

# A diagnostically challenging spindle cell carcinoma of the neck: Treatment with Mohs micrographic surgery and clinical course



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**Key words:** head and neck; melanoma; metastasis; Mohs micrographic surgery; spindle cell carcinoma.

## INTRODUCTION

Cutaneous spindle cell carcinoma describes a heterogeneous group of tumors that share an atypical spindle cell morphology and infiltrative growth pattern. Examples include dermatofibrosarcoma protuberans, desmoplastic melanoma, malignant fibrous histiocytoma, leiomyosarcoma, malignant peripheral nerve sheath tumor, and spindle cell squamous cell carcinoma.<sup>1</sup> Given their common histopathologic appearance with hematoxylin and eosin stain, immunohistochemistry is typically used to distinguish their cell of origin, although despite this, rare variants may obscure a clear diagnosis.<sup>1</sup> We report a patient with a diagnostically challenging cutaneous spindle cell carcinoma, its treatment with Mohs micrographic surgery, and subsequent aggressive clinical course.

## CASE DESCRIPTION

An immunocompetent 74-year-old woman with a history of basal cell carcinoma presented to her dermatologist with a painful “marble-sized” tumor on the left lateral side of the neck for several weeks. Concerned about hardware contamination from spinal surgery a month before, her orthopedic surgeon cautioned against a biopsy. The tumor was treated twice during a 2-month period with cryotherapy, but it more than tripled in size. Subsequent biopsy revealed a spindle cell tumor proliferation with immunohistochemistry negative for SOX10, AE1/AE3, CK5/6, HMB-45, Melan-A, S-100, and desmin. CD31 and CD34 highlighted background blood vessels only. Smooth muscle actin was focally

positive. The tumor was diagnosed as a “pseudotumor” or “low-grade” spindle cell sarcoma, which prompted a referral for Mohs micrographic surgery. Our working clinical diagnosis was a sarcomatoid dermatofibrosarcoma protuberans or spindle cell squamous cell carcinoma. Given our success with Mohs micrographic surgery as a curative treatment for these tumors, combined with the obscure pathology, we proceeded with Mohs micrographic surgery to remove the 3.5 × 3.0-cm ulcerated mass (Fig 1). There was no palpable lymphadenopathy on examination.

Histologically tumor-free margins were obtained after 2 stages of Mohs micrographic surgery. Fascicles of atypical spindled cells were noted within subcutaneous fat and skeletal muscle on frozen histopathologic sections (Fig 2). The surgical wound measured 5.4 × 4.8 cm and extended around major neck vasculature and nerves, which were carefully dissected (Fig 3). The surgical defect was repaired with a Mercedes advancement flap. Frozen sections obtained during Mohs micrographic surgery were sent for pathologic consultation. In comparison with those in the original biopsy, the spindle cells appeared more cellular and atypical, and were thought to represent a fibrosarcoma or dermatofibrosarcoma protuberans.

Given the history and presentation, histopathology, deep surgical extension adjacent to cervical lymphatic draining basins, and known predisposition of spindle cell tumors for metastasis,<sup>2</sup> additional evaluation and referrals to a medical oncologist and an otolaryngologist were initiated. Chest radiograph

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Funding sources: None.

Conflicts of interest: None disclosed.

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JAAD Case Reports 2020;6:305-7.

2352-5126

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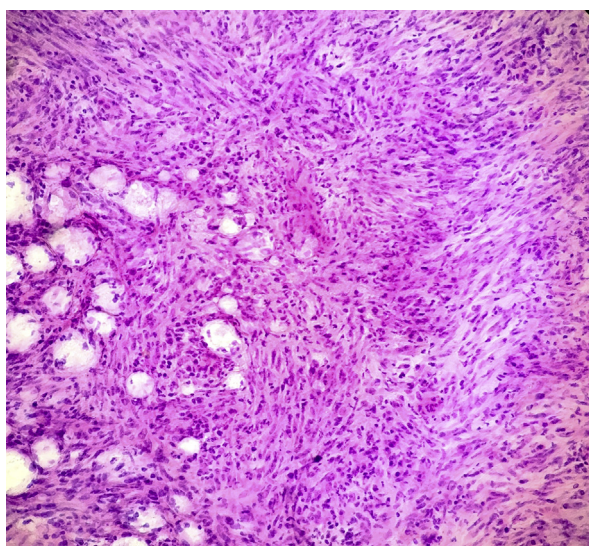
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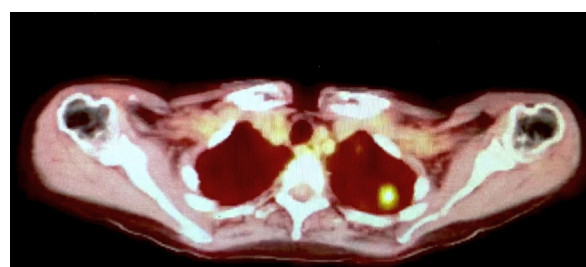
**Fig 1.** A painful, rapidly growing, 3.5 × 3.0-cm tumor that had been present for 3 months on the left lateral side of the neck.



