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Case Report

Plexiform Fibrohistiocytic Tumor with Pulmonary Metastases: A Case Report

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Abstract

Plexiform fibrohistiocytic tumor is a rare mesenchymal neoplasm classified as an intermediate-grade malignancy because of its locally aggressive behavior. We report the case of a 35-year-old chronic smoker who presented with a recurrent inguinal cutaneous tumor. Initial histopathological examination suggested a fibrohistiocytic tumor. Dermatological examination revealed a pigmented, retractile verrucous inguinal mass associated with multiple subcutaneous nodules distributed throughout the body, occurring in a context of general deterioration. Staging investigations, including lymph node ultrasonography and thoraco-abdomino-pelvic computed tomography, demonstrated multiple pulmonary nodules suggestive of metastatic disease.

Histopathological and immunohistochemical findings demonstrated a plexiform spindle-cell proliferation with histiocytic differentiation, showing CD68/CD163 positivity and negative CD34 and HMB45 staining, consistent with the rare monophasic spindle-cell variant of plexiform fibrohistiocytic tumor with pulmonary metastases. Following a multidisciplinary discussion, systemic chemotherapy was initiated because surgical management was not feasible owing to disseminated pulmonary involvement. Follow-up was incomplete, as the patient was lost to follow-up after two cycles of chemotherapy.

This case highlights an exceptionally rare presentation of plexiform fibrohistiocytic tumor with pulmonary metastases and underscores the diagnostic challenges posed by this entity, which may clinically and histologically mimic more aggressive soft tissue sarcomas.

Introduction

Plexiform fibrohistiocytic tumor is a rare mesenchymal neoplasm. First described by Enzinger and Zhang in 1988, it remains an uncommon and often underrecognized entity [1]. It predominantly affects children and young adults [2] and is characterized by a distinctive histopathological architecture. Owing to its rarity and the absence of specific clinical features, its diagnosis may represent a significant diagnostic challenge. We report a case of plexiform fibrohistiocytic tumor with pulmonary metastases.

Case Report

A 35-year-old man with chronic tobacco use and a history of schizophrenia treated with antipsychotic medication presented with a recurrent cutaneous tumor evolving over 8 years. Three previous biopsies, including immunohistochemical studies, had been interpreted as dermatofibroma.

Dermatological examination revealed an 8-cm painful pigmented verrucous retractile tumor with an infiltrated base located in the left iliac region, surrounded by brownish hyperpigmentation (Figure 1), along with a pigmented linear inguinal scar. Dermoscopy revealed a brownish-yellow hyperkeratotic lesion with a papillomatous architecture, fissures, multiple milia-like cysts, and comedo-like openings , creating a cerebriform appearance (Figure 1A).

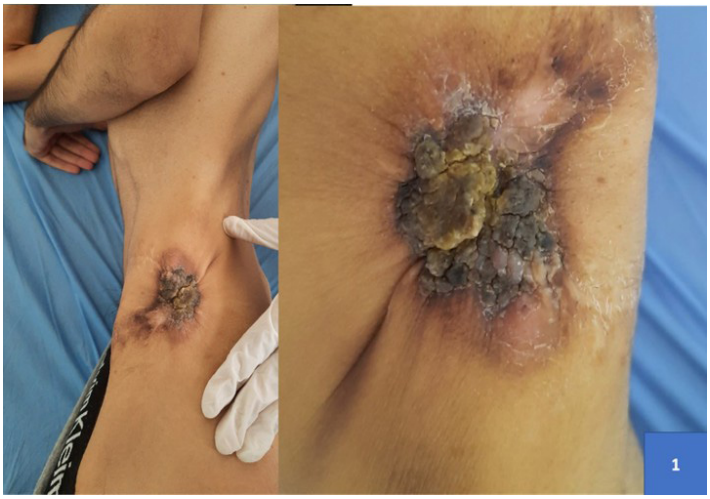

Figure 1: *An approximately 8-cm, poorly demarcated, oval, retractile tumor with irregular borders, a verrucous surface, and an infiltrated base, located on the left iliac region and surrounded by brownish pigmentation.*

