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Dermal Pleomorphic Sarcoma of the Scalp: A Case Report

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Abstract

Pleomorphic dermal sarcoma (PDS) is a rare cutaneous malignancy that predominantly affects elderly individuals and typically arises in chronically sun-exposed areas, particularly the scalp and face. It is distinguished from atypical fibroxanthoma by the presence of high-risk histopathological features, including subcutaneous invasion, tumor necrosis, and perineural or lymphovascular invasion.

We report the case of a 90-year-old man admitted to Mohammed VI University Hospital in Oujda, Morocco, in November 2025, for a progressively enlarging occipital scalp mass that had become painful and ulcerated. Clinical examination revealed a large, exophytic, ulcerated tumor measuring approximately 20 cm. Histopathological examination of the excision specimen demonstrated a markedly pleomorphic spindle and epithelioid cell proliferation with atypical mitotic figures, tumor necrosis, and perineural invasion. Immunohistochemistry revealed diffuse, strong CD10 expression and heterogeneous CD68 positivity, while epithelial, melanocytic, and endothelial markers were negative, supporting the diagnosis of PDS after exclusion of relevant differential diagnoses.

This case illustrates the importance of recognizing the characteristic histopathological features of PDS and integrating immunohistochemistry into the diagnostic workup. It also highlights the diagnostic challenges posed by a giant scalp tumor in an elderly patient and the need for appropriate management and clinical follow-up, given the potential for local recurrence and metastasis.

Categories: Pathology, Anatomy, Dermatology
Keywords: atypical fibroxanthoma, cd10, dermal pleomorphic sarcoma, differential diagnosis, scalp

Introduction

Pleomorphic dermal sarcoma (PDS) is a rare, poorly differentiated cutaneous mesenchymal malignancy that predominantly affects elderly individuals, particularly men, and most frequently arises in chronically sun-exposed areas, especially the scalp, face, and neck [1,2]. Chronic ultraviolet (UV) radiation exposure and advanced age are considered important risk factors. Clinically, PDS typically presents as a rapidly growing dermal or subcutaneous nodule that may become ulcerated, hemorrhagic, or painful, although its clinical appearance is nonspecific and may overlap with that of other cutaneous malignancies.

Histologically, PDS is characterized by a highly pleomorphic proliferation of spindle-shaped and epithelioid cells, often accompanied by marked nuclear atypia, atypical mitotic figures, and variable tumor necrosis. A defining feature of PDS is the presence of one or more adverse histopathological findings, including invasion of the subcutaneous tissue, tumor necrosis, lymphovascular invasion, or perineural invasion. These features distinguish PDS from atypical fibroxanthoma (AFX), a closely related cutaneous neoplasm that generally remains confined to the dermis and lacks these high-risk characteristics [2,3].

The diagnosis of PDS is based on the correlation of clinical findings, histopathological examination, and immunohistochemistry. Its immunophenotype is nonspecific, and immunohistochemical studies are primarily used to exclude other pleomorphic cutaneous malignancies, including poorly differentiated carcinomas, melanomas, and vascular or other mesenchymal tumors. PDS generally expresses CD10 and may show variable CD68 expression, whereas epithelial, melanocytic, and endothelial markers are typically negative. However, these findings must be interpreted in conjunction with the morphological features and cannot independently establish the diagnosis.

Compared with AFX, PDS carries a greater risk of local recurrence and distant metastasis, making complete surgical excision and appropriate clinical follow-up essential. Its management may be challenging because of its locally aggressive behavior, particularly when it involves anatomically complex sites such as the scalp.

We report a case of a giant PDS of the scalp in a 90-year-old man, presenting as a rapidly enlarging, ulcerated mass. This case highlights the diagnostic value of identifying high-risk histopathological features, the role of immunohistochemistry in excluding morphological mimics, and the clinical challenges associated

with an exceptionally large tumor arising in an elderly patient.

