Rapidly growing asymptomatic violaceous nodule

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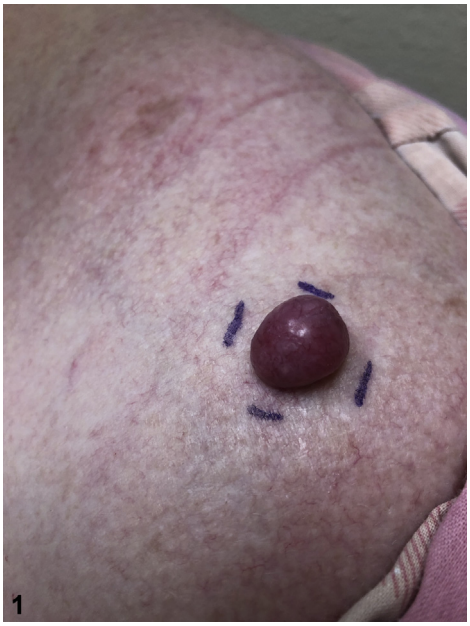

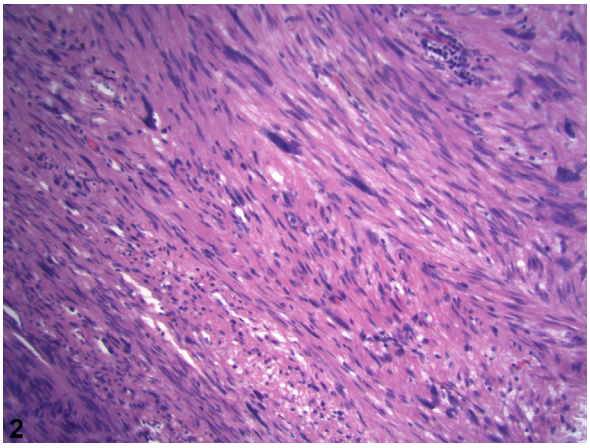

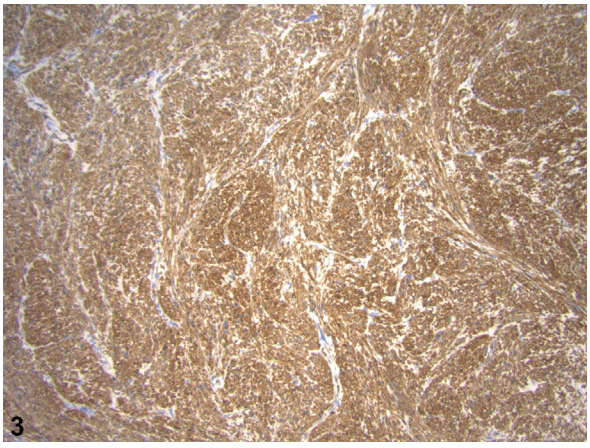

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A 58-year-old woman presented with a solitary lesion on her left anterior shoulder that started developing approximately 9 months prior to the examination (Fig 1). She reported a rapid growth of the lesion but denied any symptoms. A review of systems was negative. Physical examination demonstrated a 1.5-cm violaceous polypoid nodule without any lymphadenopathy. She denied any history of trauma or radiation to the area. Histology revealed atypical smooth muscle cells with scattered mitotic figures arranged in fascicles (Fig 2). The lesion was positive for smooth muscle actin (SMA) and desmin immunohistochemical stains (Fig 3). The stains for pan-cytokeratin, cytokeratin 5/6, S100, SRY-related HMG-box 10, CD34, and factor XIIIa were negative.

Question 1: What is the most likely diagnosis?

A. Desmoplastic melanoma

B. Dermal leiomyosarcoma (LMS)

C. Spindle cell squamous cell carcinoma (SCC)

D. Merkel cell carcinoma

E. Atypical fibroxanthoma (AFX)

Answers:

A. Desmoplastic melanoma — Incorrect. The histologic differential diagnosis of atypical spindle cell neoplasms should include desmoplastic melanoma, AFX, spindle cell SCC, and LMS. Immunostains such as SMA and desmin will not be positive in desmoplastic melanomas; rather, neuroectodermal markers such as S100 and SRY-related HMG-box 10 will be positive.[1]

