Surgical Approach of Pleomorphic Dermal Sarcoma on the Scalp

Gloria M. M. Maldonado\(^1\), Ross M. Meaden\(^1\), Alexander J. Kaminsky\(^2\)

\(^1\)??? SUNY Upstate Medical University, Syracuse, \(^2\)Section of Plastic Surgery, United Health Services, Binghamton, NY, USA

Abstract

Pleomorphic dermal sarcoma (PDS) is a fast-growing mesenchymal tissue tumor with similar characteristics to an atypical fibroxanthoma (AFX) presenting a significant clinical challenge to diagnose for physicians. We report a 79-year-old male presenting with a 3-month history of a lesion on his scalp that had been previously superficially biopsied yielding a diagnosis of PDS or AFX. Following a second biopsy, new findings led to the diagnosis of PDS. A wide local excision with 2 cm margin with delayed split-thickness skin graft reconstruction was performed. Treatment for PDS favors wide local excision with a 2–3 cm margin and adjuvant radiotherapy if perineural involvement. We support the following recommendations: full-thickness appropriate tissue diagnosis, pulmonary computed tomography scan, or X-ray to rule out metastasis, followed by a multidisciplinary team evaluation. These complex cases should be presented at a tumor board and tailored treatments should be based on patient risk factors and relevant history.

Keywords: Atypical fibroxanthoma, cutaneous sarcoma, pleomorphic dermal sarcoma, surgical treatment

Introduction

Pleomorphic dermal sarcoma (PDS) is a rare mesenchymal tissue tumor, presenting as a fast-growing, exophytic, ulcerated, bleeding lesion on sun-exposed areas\(^{1,2}\). It is common in older men, typically presenting on the scalp, face, and neck\(^{1,2}\). Sun-exposed areas are primarily affected because PDS has a high ultraviolet-induced genetic mutation signature\(^{1,2}\). The incidence and prevalence of PDS are unknown due to its clinical and histological similarity to atypical fibroxanthoma (AFX)\(^{1,4-6}\). It is uncertain whether PDS represents a more aggressive AFX variant or a distinct pathologic entity, and thus, requiring a more aggressive treatment. We present the surgical intervention of a scalp PDS.

Case Report

A 79-year-old male, with past medical history of left parotid tumor, type 2 diabetes, chronic lung nodules, severe bronchiectasis requiring chronic steroid use, and multiple actinic keratoses, presented for a 20 mm × 13 mm lesion on the posterior scalp vertex present for 3 months [Figure 1a]. Previously tangential biopsy by dermatology showed superficial portions of atypical spindle cells suspicious for AFX or PDS. An incisional biopsy taken to better categorize the lesion showed pleomorphic spindle cell neoplasm occupying the dermis up to the upper subcutis with typical and atypical spindle cells dermal proliferation and many mitotic figures with neoplastic processes surrounding nerve bundles and blood vessels. Based on these new histological findings, a PDS diagnosis was given.

It was decided to perform a wide local excision with a 2 cm margin and intraoperative frozen pathology yielding an 8 cm × 7 cm × 0.4 cm final defect [Figure 1b]. The frozen section showed clear margins with no lymphovascular invasion but with multifocal perineural involvement. Two weeks postexcision, a full-thickness skin graft harvested from the abdomen with calvarium burring was performed [Figure 1c]. Approximately 23% of graft nontake developed and a conservative approach with local wound care was taken [Figure 1d]. The wound improved with Promogran and Mepilex covering [Figure 1e], and the final full-thickness skin graft was successful [Figure 1f].

Address for correspondence: Gloria M. M. Maldonado, 500 Harrison St., Apt 806B, Syracuse 13202, NY, USA. E-mail: munaycog@upstate.edu

Received: 26 October 2023, Revised: ???, Accepted: 05 February 2024, Published: ***

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Maldonado GM, Meaden RM, Kaminsky AJ. Surgical approach of pleomorphic dermal sarcoma on the scalp. J Dermatol Dermatol Surg 2024;XX:XX-XX.
Due to the concern for perineural involvement, the patient was referred to a tertiary academic center for further evaluation. Biopsies obtained by the tertiary referral otolaryngologist 3 months postlocal wide excision revealed granulation tissue with no malignancy and a repeat review of the original pathology slides revealed no perineural invasion. The patient followed up every 6 months with dermatology for recurrence monitoring and regular skin cancer surveillance. Unfortunately, he passed away from reasons not related to his PDS.

**Discussion**

PDS and AFX are difficult to differentiate due to their overlapping clinical, morphologic, and histologic presentation on superficial biopsy.\[^{1,5,6}\] In a case series, 75% of PDS had initially been diagnosed as AFX.\[^{6}\] A deeper biopsy is necessary to differentiate these two entities as PDS, unlike AFX, must have \(\geq 1\) of the following: tumor necrosis, deep subcutaneous tissue invasion, and lymphovascular and/or perineural invasion.\[^{1,2,5,7}\] Mortality increases with a tumor size \(\geq 2\) cm, advanced age, any kind of immunosuppression, and lymphovascular invasion.\[^{3,7,8}\] PDS recurrence rate is higher than AFX, with a reported 10%–20% PDS recurrence rate.\[^{1,7}\] and distant organ metastasis most commonly being the lungs.\[^{3,6-8}\] Thus, PDS staging should include a pulmonary computed tomography (CT) scan or X-ray to rule out lung and lymph node metastasis.\[^{9}\]

PDS has no gold standard treatment; first-line management is wide local excision.\[^{9}\] Even though the 2 cm margin was widely employed, a recent publication found that an excision margin of \(\geq 3\) cm is preferred because it reduces the recurrence rate based on probability models.\[^{3}\] Radiotherapy is the preferred adjuvant treatment for metastasis, local recurrence, and perineural or lymphovascular invasion.\[^{10}\] In our case, multifocal perineural involvement was initially reported, but adjuvant radiotherapy was not pursued because pathology specimens re-review found no perineural involvement. Our recommendations include the following: full-thickness appropriate tissue diagnosis, pulmonary CT scan, or X-ray to evaluate for lung and lymph node metastasis, followed by a multidisciplinary team approach which includes dermatology, oncology, surgical oncology, radiation oncology, and plastic and reconstructive surgery, case presentation at a tumor board, and tailored treatments based on patient risk factors and relevant history.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.
Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

References
1. Lo AC, McDonald S, Wong KY. Case of pleomorphic dermal sarcoma with systematic review of disease characteristics, outcomes and management. BMJ Case Rep 2021;14:e244522.

Author Queries???
AQ1: Kindly provide department.
AQ2: Kindly provide department if applicable.
AQ3: Kindly mention Dr. or Prof.
AQ4: Kindly check the formation of address.
AQ5: Kindly provide revised date