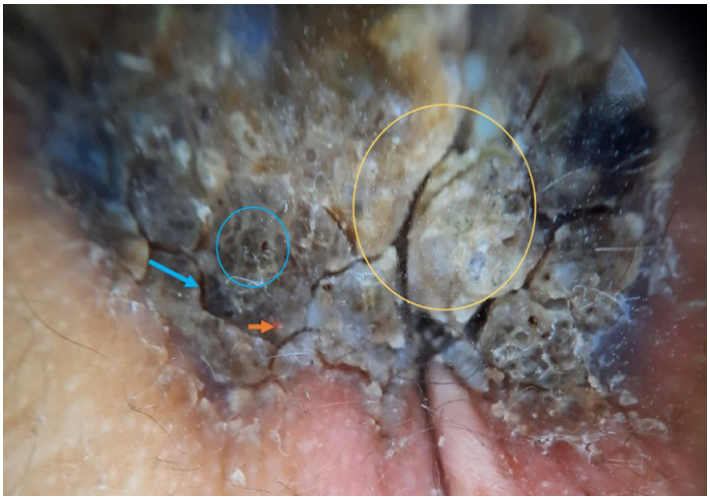

Figure 1A: *Dermoscopy revealed a brownish-yellow hyperkeratotic (yellow circle) lesion with a papillomatous architecture, fissures (blue arrow), multiple milia-like cysts (orange arrow), and comedo-like openings (blue circle), creating a cerebriform appearance.*

A similar 1.5-cm lesion was observed in the right inguinal region (Figure 2). Multiple subcutaneous nodules of variable size were also noted in the paraumbilical, axillary, and cervical regions (Figure 3). These nodules were attached to the deep plane but remained mobile relative to the skin. Bilateral inguinal lymphadenopathy was present. The lesions had developed in a context of anorexia and significant weight loss.

The main differential diagnosis is recurrent dermatofibroma and dermatofibrosarcoma protuberans (DFSP), based on the patient's age, the infiltrative growth pattern, and the history of repeated local recurrence following previous surgical excisions. However, the multifocal distribution of the lesions, associated constitutional symptoms, and the subsequent histopathological and immunohistochemical findings ultimately ruled out this diagnosis.

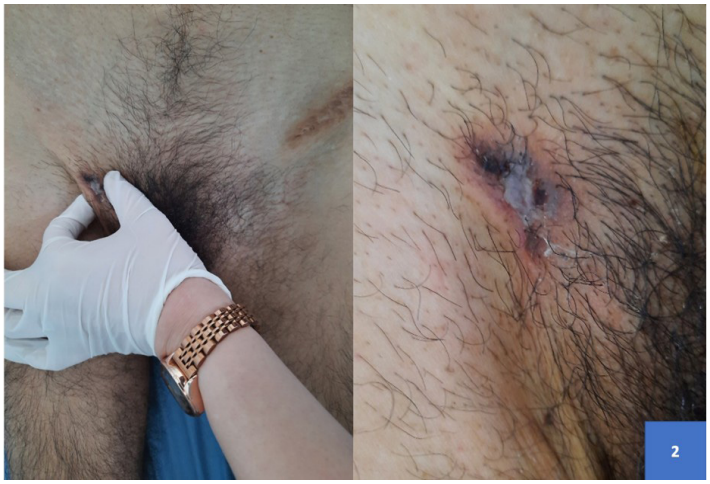

Figure 2: *A pigmented plaque measuring approximately 1.5 cm, with a slightly hyperkeratotic surface and a positive dimple sign, located in the right inguinal region.*