Case Presentation

A 90-year-old man with no significant medical history presented with an eight-month history of an occipital scalp nodule. The lesion was initially asymptomatic but progressively enlarged, followed by a period of rapid growth associated with pain and ulceration. An initial biopsy suggested PDS.

On physical examination, a large left occipital scalp mass measuring approximately 20 cm in greatest dimension was observed. The tumor was exophytic, erythematous with violaceous areas, and had an ulcerated, lobulated surface. It was firm, non-tender, and fixed to the underlying tissues. Whole-body computed tomography (CT) revealed a large soft-tissue mass involving the left occipital scalp, without intracranial extension or evidence of distant metastasis.

The surgical specimen measured 14 × 13 × 9.5 cm, and the tumor measured 11.2 × 7.2 × 6 cm on gross examination. These measurements differed from the approximately 20 cm clinical estimate obtained on physical examination, as they were obtained from the resected specimen during gross pathological assessment.

Given its large size, rapid growth, ulceration, and clinical appearance, the lesion raised suspicion for a malignant cutaneous tumor. The clinical differential diagnosis included cutaneous squamous cell carcinoma, basal cell carcinoma with an aggressive growth pattern, amelanotic melanoma, and other pleomorphic dermal neoplasms, particularly AFX and PDS. Histopathological examination and immunohistochemistry were essential to establish the final diagnosis and exclude these mimicking entities.

Pathological findings

Gross Examination

The surgical specimen consisted of a scalp excision measuring 14 × 13 × 9.5 cm, with an exophytic tumor measuring 11.2 × 7.2 × 6 cm. On sectioning, the tumor showed cystic changes and hemorrhagic areas (Figure *1*).

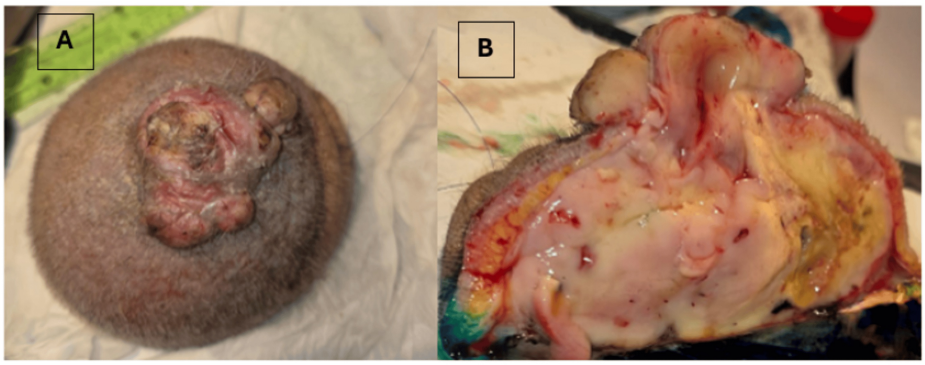


FIGURE 1: Gross photograph of the surgical resection specimen.

A: Resected tumor specimen. B: Internal appearance of the tumor on the bisected specimen.

Histological Examination

Microscopic examination revealed skin tissue covered by a regular epidermis with overlying orthokeratotic hyperkeratosis. The underlying dermis contained an encapsulated, nodular, and diffusely arranged tumor proliferation (Figure *2*). The tumor was composed predominantly of spindle-shaped cells, with occasional epithelioid cells showing marked cytologic atypia and pleomorphism (Figure *3A*). The tumor cells exhibited enlarged nuclei with irregular nuclear contours, and the chromatin ranged from dense to vesicular, with prominent nucleoli (Figure *3B*). Numerous mitotic figures, including atypical mitoses, were identified. The tumor stroma was fibrous and contained congested blood vessels. Multifocal tumor necrosis was present (Figure *4A*), along with perineural invasion (Figure *4B*).

