

Pleomorphic soft tissue sarcoma metastatic to the skin of the scalp and groin

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INTRODUCTION

Rarely, the dermatologist may encounter a primary cutaneous pleomorphic sarcoma (also known as a *malignant fibrous histiocytoma*), an undifferentiated neoplasm thought to be of mesenchymal origin.¹ These uncommon skin tumors typically manifest as tender subcutaneous nodules. In contrast to primary cutaneous sarcomas, we present a case of a soft tissue pleomorphic sarcoma with multiple metastases to the cutaneous scalp and the groin. Although previous reports have described leiomyosarcoma metastatic to the skin, no report to our knowledge has detailed a widely metastatic undifferentiated sarcoma with lesions in multiple skin sites.

CASE REPORT

A 75-year-old white man with a history of prostate cancer treated with radiotherapy presented to his primary care physician with reproducible chest, back, and shoulder pain; painless swelling in his left leg; and nodules on the skin of his scalp and groin. The patient returned home with a trial of cyclobenzaprine for presumed musculoskeletal strain and a planned outpatient dermatology appointment. Within a month, the patient returned to the emergency department with severe weakness, confusion, anorexia, and purulent, bloody drainage from several scalp nodules. Skin examination found multiple flesh-colored to violaceous nodules, the most prominent located on the patient's inguinal groin and vertex scalp (Fig 1). The largest scalp lesion measured approximately 2 cm³ and had central crusting and ulceration (Fig 1, right panel).

Musculoskeletal examination found a left thigh larger than the right, but strength, range of motion, and gait were normal. Initial laboratory results were remarkable for a white blood cell count of 20,000/ μ L, Hgb of 10 g/dL, Ca²⁺ of 10.5 mg/dL, and an alkaline phosphatase level of 326 IU/L. Chest, abdomen, and pelvis computed tomography scan showed destruction of the rib architecture and multiple small pulmonary masses. Brain magnetic resonance images showed enhancing lesions in the temporal lobe, frontal lobe, and the scalp. None of these scalp lesions arose in or invaded the calvaria. A left femoral radiograph showed a large (>10 cm) soft tissue mass. Before transfer to our facility, the patient underwent biopsies of the thigh mass and left femur. Both biopsies showed indeterminate findings.

After transfer to our facility, the dermatology service obtained a punch biopsy section of a cutaneous lesion from the right inguinal area. Routine histology found a dense proliferation of atypical dermal spindle cells with approximately 25 mitotic figures per square millimeter (Fig 2). Lesional cells completely effaced the normal cutaneous architecture, and the biopsy section transected the base of the lesion. Lesional cells were positive for vimentin, negative for pancytokeratin, S-100, SMA, desmin, CD34, CD31, c-Kit, HMB45, and Melan-A. Esophagogastroduodenoscopy-guided biopsy of gastric and duodenal ulcers showed similar vimentin-positive atypical spindle cells. Given this immunohistochemical profile—and the lack of any evidence of spindle cell maturation from either biopsy—no additional immunohistochemistry (eg,

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Fig 1. Examination found multiple flesh-colored to violaceous nodules, the most prominent located on the inguinal groin and vertex scalp (*left and right panel, respectively*).