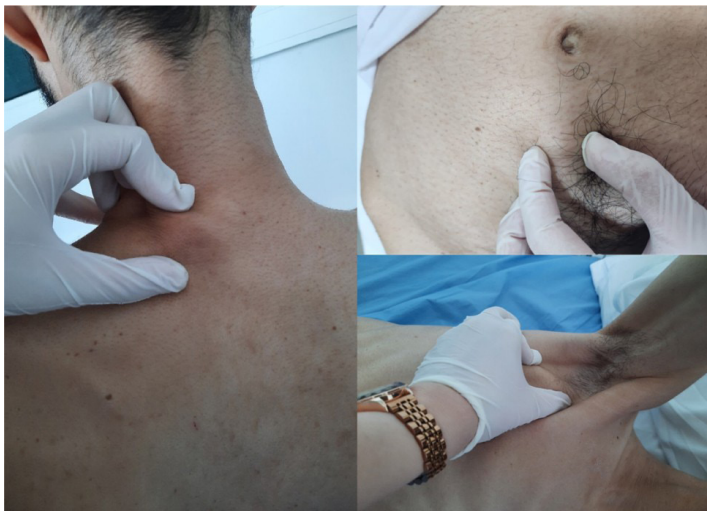

Figure 3: *Multiple subcutaneous nodules of variable size, the largest measuring 4 cm, located in the paraumbilical region, axilla, and nape of the neck. The nodules were adherent to the underlying structures while remaining mobile relative to the overlying skin.*

Pecoma was also considered because of the presence of pigmented nodules occurring in an adult patient, although the cutaneous location of this tumor is exceptionally rare.

Verrucous squamous cell carcinoma was discussed in view of the verrucous appearance of the lesion; however, this diagnosis was considered unlikely owing to the absence of ulceration, bleeding, or foul-smelling discharge.

Given the patient's constitutional symptoms, including weight loss, cachexia, chronic tobacco use, and persistent cough, cutaneous metastases from an underlying solid malignancy, particularly of pulmonary or renal origin, were also suspected.

Finally, the infiltrative pattern, local recurrence, and multifocal distribution of the lesions raised the possibility of an aggressive soft tissue sarcoma.

Histopathological examination showed a dermal and

deep-seated spindle-cell proliferation arranged in short interlacing fascicles with a storiform pattern (Figure 4). Cellularity was high, with occasional non-atypical mitotic figures. Xanthomatous histiocytes and hemosiderin-laden cells were present, with positive Perls staining and negative Fontana-Masson staining. Dense collagen bundles and vascular clefts were also identified (Figure 5).

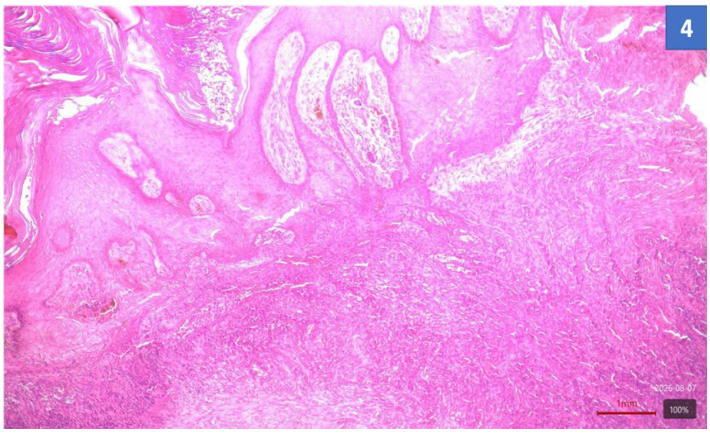

Figure 4: *Histopathological examination (H&E, ×50) showing a hyperacanthotic verrucous epidermis overlying a dermal spindle-cell proliferation composed of fibroblast-like cells arranged in fascicles and focally in a storiform pattern.*

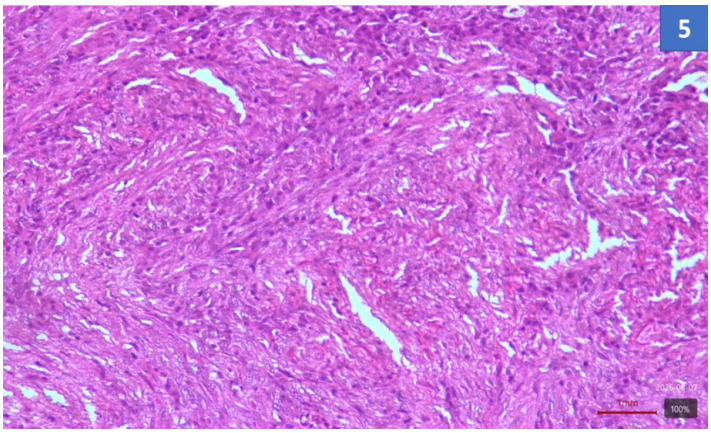

Figure 5: *Histopathological examination (H&E, ×200) showing a highly cellular spindle-cell proliferation arranged in short interlacing fascicles, associated with slit-like vascular spaces.*

Immunohistochemical analysis showed expression of CD68 and CD163 in histiocytic cells (Figure 6A), with CD163 also positive in spindle cells. CD10 was diffusely expressed, while smooth muscle actin showed focal positivity. Focal S100 expression was observed in histiocytoid cells. CD34 and HMB45 were negative. Nuclear p53 staining was weak and heterogeneous, and the Ki-67 proliferation index was low (approximately 3%) (Figure 6B).