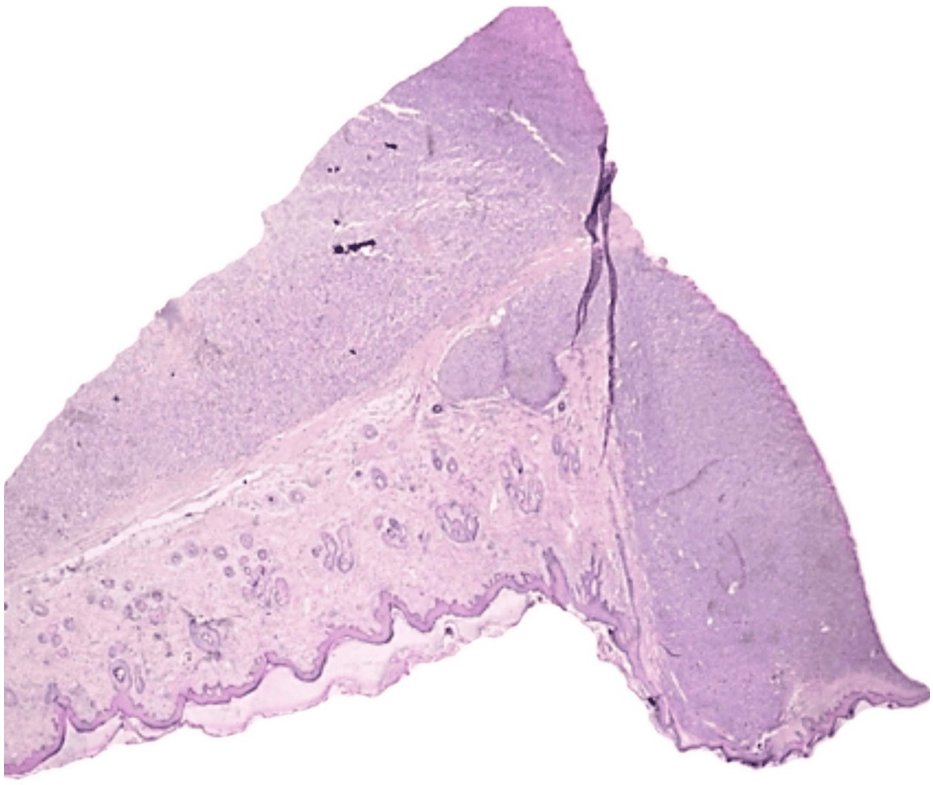


FIGURE 2: Photomicrograph showing a dermal tumor proliferation with preservation of the overlying epidermis.

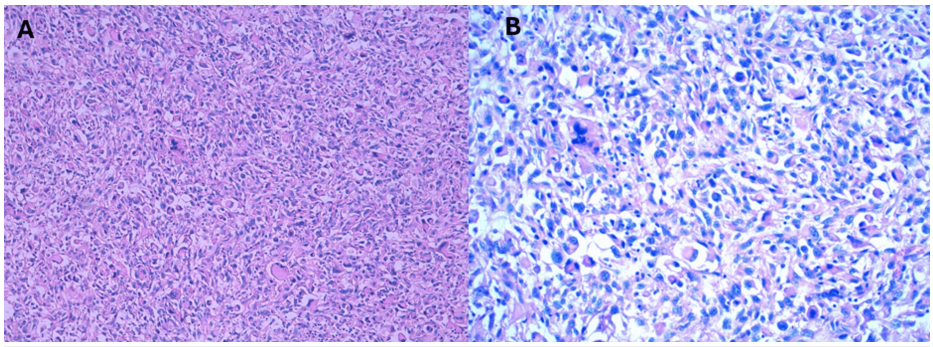


FIGURE 3: H&E-stained photomicrographs illustrating the histological features of pleomorphic dermal sarcoma.

A: Low-power photomicrograph (×10) showing the spindle-cell morphology and marked cellular pleomorphism. B: Higher-power photomicrograph (×20) highlighting the cytological atypia.

