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Leiomyosarcoma of the inferior turbinate

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ABSTRACT

We report a case of leiomyosarcoma of the inferior nasal turbinate. The patient, a 68-year-old Caucasian male, presented with 4–6 weeks of epistaxis that was resistant to nasal packing and septal cautery. Upon inspection in the operating room, a small mass was excised from the inferior turbinate. High-power H&E-stained microscopy demonstrated bundles of malignant smooth muscle cells, and immunohistochemical stains were strongly positive for desmin, smooth muscle actin and vimentin, while negative for pankeratin EA1/EA3 and CaM 5.2, suggesting leiomyosarcoma as the diagnosis. Clear margins were obtained at a second surgery. At the time of this writing it is 8 months since his last surgery and he has remained symptom free.

Introduction

Approximately 8.4% of all sarcomas are located in the head and neck. Leiomyosarcoma (LMS) is a malignant tumor of smooth muscle origin frequently found in the uterus and digestive tract. In the head and neck, however, LMS makes up only 2.3% of non-epithelial tumors of the paranasal sinuses, nasopharynx and nose [1]. This is in part due to the small amounts of smooth muscle in the head and neck, largely limited to the tunica media of vasculature [2]. Moreover, it has been shown that LMS isolated to the head and neck are more aggressive and carry a poorer prognosis than gastrointestinal variants. To our knowledge, less than 20 cases of nasal leiomyosarcoma have been reported globally. Here we present a case of this rare entity and discuss appropriate treatment.

Case presentation

The patient, a 68-year-old Caucasian male, was referred to our clinic with recurrent intermittent epistaxis for 4–6 weeks. The epistaxis was intermittently treated with nasal packing and septal cautery but failed to resolve. He denied fatigue, anorexia or weight loss and admitted to chronic rinosinusitis and nasal airway obstruction for most of his adult life with a history of melanoma of the face.

In-office nasal examination revealed a significant septal deviation on the side of the recurrent epistaxis and inferior turbinate hypertrophy. No masses or lesions were appreciated during office endoscopy. The decision was made to proceed with nasal endoscopy, septoplasty and submucosal reduction (SMR) of the inferior turbinates to control epistaxis and improve his nasal obstruction. While performing the SMR of the left inferior turbinate, an irregular mass was encountered on the lateral undersurface of the turbinate, abutting the medial wall of the maxillary sinus. This only became visible after a reduction in the inferior turbinate had been performed. Multiple biopsies of this area were sent as permanent specimens for pathologic review. The remainder of the SMR, septoplasty and endoscopy were unremarkable.

Histopathologic examination demonstrated submucosal involvement by a high-grade sarcoma with areas of necrosis. High-power H&E-stained microscopy demonstrated bundles of malignant smooth muscle cells (Fig. 1). Immunohistochemical (IHC) stains (Figs. 2 and 3) were strongly positive for desmin, smooth muscle actin and vimentin while negative for pankeratin EA1/EA3 and CaM 5.2.

A diagnosis of grade 3 leiomyosarcoma of the left inferior nasal turbinate was made. Subsequent PET and CT scans were negative for distal and local malignancies.

Given the need to assess surgical margins, the patient was taken back to the operating room for left inferior turbinectomy, maxillary antrostomy/partial medial maxillectomy and anterior ethmoidectomy. All margins were negative for residual leiomyosarcoma.

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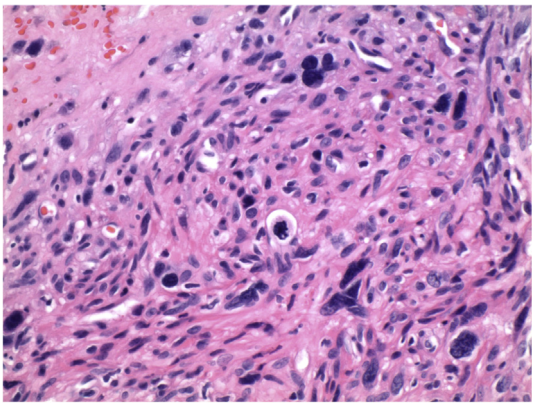

Fig. 1. High-powered (40X) H&E-stained micrograph showing fascicles of malignant spindle cells with cigar-shaped nuclei.