Lymph node ultrasonography demonstrated axillary, inguinal, and cervical lymphadenopathy. Thoraco-abdomino-pelvic computed tomography revealed multiple pulmonary nodules consistent with metastatic disease, left external iliac lymphadenopathy with central necrosis, and a small left pleural effusion. Serum LDH, β-hCG, CA19-9, and

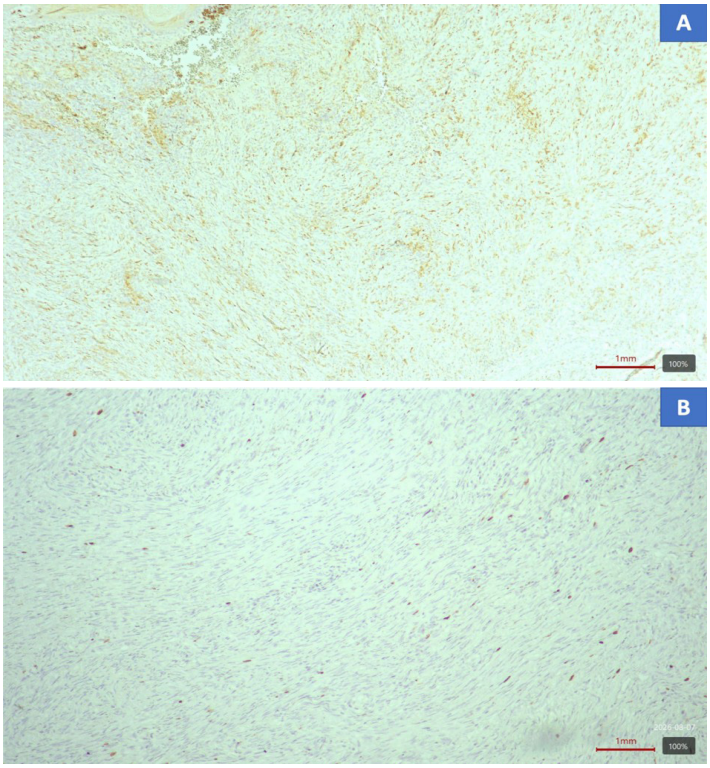

Figure 6: *Immunohistochemical findings*
(A) diffuse CD163 positivity in the tumor cells (×50)
(B) low Ki-67 proliferative index, estimated at 3% (×100).

alpha-fetoprotein levels were within normal limits.

Based on the clinical, radiological, histopathological, and immunohistochemical findings, a diagnosis of plexiform fibrohistiocytic tumor, specifically the rare monophasic myofibroblastic spindle-cell variant, was favored. The pulmonary lesions were considered highly suggestive of metastatic spread.

Following multidisciplinary discussion and pathological review, the differential diagnosis with monophasic synovial sarcoma remained a concern because of the unusually aggressive clinical course. As curative surgery was not feasible due to disseminated pulmonary involvement, the patient was treated according to a sarcoma protocol with doxorubicin monotherapy. After two cycles of chemotherapy, he was lost to follow-up.

Discussion

Plexiform fibrohistiocytic tumor is a rare mesenchymal neoplasm of intermediate malignancy, characterized by local aggressiveness, a tendency for recurrence, and a low metastatic potential [1]. Its diagnosis remains challenging because of the lack of specific clinical features and its rarity.

Clinically, it usually presents as a solitary, firm nodule with poorly defined margins. Its size is generally less than 3 cm, although it may occasionally manifest as a slow-growing, painless subcutaneous mass. The upper extremities, particularly the hand and wrist, are the most common

sites, accounting for approximately 50% of cases. Other reported locations include the lower extremities (20%), as in our patient, the trunk (15%), and the head and neck region (10%) [3]. To date, no specific dermoscopic features have been described in the literature. Our case is unusual because of its inguinal location, which is only exceptionally reported in the literature.