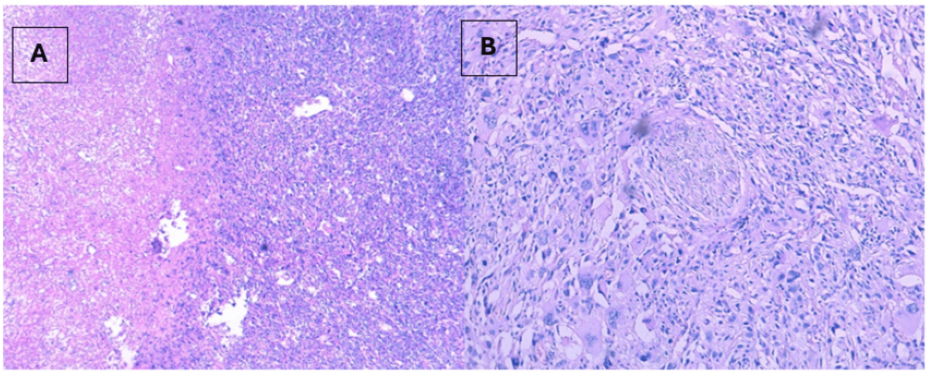


FIGURE 4: H&E-stained photomicrographs of pleomorphic dermal sarcoma.

A: Photomicrograph showing tumor necrosis. B: Photomicrograph showing perineural invasion.

Immunohistochemistry

Immunohistochemical analysis demonstrated strong and diffuse CD10 positivity (Figure *5*) and heterogeneous CD68 positivity (Figure *6*). The tumor cells showed negative staining for epithelial markers, including pancytokeratin and p40 (Figure *7*), as well as negative staining for melanocytic markers, including S100 and Melan-A (Figure *8*). Vascular markers, including CD34 and CD31, were also negative (Figure *9*). Taken together with the morphological features and the presence of aggressive histological criteria, these immunohistochemical findings supported the diagnosis of PDS.

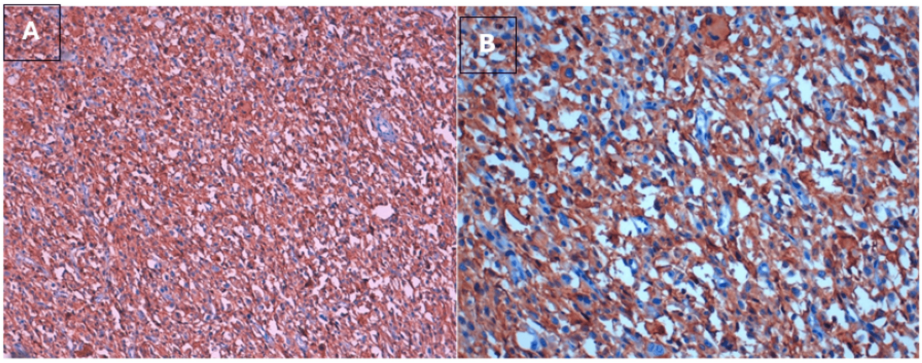


FIGURE 5: Immunohistochemical staining showing strong and diffuse CD10 expression.

A: Low-power view (×10). B: Higher-power view (×20).

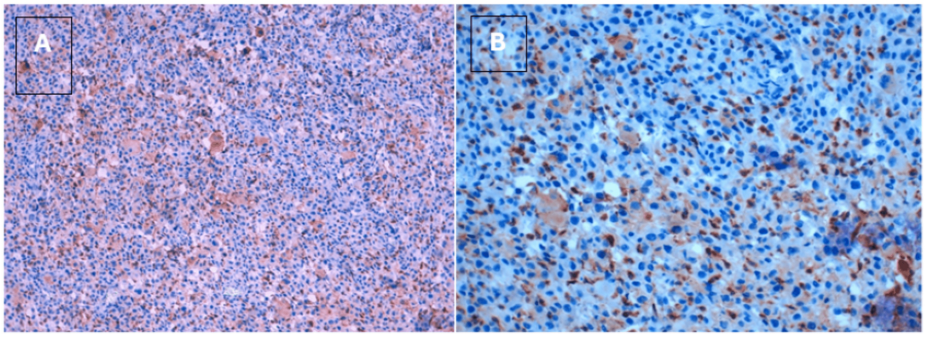


FIGURE 6: Immunohistochemical staining showing heterogeneous CD68 expression in pleomorphic dermal sarcoma.

