An atypical presentation of a primary dermal leiomyosarcoma

Robert Dazé, DO, PGY-3[1], Krina Chavda, DO, FAOCD[1], Keith Baribault, MD[2], Richard Miller DO, FAOCD[1]
[1]Largo Medical Center, Department of Dermatology, Largo, Florida
[2]Dermpath Diagnostics, Tampa, Florida

HCA Healthcare/USF Morsani College of Medicine GME Programs

Introduction

Leiomyosarcoma (LMS) is a rare malignant tumor with smooth muscle differentiation. LMS is classified into dermal or subcutaneous variants that are derived from the arrector pili muscles or the small- to medium-sized blood vessels of the subcutaneous tissue, respectively. These tumors usually present in the 5th to 7th decade of life and affect men twice as frequently as women. The most common area of cutaneous presentation is the lower extremity. While metastasis is rare from a dermal LMS, the current therapeutic modalities include excision, radiation, and Mohs microscopic surgery. We report a rare case of a primary cutaneous LMS that was successfully treated with a wide local excision using 1-cm margins.

Case Report

A 58-year-old Caucasian female with a past medical history of atrial fibrillation, hypertension, hyperlipidemia, and obstructive sleep apnea presented for evaluation of a solitary lesion present on her left anterior shoulder that had been present for approximately 9 months prior to examination. She reported rapid growth of the lesion since its onset, but denied any symptoms including pain, pruritus, bleeding, or ulceration. Physical exam demonstrated a 1.5-cm erythematous to violaceous polypoid, well circumscribed nodule with superficial telangiectasias (Figures 1 & 2). There was no significant lymphadenopathy. Review of systems was negative for constitutional, cardiopulmonary, gastrointestinal, and genitourinary symptoms. Microscopic examination of a shave biopsy revealed cellular fascicles of atypical smooth muscle cells with scattered atypical mitotic figures (Figures 3-6). The lesion was diffusely positive for smooth muscle actin and desmin immunohistochemical stains (Figures 7-8). P63 stain was focally positive, but mostly negative. Stains for pan-cytokeratin, cyokeratin 5/6, S100, SOX-10, CD34 and Factor XIIIa were negative. The above clinical and histopathological correlation confirmed the diagnosis of a primary cutaneous leiomyosarcoma.

Clinical Images

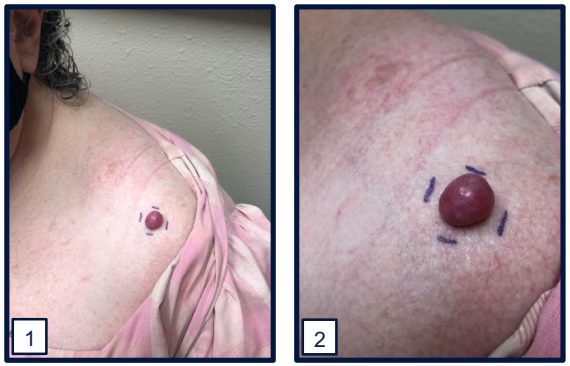

Figure 1 & 2: 1.5-cm erythematous to violaceous polypoid nodule present on the left anterior shoulder

Histopathology

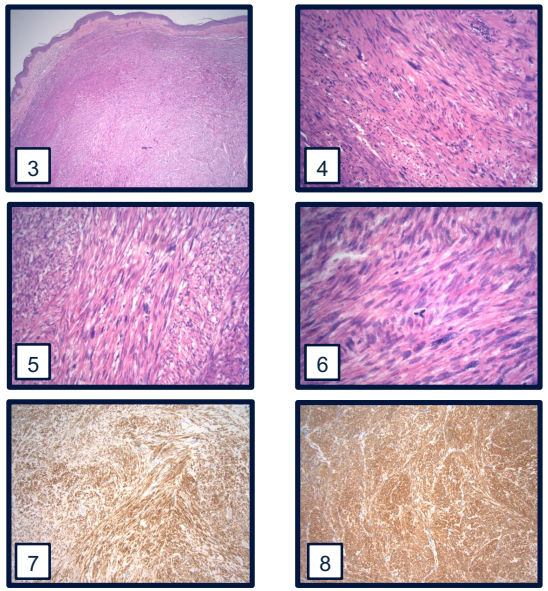

Figure 3 (H&E 40x), Figure 4 (H&E, 200x), Figure 5 (H&E, 200x), Figure 6 (H&E, 400x): Proliferation of atypical smooth muscle cells with scattered mitotic figures

Figure 7 (desmin, 100x), Figure 8 (smooth muscle actin, 100x): both immunohistochemical stains were diffusely positive

Discussion

With its clinical ambiguity, initial differentials included basal cell carcinoma, merkel cell carcinoma, and a cutaneous metastasis as the patient was not up to date on age appropriate cancer screenings proposed by the United States Preventative Services Task Force. However, the LMS was confirmed on histopathological examination and further confirmed with appropriate immunohistochemical stains such as smooth muscle actin and desmin.

The current treatment modalities include local excision with narrow margins, wide local excision with at least a 1-cm margin, wide local excision plus radiation, and Mohs microscopic surgery. With all treatment modalities, margin control is the strongest predictor of clinical prognosis in the treatment of LMS. Therefore, given the absence of lymphadenopathy and systemic symptoms, the decision was made to proceed with wide local excision with a 1-cm margin. The tumor was completely excised, and she was referred to hematology-oncology for further evaluation.

Due to the lack of standardization of ongoing surveillance and management, oncology currently recommends a computed tomography of the chest yearly for the first two years and then a plain chest radiograph every year thereafter to monitor for distal metastasis. The most common site for internal metastasis is the lung and the most common site for cutaneous metastasis is the scalp. Furthermore, the patient is recommended to have full body skin examinations every 6 months for the first two years, then yearly thereafter.

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