An unusual painful scar

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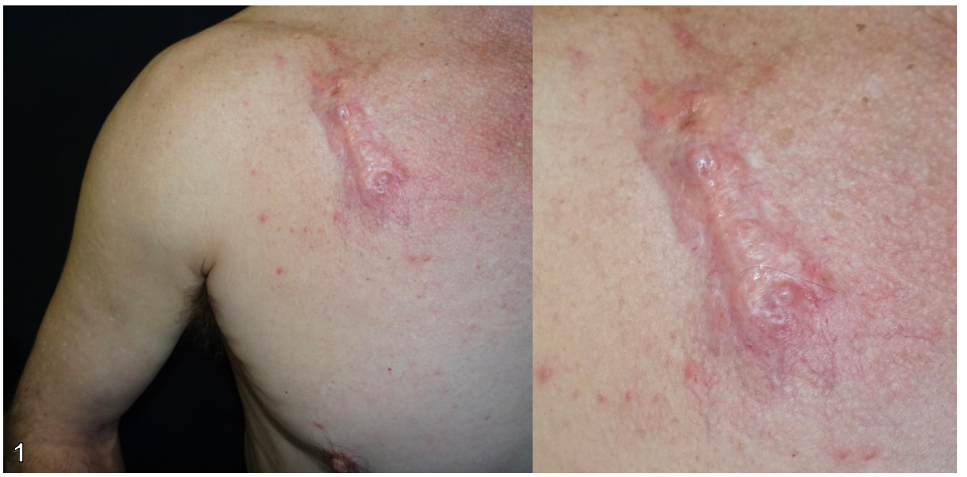

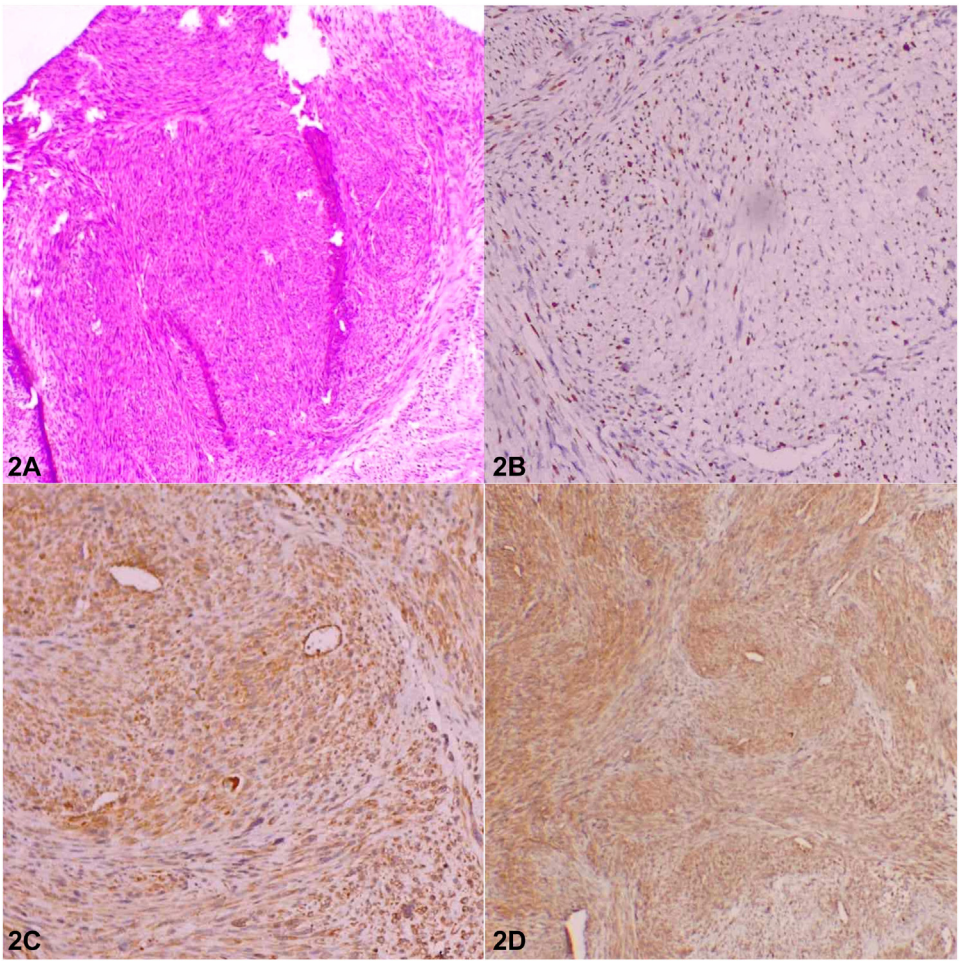

A 50-year-old Hispanic man presented to our dermatology department with a 2-year history of a painful scar on his right pectoral region (Fig 1). The lesion developed 1 month after suffering a traumatic injury with a metal object. He underwent treatment with triamcinolone injections and surgical resection. Despite these treatments, the scar recurred within a few weeks. A skin biopsy reported a poorly circumscribed spindle cell neoplasm in the dermis, characterized by elongated nuclei, nuclear pleomorphism, and mitosis (Fig 2, *A*). Immunostaining was positive for, Ki67 (Fig 2, *B*), smooth muscle actin (Fig 2, *C*) and vimentin (Fig 2, *D*), while S-100 stain was negative.

Question 1: What is the most likely diagnosis?