A: Low-power view illustrating the heterogeneous distribution of CD68-positive cells. B: Higher-power view highlighting the heterogeneous cytoplasmic CD68 positivity.

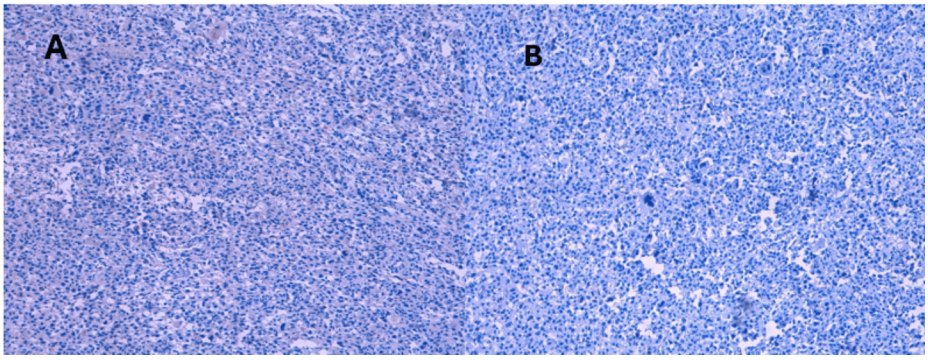


FIGURE 7: Immunohistochemical staining showing negative expression for p40 (A) and pancytokeratin (B).

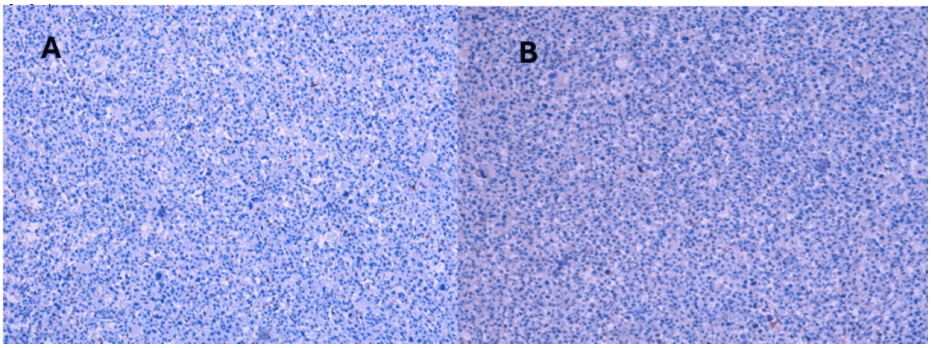


FIGURE 8: Photomicrographs showing negative immunostaining for S100 (A) and Melan-A (B).

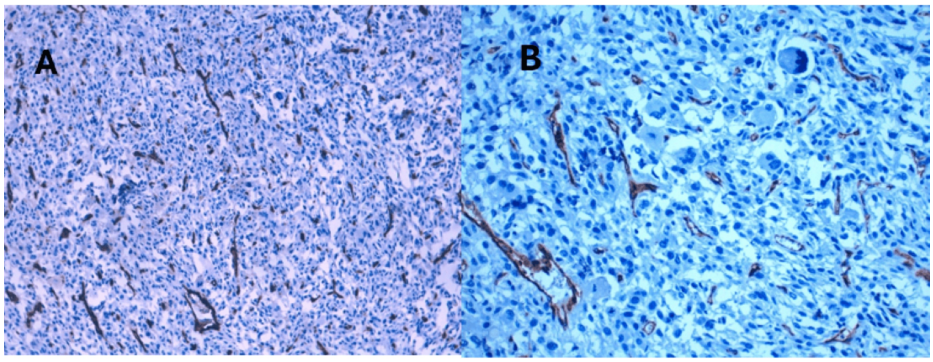


FIGURE 9: Immunohistochemical staining showing negative expression for CD34 (A) and CD31 (B).

