in the literature.

In our case, we have the stasis factor. We therefore suspected that venous stasis was an etiological factor.

**Conclusion:**

Purpura is a rare sign of deep vein thrombosis. Our observation is original as it is the third case described.



**Abstract N°: 6017****Medallion-like dermal dendrocyte hamartoma - case report of a rare neoplasm**

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**Introduction & Objectives:**

Medallion-like dermal dendrocyte hamartoma (MLDDH), also called “plaque-like CD34-positive dermal fibroma”, was first described in 2004 by Rodriguez-Jurado et al as solitary, slightly atrophic, and asymptomatic lesions with a distinct medallion shape. Typically, MLDDH is noticed during infancy or at birth on the neck or upper trunk and the lesions are characterized by their pliable nature or slightly indurated plaque appearance. We report a case of this rare neoplasm.

**Materials & Methods:**

Retrospective review of medical records from a patient diagnosed with MLDDH.

**Results:**

We describe the case of a 10-year-old girl referred to our dermatology department due to a congenital, asymptomatic foot lesion, proportionally increasing in size with her growth. Upon physical examination, a slightly infiltrated erythematous-violaceous plaque was evident on the dorsum of the right foot, measuring approximately 6x5 cm. The plaque was oval-shaped with a smooth surface, irregular borders, and well-defined limits.

A skin biopsy of the lesion was performed, revealing a moderately cellular, spindle cell mesenchymal proliferation. The cells were monomorphic and organized in a parallel orientation to the epidermal surface, sparing the cutaneous appendages and occupying the entire thickness of the reticular dermis and superficial hypodermis. No cytological atypia or mitotic figures were found. The stroma showed a mix of collagen with occasional scattered mast cells. The immunohistochemical profile showed diffuse positivity for CD34 and negativity for SMA, SOX10, and HMB45. These findings were suggestive MLDDH, although they could not exclude the diagnosis of dermatofibrosarcoma protuberans (DFSP). Therefore, further investigation was conducted using fluorescence in situ hybridization (FISH) to search for rearrangements in the *COL1A1* gene, which were not found in our case. Surgical treatment was offered in order to obtain a complete removal of the lesion, but the parents declined. As such, we maintain a regular clinical follow-up of the patient.

**Conclusion:**

Despite being described almost twenty years ago, and with less than 30 cases reported in the literature, there are still many aspects of this condition that remain unclear. Typically, the majority of affected individuals are females, and it is generally believed to be a congenital or early-life occurrence. Given its clinical presentation, differential diagnoses to consider include aplasia cutis, atrophoderma, anetoderma, and congenital (DFSP). Therefore, performing a skin biopsy is of utmost importance for establishing an accurate diagnosis. In light of the dermal proliferation of spindle cells, it is also important to consider histologic differentials such as dermatofibroma, neurofibroma or fibroblastic connective tissue nevus. As described in our case, molecular studies may be necessary to exclude the diagnosis of DFSP, which displays the gene translocation at t(17;22)(q22;q13), expressed in the fusion gene *COL1A1-PDGFB*. While some authors recommend excision of the lesions due to the possibility

of malignancy, MLDDH is benign, and no cases of malignant transformation have been reported thus far.

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