B. Dermal leiomyosarcoma (LMS) — Correct. LMS is a rare malignant sarcoma with smooth muscle differentiation that typically presents in men in a ratio of approximately 3:1.[2,3] It is characterized by interlacing bundles or fascicles of spindled tumor cells with atypical cigar-shaped nuclei that stain positive for mesenchymal markers such as SMA and desmin, with SMA present in virtually all tumors.[1,3]

C. Spindle cell squamous cell carcinoma (SCC) — Incorrect. Spindle cell SCC would stain positive for high molecular weight keratins such as AE1/AE3, CK903, and p63. Moreover, SCC would be negative for myogenic markers such as SMA and desmin.[1]

D. Merkel cell carcinoma — Incorrect. Merkel cell carcinoma histologically presents as a small blue cell tumor highlighted by neuroendocrine markers such as chromogranin, neuron-specific enolase, and synaptophysin.[1] Additionally, it will demonstrate a classic perinuclear dot pattern with CK20.

E. Atypical fibroxanthoma (AFX) — Incorrect. AFX is considered a diagnosis of exclusion as a number of immunohistochemical stains, such as CD10, will react but none of these are entirely specific.[1] Up to one-third of AFXs can be positive for SMA, thus, requiring an additional myogenic marker such as desmin, which would be negative.[1]

Question 2: Where does this tumor most commonly metastasize to?

A. Scalp

B. Small bowel

C. Liver

D. Lungs

E. Brain

Answers:

A. Scalp — Correct. The skin was the most common site for distal metastasis of a primary dermal LMS, with the scalp as the most common location.[2,4]

B. Small bowel — Incorrect. Only subcutaneous LMS has been known to metastasize to the small bowel, not primary dermal LMS. Small bowel metastases account for only a limited number of cases.[2]

C. Liver — Incorrect. Hepatic metastases have been known to occur in limited cases originating from subcutaneous LMS, not dermal LMS. In addition, subcutaneous tumors are associated with a greater disease-specific mortality compared with its dermal counterpart.[2]

D. Lungs — Incorrect. Primary dermal LMS has been documented to metastasize to the lungs; however, this is not the most common site.[2] In contrast, subcutaneous LMS carries a greater metastatic potential with the most common visceral site being the lungs.[2,3]

E. Brain — Incorrect. To date, there have been no documented cases of primary dermal LMS metastasis to the brain.[2]

Question 3: Which of the following modalities is considered the gold standard of treatment for this particular tumor?

A. Excision with narrow margins

B. Wide local excision

C. Radiation

D. Mohs micrographic surgery

E. Doxorubicin

Answers:

A. Excision with narrow margins — Incorrect. As margin control is the strongest predictor of clinical outcomes, excision with narrow margins is not recommended. Comparatively, narrow surgical margins are associated with the highest rates of recurrence, metastasis, and disease-specific mortality.[2,5]

B. Wide local excision — Correct. Because of the increased risk of local recurrence and metastasis, wide local excision is considered the gold standard of treatment. Most of the literature recommends a 1-cm margin, whereas other sources advocate 2-cm to 5-cm margins.[2,3,5]

C. Radiation — Incorrect. Radiation can be used adjunctively with primary surgical resection for deep LMS.[2] This adjuvant treatment is typically reserved for patients with large lesions (>5 cm), tumor-positive excision margins, high-grade LMS, and after local relapse.[3]

D. Mohs micrographic surgery — Incorrect. Although Mohs micrographic surgery is associated with superior results in terms of lower recurrence rates, it is not the current standard of care as the available data supporting this modality are limited to small case reports and case series.[3,5] For sites where wide local excision may not be appropriate, Mohs micrographic surgery is a valid alternative.[3]

E. Doxorubicin — Incorrect. Doxorubicin in combination with ifosfamide or dacarbazine is considered the first-line therapy for metastatic disease as LMS is considered an anthracycline-sensitive sarcoma.[4] This therapy would be inappropriate for the treatment of a primary dermal LMS as surgical resection with wide margins is the treatment of choice.[3,5]

Abbreviations used:

AFX: atypical fibroxanthoma
LMS: leiomyosarcoma
SCC: squamous cell carcinoma
SMA: smooth muscle actin

Conflicts of interest

None disclosed.

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