A. Keloid scar

B. Dermatofibrosarcoma protuberans

C. Cutaneous leiomyosarcoma (LMS)

D. Cutaneous leiomyoma

E. Fibrosarcoma

Answers:

A. Keloid scar — Incorrect. Keloids are more common among Asian, Hispanic, and African American populations. Clinically, they present as firm lesions with a smooth surface that extends beyond the borders of the original wound and can be associated with itching or pain. The diagnosis is less likely due to the resistance to treatment; furthermore, histological findings are not consistent with a nonmalignant entity.

B. Dermatofibrosarcoma protuberans — Incorrect. Dermatofibrosarcoma protuberans is the most common type of skin sarcoma. Typically presenting as an asymptomatic and slowly growing plaque with an atrophic or sclerodermiform surface on the trunk or limbs. Histopathology shows spindle cells neoplasm arranged in a storiform pattern with variable mitotic activity.

C. Cutaneous leiomyosarcoma (LMS) — Correct. Cutaneous LMS is a malignant tumor derived from smooth muscle with an estimated incidence of 0.6/1,000,000/y.[1] Caucasian men over the age of 50 years are more commonly affected. Cutaneous LMS appears as a firm isolated or multiple and confluent nodules with a violaceous surface. Usually ranging from 1 to 5 cm in size, mostly arising in the extremities, and the head and neck region. The metastatic form primarily affects the scalp and the trunk.[2] Associated symptoms like pruritus, paresthesia, and pain, are present in 63% of patients.[3]

D. Cutaneous leiomyoma — Incorrect. Cutaneous leiomyoma is a benign smooth muscle tumor with variable clinical appearance. Normally, cutaneous leiomyoma presents as small symmetrical papules or nodules with a slow progression and does not affect deep tissues, unlike cutaneous LMS.

E. Fibrosarcoma — Incorrect. Fibrosarcoma is a slow growing deep soft tissue sarcoma that mainly affects young adults as a subcutaneous lesion with secondary involvement of the skin. The extremities are the most common site of appearance, frequently associated with radiation exposure or burns.

Question 2: Which of the following is not considered a poor prognostic factor?

A. The subcutaneous histopathological variant

B. Positive vimentin and smooth muscle-specific actin immunophenotype

C. Tumor size over 5 cm

D. Fascia involvement

E. High histological grade

Answers:

A. The subcutaneous histopathological variant — Incorrect. Cutaneous LMS has 3 main

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clinicopathological variants: the superficial or dermal form, the subcutaneous form, and the metastatic form. The prognosis varies between superficial and subcutaneous LMS, with a local recurrence rate of 24.4% and 36.63%, and a mortality rate of 3.3% and 37.82% respectively.[4] Metastatic LMS has an average survival of 16 months.

B. Positive vimentin and smooth muscle-specific actin immunophenotype — Correct. Cutaneous LMS is distinguished from other fusiform cell neoplasms by positive smooth muscle markers such as vimentin, desmin, h-caldesmon, smooth muscle-specific actin, alpha-smooth muscle actin, and myosin. Of note, these markers are also positive in its benign counterpart — cutaneous leiomyoma.

C. Tumor size over 5 cm — Incorrect. The American Joint Committee on Cancer established a size of 5 cm or greater as a distinct poor prognosis factor of neoplasms in their staging system for soft tissue sarcomas.

D. Fascia involvement — Incorrect. The depth of the tumor with involvement of the fascia has been associated with an increased risk of metastasis, local recurrence, and mortality.

E. High histological grade — Incorrect. The histologic grade is determined based on the degree of cellular differentiation, the mitotic count, and the percentage of tumor necrosis. It is a determinant in the prognosis of soft tissue sarcomas.

Question 3: What would be the treatment of choice?

A. Cryosurgery

B. Surgical resection

C. Mohs surgery

D. Chemotherapy with doxorubicin and dacarbazine

E. Tyrosine kinase inhibitors

Answers:

A. Cryosurgery — Incorrect. Cryosurgery has been successfully used in cases of cutaneous leiomyomas, with good esthetic results and low tumoral recurrence. However, the malignant nature of cutaneous LMS makes the use of this therapy inappropriate.

B. Surgical resection — Correct. There are no randomized clinical trials defining the best therapeutic modality for cutaneous LMS. The available evidence to date, based on reports and case series, supports surgical resection with wide margins as the first-line therapy, with peripheral margins of 3 to 5 cm and deep margins to the fascia.[5]

C. Mohs surgery — Incorrect. While effective, Mohs surgery has not shown superiority over wide surgical resection. Mohs surgery may be preferred in cosmetically sensitive anatomical regions.

D. Chemotherapy with doxorubicin and dacarbazine — Incorrect. Recommendations for the neoadjuvant treatment of cutaneous LMS with chemotherapy are not specific. Treatment is established on an individual basis in response to indicators of poor prognosis. Based on review cases in medical literature, nonsurgical treatments are not considered first-line treatments.

E. Tyrosine kinase inhibitors — Incorrect. The overexpression of tyrosine kinase receptors such as insulin-like growth factor receptor and platelet-derived growth factor receptor has been shown in cutaneous LMS. Currently, tyrosine kinase inhibitors such as pazopanib, are being studied for recurrent, metastatic, or refractory cases.

Abbreviation used:

LMS: cutaneous leiomyosarcoma

Conflicts of interest

None disclosed.

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