Histological examination demonstrated a pleomorphic spindle-cell tumor extending from the dermis into the subcutaneous adipose tissue, with marked cytological atypia, atypical mitotic figures, multifocal tumor necrosis, and perineural invasion. Immunohistochemical analysis supported the diagnosis of PDS after exclusion of relevant differential diagnoses. The surgical margins were negative, with a minimum clearance of more than 5 mm.

The patient underwent surgical excision followed by adjuvant radiotherapy. At one year of follow-up, no local recurrence or metastatic disease had been documented.

Discussion

The morphological description of malignant pleomorphic sarcoma was first reported in 1963 as a predominantly dermal spindle cell neoplasm [4]. However, the term pleomorphic dermal sarcoma was first introduced into the literature in 2011 in an editorial by Dr. McCalmont devoted to AFX. In this editorial, he stated that a tumor confined to the dermis or associated with only minimal subcutaneous involvement has a low malignant potential and may therefore be designated as either AFX or PDS [5,6].

In 2012, Dr. McCalmont [7], echoing the views of Dr. Fletcher, further clarified that AFX and PDS are not synonymous entities. AFX refers to tumors strictly confined to the dermis and lacking adverse histopathological features, whereas PDS describes morphologically similar lesions that exhibit invasive characteristics and an uncertain biological behavior. This distinction, based on well-defined histopathological criteria, was subsequently adopted by Miller et al. [8] and has since been incorporated into the current World Health Organization (WHO) classification.

The morphological distinction between PDS and AFX is extremely challenging, as both lesions share highly similar histopathological features. Nevertheless, the presence of specific adverse pathological findings strongly supports the diagnosis of PDS. These include deep invasion beyond the dermis, lymphovascular invasion, perineural invasion, tumor necrosis, and metastatic spread.

Morphologically, PDS is characterized by a proliferation of markedly pleomorphic spindle to epithelioid cells with abundant eosinophilic cytoplasm and hyperchromatic or vesicular nuclei, accompanied by a high mitotic rate. The tumor may also exhibit a myxoid or desmoplastic stroma, pseudoangiomatous changes, or a storiform growth pattern. These histological features may overlap with those of other neoplasms, particularly myxofibrosarcoma, desmoplastic melanoma, and cutaneous squamous cell carcinoma, making careful exclusion of these differential diagnoses essential, as PDS remains fundamentally a diagnosis of exclusion [9].

The differential diagnosis of PDS includes several cutaneous malignancies with overlapping morphological features, particularly AFX, spindle cell squamous cell carcinoma, desmoplastic melanoma, angiosarcoma, and leiomyosarcoma [1,10]. AFX represents the closest diagnostic counterpart to PDS, as both tumors share similar cytological features and immunohistochemical profiles. Their distinction relies primarily on histopathological criteria, with invasion of the subcutaneous tissue, tumor necrosis, and perineural or lymphovascular invasion favoring PDS over AFX [1].

Spindle cell squamous cell carcinoma may exhibit marked cellular pleomorphism and a predominantly spindle cell morphology. Epithelial differentiation is supported by immunohistochemical expression of pancytokeratin and p40; therefore, the negativity of both markers in the present case argued against this diagnosis [1,11]. Desmoplastic melanoma may also mimic PDS, particularly when composed of pleomorphic spindle cells infiltrating the dermis. The absence of S100 and Melan-A expression, together with the

morphological findings, did not support this diagnosis [1,10]. Angiosarcoma was considered less likely, given the absence of endothelial marker expression, including CD31 and CD34 [1,10].

