

Pleomorphic dermal sarcoma of the scalp: Review of management and distinguishing features from atypical fibroxanthoma



Alexa J. Cohen, BS, Sahithi Talasila, BS, Bojan Lazarevic, BA, Laura Gleason, BS, Viral Patel, MD, and Neda Nikbakht, MD, PhD

Key words: atypical fibroxanthoma; dermatopathology; management; pleomorphic dermal sarcoma.

INTRODUCTION

Pleomorphic dermal sarcoma (PDS) and atypical fibroxanthoma (AFX) are unusual and rare tumors of the tissue mesenchyme. Because the morphology and clinical presentation of PDS and AFX are similar, there is disagreement over whether AFX and PDS are separate clinical conditions or variants of the same condition. Researched and studied far more heavily than PDS, AFX typically affects elderly patients and those with excessive sun exposure.¹ Several other risk factors for AFX include organ transplantation, radiation, and xeroderma pigmentosum.^{2,3}

Although PDS shares histopathologic characteristics with AFX, PDS presents with a more aggressive clinical course, including the potential for metastases and recurrence.⁴ Because of the rare occurrence of PDS, the exact histogenesis, clinical course, and treatment have yet to be elucidated. Here, we report a case of PDS, affecting the scalp of an elderly man and provide a review of the literature focused on the diagnosis and treatment of PDS as well as its distinction from AFX.

CASE REPORT

A 72-year-old white male presented to the dermatology clinic for evaluation of a new scalp lesion of 1-week duration. The patient reported intermittent bleeding with mechanical irritation but denied pain or pruritus. On examination, he had a soft, pink-to-purple, ulcerated, ill-defined nodule with hemorrhagic crusting measuring approximately 2.5 cm on

Abbreviations used:

AFX: atypical fibroxanthoma
MFH: malignant fibrous histiocytoma
PDS: pleomorphic dermal sarcoma

the vertex scalp (Fig 1, A). A shave biopsy of the lesion was performed, and histologic examination revealed a dermal neoplasm with pleomorphic spindle-shaped to epithelioid cells, enlarged nuclei, prominent nucleoli, and several atypical mitotic figures extending to the base of the lesion (Fig 2, A and B). The neoplasm was stained strongly with CD10 and faintly with CD163 and smooth muscle actin. S100, avian v-ets erythroblastosis virus E26 oncogene homolog (ERG), and pan-cytokeratin (PANCKER) stains were negative (Fig 2, C-H). These histologic findings, including margin involvement by the tumor, and the clinical findings of lack of definition, asymmetry, and size >2 cm, raised concern for PDS. Given this concern, a positron emission tomography/computed tomography scan was performed to rule out distant metastasis and revealed no lymph node or systemic involvement.

The lesion was treated with wide local excision with 3-cm margins to the level of the pericranium (Fig 1, B). Resection sized the residual tumor at 1.5 cm with an invasion into subcutaneous tissue with a mitotic rate of 10 mitoses per 10 high-power fields, histologic grade 2, and tumor staging of T1.

From the Department of Dermatology and Cutaneous Biology, Thomas Jefferson University, Philadelphia, Pennsylvania.

Funding sources: Dr Nikbakht is supported by the Skin Cancer Foundation, United States Todd Nagel Memorial Research Grant.

IRB approval status: Not applicable.

Correspondence to: Neda Nikbakht, MD, PhD, Department of Dermatology and Cutaneous Biology, Thomas Jefferson University 233 S. 10th 23 Street, Rm 410 Philadelphia PA 19107. E-mail: neda.nikbakht@jefferson.edu.

JAAD Case Reports 2022;29:123-6.

2352-5126

© 2022 Published by Elsevier on behalf of the American Academy of Dermatology, Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<https://doi.org/10.1016/j.jdc.2022.09.002>



Fig 1. **A**, Initial presentation; pink-to-purple, ulcerated, ill-defined nodule with hemorrhagic crusting measuring approximately 2 cm on the posterior aspect of vertex of the scalp. **B**, One-week following wide local resection and O-Z flap repair; well-healing incision with sutures in place. **C**, Four-months postoperation; well-healed incision with slight atrophy.