**Fig 3.** Mohs micrographic surgical defect measuring 4.8 × 5.4 cm. A communicating vein, the external jugular, transverse cervical nerve, and posterior border of the sternocleidomastoid muscle are shown.



**Fig 2.** Representative histologic features on frozen sections from the first stage of Mohs micrographic surgery. The cells have enlarged, hyperchromatic, and vesicular chromatin. Mitotic figures are present. Focally, there is a vague storiform pattern and honeycomb permeation of fat. (Hematoxylin-eosin stain; original magnification: ×10.)



**Fig 4.** Fluorine-18 fluorodeoxyglucose positron emission tomography–computed tomography scan after Mohs micrographic surgery, revealing fluorine-18 fluorodeoxyglucose–avid left-sided pulmonary and mediastinal metastases.

revealed 2 nodular densities within the mid right lung and apical left lung, and magnetic resonance imaging revealed enlarged lymph nodes or recurrent tumor on the left side of the neck. A subsequent fluorine-18 fluorodeoxyglucose positron emission tomography–computed tomography scan revealed metastatic disease in the left side of the neck, multiple pulmonary parenchymal lesions, mediastinal lymph node involvement, and multiple subcutaneous chest tumors in the right side of the chest (Fig 4). The patient underwent a radical neck dissection a week later.

Comprehensive genetic studies on residual tumor during the neck dissection revealed numerous mutations, including a *TP53* mutation. The patient

was referred to a medical oncologist, who initiated adjuvant chemotherapy (doxorubicin) and olaratumab (a PDGFR- $\alpha$  antibody). After a good response initially, the patient developed congestive heart failure and began receiving pembrolizumab instead (anti-PD1 antibody), and she currently has no disease progression.

## DISCUSSION

Although immunohistochemical stains are often useful in differentiating cutaneous spindle cell tumors, special stains may be insufficient. The immunostaining studies showed negative staining results for SOX-10, CD34, desmin, intratumoral CD31, AE1/AE3, and CK5/6, which are typical markers in desmoplastic melanoma, dermatofibrosarcoma protuberans, myofibrosarcoma, cutaneous angiosarcoma, and spindle cell squamous cell carcinoma, respectively. Although these results were inconclusive, the tumor's rapid growth and our patient's pain prompted expedient surgical intervention and subsequent metastatic evaluation. Her marked clinical response to pembrolizumab, a checkpoint protein antibody approved for nonresectable metastatic

melanoma, made spindle cell melanoma the leading differential diagnosis.

Despite thorough evaluations, ambiguous diagnoses and diagnostic challenges exist. Reaching a definitive diagnosis before surgery is not always feasible, given the inherent delays in tissue testing and the necessity at times for clinical action. In hindsight, although Mohs micrographic surgery did not provide the intended cure, it debulked a rapidly growing tumor, offered immediate pain relief, secured additional tissue for histopathologic examination, and established a more definitive course of action. This scenario may be encountered during rare Mohs micrographic surgery cases in which the infiltrative growth of squamous cell carcinomas or other spindle cell neoplasms (eg, atypical fibroxanthoma, dermatofibrosarcoma protuberans), and in some unfortunate instances aggressive basal cell carcinomas, cannot be appreciated beforehand.

In most academic or university settings, presentation of diagnostic and management conundrums at multidisciplinary tumor boards is particularly beneficial for collaborative learning and therapeutic guidance. A recent report highlighted the benefits of such a multidisciplinary approach in dermatologic surgery,<sup>3</sup> but it also noted the practical difficulties in gathering

the numerous specialists, which can be particularly challenging for private-practice clinicians. Our patient would have benefitted from tumor board discussion, in which evaluation and treatment could have initially been guided by numerous informed subspecialists. Furthermore, our intention was to minimize morbidity and maximize cost-effectiveness by performing Mohs micrographic surgery, given the information we had at patient presentation. In retrospect, although Mohs micrographic surgery was not curative, it was used to facilitate the patient's subsequent evaluation and treatment.

Our case demonstrates that expedient interdisciplinary care and an oncologic evaluation for extracutaneous involvement for aggressive, diagnostically challenging cutaneous spindle cell carcinomas may be a reasonable approach despite ostensibly clear margins involving Mohs micrographic surgery.

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