Other potential mimics include pleomorphic leiomyosarcoma and other undifferentiated pleomorphic spindle cell neoplasms. However, smooth muscle differentiation could not be formally assessed in the present case because smooth muscle markers were not performed. Therefore, the final diagnosis relied on the integration of the histomorphological findings, the available immunohistochemical profile, and the presence of adverse histopathological features characteristic of PDS [1,10].

Strong, diffuse CD10 positivity, together with heterogeneous CD68 expression, provided supportive but nonspecific evidence for the diagnosis of PDS. Negative staining for epithelial markers, particularly pancytokeratin and p40, vascular markers (CD34 and CD31), and melanocytic markers (S100 and Melan-A) helped exclude several important diagnostic mimics. Although several studies have investigated immunohistochemical markers capable of distinguishing AFX from PDS, none has demonstrated sufficient diagnostic accuracy for routine clinical practice [12]. Furthermore, smooth muscle markers were not performed in the present case; therefore, smooth muscle differentiation could not be formally excluded immunohistochemically. The diagnosis was consequently based on the correlation between the morphological findings and the available immunohistochemical results.

The diagnosis of PDS should generally not be established based on a superficial biopsy alone, as such specimens may fail to adequately assess the depth of invasion, thereby preventing reliable differentiation between AFX and PDS [12].

Pleomorphic dermal sarcoma is characterized by a molecular signature of UV radiation-induced genetic mutations associated with an exceptionally high tumor mutational burden, exceeding that observed in other UV-related cutaneous malignancies, including cutaneous squamous cell carcinoma and malignant melanoma. The most frequently reported genetic alterations are inactivating mutations of TP53, followed by alterations involving the CDKN2A/B locus [1].

This high tumor mutational burden provides a rationale for investigating immunotherapeutic strategies in advanced PDS. Immune checkpoint inhibitors, particularly anti-PD-1 agents such as pembrolizumab, have shown promising results in isolated case reports; however, the available evidence remains limited, and their role in the management of PDS has not yet been established [13]. In the present case, treatment consisted of surgical excision followed by adjuvant radiotherapy, and no immunotherapy was administered.

Although most cases of PDS can be successfully treated by complete surgical excision, approximately one-third of patients develop local recurrence or distant metastases. To date, no standard treatment has been established for advanced disease [14].

In the present case, the unusually large tumor size, together with extension into the subcutaneous adipose tissue, tumor necrosis, and perineural invasion, highlights the aggressive histopathological features of this neoplasm. Despite complete surgical excision with negative margins followed by adjuvant radiotherapy, continued follow-up remains important, given the potential risk of recurrence and metastasis. At one year of follow-up, no local recurrence or metastatic disease had been documented.

Conclusions

Pleomorphic dermal sarcoma is a rare but aggressive cutaneous malignancy. Its diagnosis relies on the correlation of morphological findings with immunohistochemistry to exclude other spindle cell neoplasms. This case highlights the importance of recognizing adverse histopathological features, particularly the large tumor size, tumor necrosis, perineural invasion, and extension into the subcutaneous adipose tissue, in distinguishing PDS from AFX. Although no local recurrence or metastatic disease was documented at one year of follow-up following surgical excision and adjuvant radiotherapy, the limited follow-up period precludes definitive conclusions regarding the long-term outcome.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Concept and design: Manal Kharkhach, Anass Haloui, Asmae Aissaoui, Nassira Karich, Amal Bennani

Acquisition, analysis, or interpretation of data: Manal Kharkhach, Anass Haloui, Asmae Aissaoui, Nassira Karich, Amal Bennani

Drafting of the manuscript: Manal Kharkhach, Anass Haloui, Asmae Aissaoui, Nassira Karich, Amal

Bennani

Critical review of the manuscript for important intellectual content: Manal Kharkhach, Anass Haloui, Asmae Aissaoui, Nassira Karich, Amal Bennani

Supervision: Anass Haloui, Asmae Aissaoui, Nassira Karich, Amal Bennani

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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