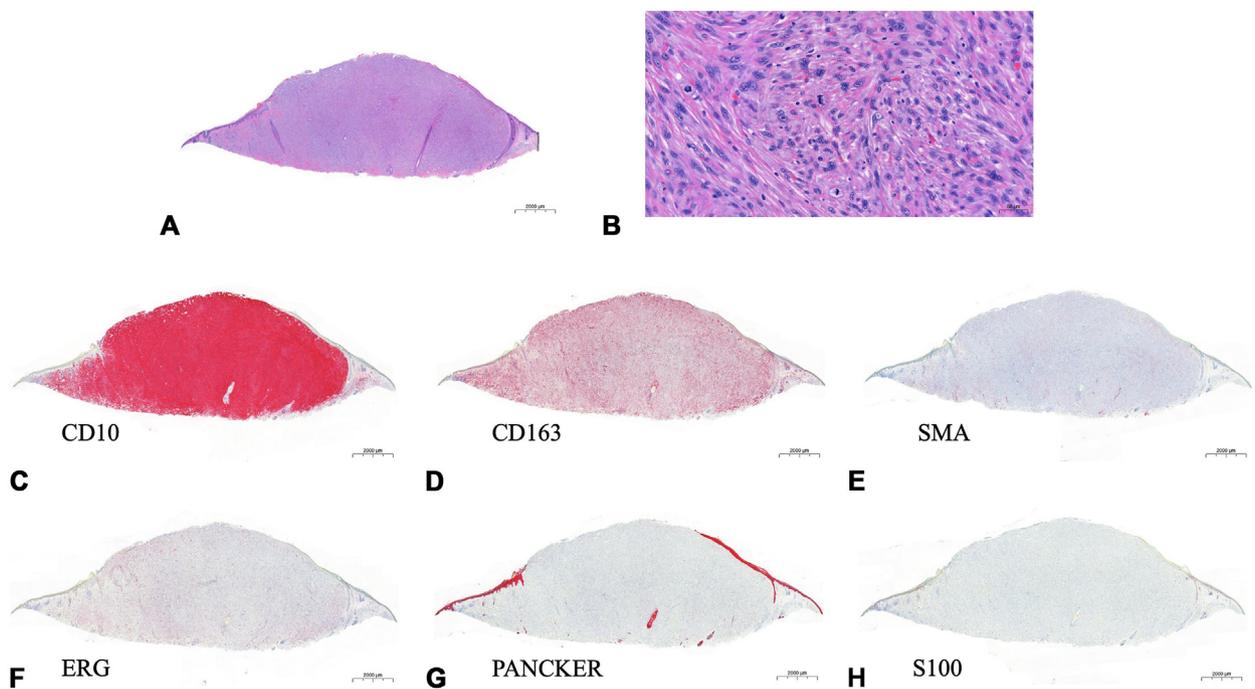


Fig 2. **A**, There is a dense, nodular proliferation within the dermis with a lateral collarette and epidermal ulceration. **B**, High-power view shows pleomorphic spindle-shaped and epithelioid cells with enlarged hyperchromatic nuclei, prominent nucleoli, and several mitotic figures. **C-H**, The neoplasm highlights strongly with CD10 and faintly with CD163 and smooth muscle actin stains. ERG, pancytokeratin (PANCKER), and S100 stains are negative. (A and B, Hematoxylin-eosin stain; original magnifications: A, $\times 10$; B, $\times 100$; C, CD10 stain; original magnification: C, $\times 10$; D, CD163 stain; original magnification: D, $\times 10$; E, SMA stain; original magnification: E, $\times 10$; F, ERG stain; original magnification: F, $\times 10$; G, PANCKER stain; original magnification: G, $\times 10$; H, S100 stain; original magnification: H, $\times 10$.)

The presence of neoplastic invasion into the subcutaneous fat and overall immunohistochemical and histopathology presentation supported the diagnosis

of PDS. Because the procedure yielded negative resection margins, adjuvant radiotherapy was not indicated.

Within 4 months of excision, he presented for follow-up with a well-healed surgical site without clinical evidence of recurrence (Fig 1, C). Although there are no standardized management guidelines for PDS, follow-up appointments were planned every 6 months for the next 3 years and annually thereafter based on the National Comprehensive Cancer Network's 2022 clinical practice guidelines for soft-tissue sarcomas.⁵

DISCUSSION

The precise definitions, diagnostic criteria, and terminology of PDS and its related neoplasms are a topic of extensive debate. Historically, the terms AFX and malignant fibrous histiocytoma (MFH) were used on a spectrum, with more superficial and deeper tumors termed AFX and MFH, respectively. As histologic techniques advanced, many tumors that were previously classified as MFH were found to be falsely diagnosed and recategorized to other types of tumors. As such, the term MFH has become antiquated and replaced by the term undifferentiated pleomorphic sarcoma. To further complicate matters, it is argued that the term PDS better describes tumors of cutaneous origin because the term undifferentiated pleomorphic sarcoma also includes a variety of malignant soft-tissue neoplasms.⁶

Because PDS presents with an aggressive clinical course, including a high rate of metastasis, a high chance of recurrence, and deep tissue invasion, the need for a timely and accurate diagnosis is apparent.^{7,8} Histologically, AFX and PDS share similar immunohistochemical profiles. The major distinguishing features of PDS are the presence of tumor necrosis, lymphovascular invasion, and perineural infiltration.⁷ In addition, AFX presents with proximity to the epidermis with limited depth of invasion, whereas PDS's presentation is diffusely infiltrative with more aggressive histopathologic behavior.^{7,8} Clinical distinguishing features include a typically <2 cm lesion size for AFX and a larger lesion diameter for PDS. Furthermore, PDS is commonly ill-defined and asymmetric, whereas AFX often presents as well-defined lesions.^{7,8}