Definitive diagnosis relies on histopathological examination [3]. Plexiform fibrohistiocytic tumor is a multinodular mesenchymal proliferation with an infiltrative plexiform architecture, preferentially involving the deep dermis and the dermo-subcutaneous junction, although purely dermal or subcutaneous forms, sometimes extending into the underlying muscle, have also been reported. The tumor is composed, in varying proportions, of three cellular populations: spindle-shaped fibroblastic cells, mononuclear histiocytoid cells, and osteoclast-like multinucleated giant cells, which may occur in mixed patterns. The plexiform architecture consists of small to medium-sized nodules. Hemorrhagic foci associated with hemosiderin deposition are frequently observed, particularly in fibroblastic-predominant lesions, whereas foci of osseous metaplasia have occasionally been reported. Inflammation is usually minimal or absent. Vascular invasion, as well as perineural, peri Pacinian corpuscle, or adnexal entrapment, may be present. In fibroblastic variants, extension into the subcutaneous adipose tissue may produce a characteristic “micro-adipocyte” appearance. Cytologically, the tumor typically lacks significant pleomorphism, marked mitotic activity, or hyalinization; however, rare cases with cellular atypia or occasional atypical mitotic figures have been reported. Immunohistochemically, spindle cells express smooth muscle actin (SMA), whereas histiocytoid and giant cells are positive for the histiocytic markers CD68, CD163, and NK1/C3. Cytokeratins, desmin, HMB-45, S100 protein, and CD34 are usually negative [4]. In our patient, the histological pattern corresponded to the fibroblastic spindle-cell variant with a plexiform architecture.

Plexiform fibrohistiocytic tumor is an extremely rare neoplasm, with fewer than one hundred cases reported in the literature. The largest published series included 66 and 22 cases, respectively [1,5].

The main differential diagnosis is cellular neurothekeoma, which shares a superficial location and multinodular architecture with plexiform fibrohistiocytic tumor; however, it differs by the absence of osteoclast-like giant cells and the lack of expression of histiocytic markers such as CD68 and CD163 [6,7]. Fibromatosis and lipofibromatosis may exhibit a similar infiltrative growth pattern but lack both the histiocytic component and the characteristic plexiform organization [8]. Malignant fibrous histiocytoma may also display a fibrohistiocytic proliferation, but it is typically associated with marked pleomorphism and high mitotic activity [9]. Finally, giant cell tumor of soft tissue likewise contains numerous osteoclast-like giant cells [10], but generally lacks the characteristic multinodular plexiform architecture of plexiform fibrohistiocytic tumor.

Plexiform fibrohistiocytic tumor carries a significant risk of local recurrence, whereas lymph node and pulmonary metastases remain exceptional [6]. No clinicopathological features have been clearly associated with prognosis. Our patient developed multiple pulmonary nodules highly suggestive of metastatic disease, representing an exceptionally uncommon evolution of PFHT.

Treatment of localized disease relies on wide surgical excision with tumor-free margins, which usually provides durable disease control [6]. Despite its generally favorable outcome, local recurrences may occur, warranting long-term follow-up. Metastatic forms, which are exceptionally reported, mainly involve lymph nodes and lungs [1]. In such situations, management requires a multidisciplinary approach combining, whenever feasible, surgical resection of metastatic lesions and systemic therapies tailored to the clinical context [4]. Management was further complicated by the absence of standardized treatment protocols for metastatic PFHT, the impossibility of curative surgery due to disseminated pulmonary involvement, and the patient's subsequent loss to follow-up after two cycles of chemotherapy.

Conclusion

Plexiform fibrohistiocytic tumor is a rare intermediate-grade neoplasm whose diagnosis may be challenging, particularly in uncommon histological variants. Although its clinical course is usually favorable following complete surgical excision, rare cases of pulmonary metastases have been reported and represent a significant diagnostic and therapeutic challenge. Our case highlights the importance of early recognition, complete surgical resection whenever feasible, and long-term follow-up to enable the timely detection of recurrences or metastatic disease.

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