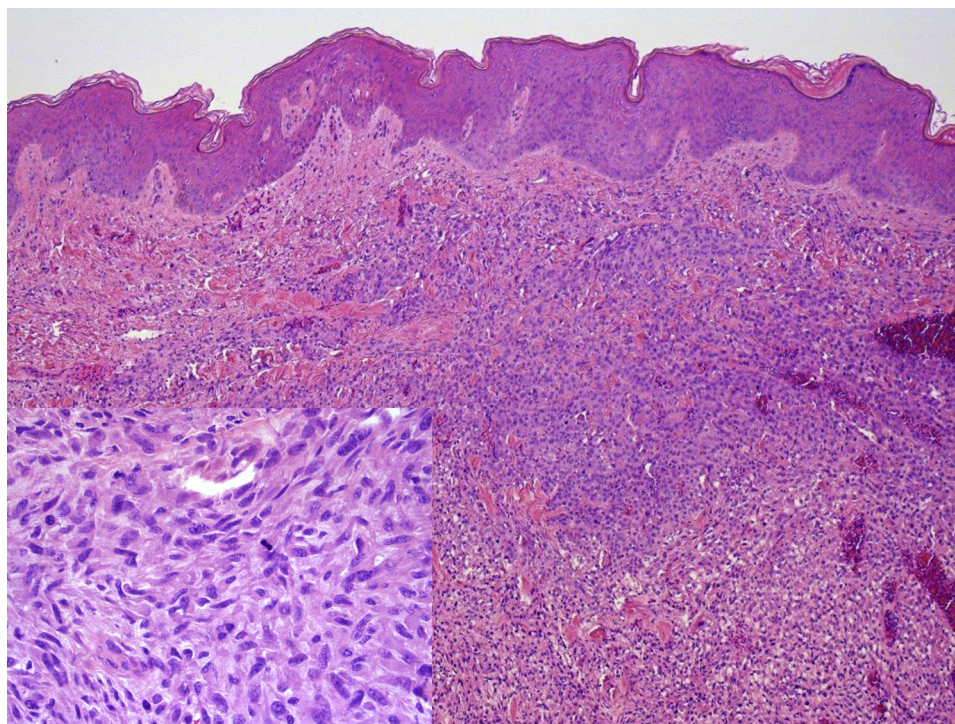


Fig 2. Histology of a cutaneous lesion from the right inguinal area shows a dense proliferation of atypical spindle cells within the dermis with approximately 25 atypical mitotic figures per square millimeter, findings consistent with a high-grade pleomorphic sarcoma. (Hematoxylin-eosin stain.)

MDM2 for dedifferentiated liposarcoma) was performed. In all, the findings led to a diagnosis of widely metastatic high-grade pleomorphic spindle cell sarcoma arising from a large mass in the left thigh.

The patient received treatment for hypercalcemia, hemorrhagic shock caused by bleeding malignant ulcers, and hypoxemic respiratory failure caused by metastatic burden in his lungs. Despite aggressive

intervention in the intensive care unit, the patient continued to deteriorate, and the family decided to transition the patient to comfort care only. The patient died from respiratory complications 7 days after initial hospitalization.

DISCUSSION

Sarcoma arising in any location has a worldwide incidence estimated at 1.8 to 5 cases per 100,000 persons per year, with most arising in individuals over the age of 55.² The most common subtypes include gastrointestinal stromal tumors (about 1 in 5), unclassified (about 1 in 6), liposarcoma (about 1 in 6), and leiomyosarcoma (about 1 in 10).³ Based on histologic features, including morphology and frequency of mitotic cells, the National Cancer Institute guidelines classify sarcomas as low, medium, or high grade, a designation carrying prognostic and therapeutic significance.⁴ For solitary lesions—including those arising in the skin—treatment consists of local resection and, depending on histology and staging, either preoperative or postoperative chemoradiotherapy. Sarcoma preferentially metastasizes to the lung and bones (as in this case), and metastatic sarcoma poses a great therapeutic challenge. Response to chemoradiotherapy hinges on tumor behavior, histologic subtype, and patient characteristics. A recent study showed the most favorable chemotherapeutic outcomes in patients with liposarcoma or synovial histology, with no bone metastases, and who were less than 40 years old.⁵ Regardless, prognosis remains guarded, and median survival in metastatic disease is approximately 1 year.

Of note, soft tissue sarcomas of all histologic subtypes rarely metastasize to the skin. A retrospective study from MD Anderson Cancer Center encompassing 20 years of data and 25,000 histologically confirmed sarcoma cases found skin metastases in only 65 cases (0.26% of all sarcoma cases). Of

those 65 cases, leiomyosarcoma was the most frequent subtype (28 of 65; 43%), with only 4 cases (6%) of pleomorphic sarcoma/malignant fibrous histiocytoma metastatic to the skin.⁶ Concordant with this study, several reports have detailed cutaneous nodules as the presenting sign of metastatic leiomyosarcoma, but very few publications have noted pleomorphic sarcoma metastatic to the skin.^{7,8} Moreover, this is the first such case of which we are aware of a high-grade pleomorphic sarcoma arising in the soft tissue and metastasizing to multiple distinct anatomic skin sites. Despite this unusual diagnosis carrying a poor prognosis, the dermatology service was able to facilitate care by quickly refining a definitive diagnosis using a minimally invasive technique.

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