At this time, there are no published guidelines on the management and follow-up of PDS.⁹ However, general management guidelines for soft-tissue sarcomas may be used for reference. Surgical treatment with wide location excision remains the first-line therapy for PDS.⁴ Although specific guidelines for margin control are not well established, recent data suggest that 95% of cutaneous undifferentiated pleomorphic sarcoma tumors can be cleared with uniform peripheral surgical margins of 3 cm.¹⁰

For patients with surgically treated low-grade soft-tissue sarcomas of the head and neck with negative margins, the National Comprehensive Cancer Network's recommends follow-up every 3 to 6 months for the first 2 to 3 years, and annually thereafter.⁵ Imaging of the primary site may not be required in situations where the area is easily followed by a physical examination. Patients with advanced disease and those for whom negative surgical margins cannot be achieved should be considered for resection or adjuvant radiotherapy to decrease the risk of local recurrence. Although no imaging guidelines exist for AFX or PDS, imaging may be particularly useful in PDS to evaluate local infiltration before surgery because of the higher risk of metastasis.

The potential for metastases and recurrence vary significantly among PDS and AFX. For PDS, the rate of metastasis is between 8.8% and 20%, whereas AFX has a metastatic potential of 1% to 2%.⁴ Following treatment, the PDS recurrence rate is 17% to 35%, whereas AFX has a recurrence rate of 4.6% to 11.3%.

We present a case of PDS occurring on the scalp of a 72-year-old male patient, successfully treated with wide local excision. This case highlights the morphologic and histologic characteristics of PDS and discusses what is known about the management of this uncommon malignancy.

Conflicts of interest

None disclosed.

REFERENCES

- Lonie S, Yau B, Henderson M, Gyorki D, Angel C, Webb A. Management of pleomorphic dermal sarcoma. *ANZ J Surg*. 2020;90(11):2322-2324. <https://doi.org/10.1111/ans.15909>
- Wollina U, Schönlebe J, Bujok V, et al. Dermal pleomorphic sarcoma of the scalp - report of two cases. *Open Access Maced J Med Sci*. 2019;7(18):2982-2984. <https://doi.org/10.3889/oamjms.2019.274>
- Griewank KG, Wiesner T, Murali R, et al. Atypical fibroxanthoma and pleomorphic dermal sarcoma harbor frequent NOTCH1/2 and FAT1 mutations and similar DNA copy number alteration profiles. *Mod Pathol*. 2018;31(3):418-428. <https://doi.org/10.1038/modpathol.2017.146>
- Miller K, Goodlad JR, Brenn T. Pleomorphic dermal sarcoma: adverse histologic features predict aggressive behavior and allow distinction from atypical fibroxanthoma. *Am J Surg Pathol*. 2012;36(9):1317-1326. <https://doi.org/10.1097/PAS.0b013e31825359e1>
- National Comprehensive Cancer Network. NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) Soft Tissue Sarcoma. May 17, 2022. Accessed July 13, 2022. https://www.nccn.org/professionals/physician_gls/pdf/sarcoma.pdf
- Soleymani T, Aasi SZ, Novoa R, Hollmig ST. Atypical fibroxanthoma and pleomorphic dermal sarcoma: updates on classification and management. *Dermatol Clin*. 2019;37(3):253-259.
- Bowe CM, Godhania B, Whittaker M, Walsh S. Pleomorphic dermal sarcoma: a clinical and histological review of 49 cases. *Br J*

- Oral Maxillofac Surg.* 2021;59(4):460-465. <https://doi.org/10.1016/j.bjoms.2020.09.007>
8. Cates JMM. The AJCC 8th edition staging system for soft tissue sarcoma of the extremities or trunk: a cohort study of the SEER database. *J Natl Compr Canc Netw.* 2018;16(2):144-152. <https://doi.org/10.6004/jnccn.2017.7042>
 9. Tardío JC, Pinedo F, Aramburu JA, et al. Pleomorphic dermal sarcoma: a more aggressive neoplasm than previously estimated. *J Cutan Pathol.* 2016;43(2):101-112. <https://doi.org/10.1111/cup.12603>
 10. Jibbe A, Worley B, Miller CH, Alam M. Surgical excision margins for fibrohistiocytic tumors, including atypical fibroxanthoma and undifferentiated pleomorphic sarcoma: a probability model based on a systematic review. *J Am Acad Dermatol.* 2022;87(4):833-840. <https://doi.org/10.1016/j.jaad.2021.09.036>