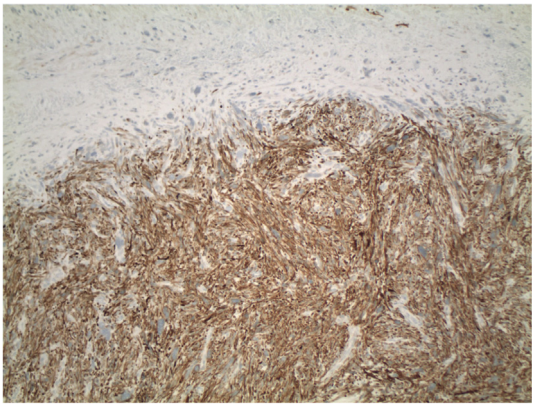

Fig. 2. Positive desmin immunohistochemical stain (10X).

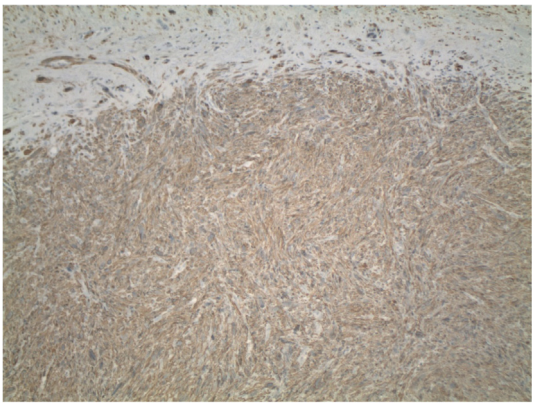

Fig. 3. Positive smooth muscle actin immunohistochemical stain (10X).

His case was presented at our multidisciplinary tumor conference and the decision was made to withhold adjuvant therapy. The patient has been compliant with close follow-up and has had no signs of recurrence as of 8 months after his second operation.

Discussion

Dobben et al. reported the first case of LMS arising from the nasopharynx in 1958, and since that time fewer than 20 cases have been reported worldwide [3].

Across reports thus far, the mean age of onset is 53, with a slight male predominance of 1.1:1. Similar to other sinonasal malignancies, the most common presenting symptoms are congestion, epistaxis, pain, local swelling, and rhinorrhea [4].

The rarity at which LMS presents in the sinonasal cavity is likely attributed to the paucity of smooth muscle in this region. Some suggest that the cells of origin are found in the tunica media of blood vessel walls, while others have suggested the tumor arises from undifferentiated mesenchymal tissue or even myoepithelial cells in submucosal glands [2].

H&E staining of LMS shows intersecting fascicles of malignant spindle cells with cigar-shaped or atypically lobulated nuclei; however, LMS can be difficult to distinguish from other soft tissue sarcomas by light microscopy alone. Differential diagnoses include fibrosarcoma, rhabdomyosarcoma, malignant schwannoma and benign spindle-cell tumors. To differentiate between these various tumors, IHC staining methods are often necessary. As was used in our case, IHC staining with antibodies to smooth muscle actin and desmin as well as vimentin as a marker for mesenchymal tissue are useful in demonstrating LMS as the diagnosis.

Wide surgical excision is the treatment of choice for nasal LMS. As LMS in the sinonasal tract has not been shown to be radiosensitive, radiation therapy of primary, recurrent or metastatic LMS is generally ineffective in reducing tumor size. Chemotherapy for sinonasal leiomyosarcoma has only been used effectively in adjuvant with surgery and may be considered in patients with multiple metastases and/or a large primary tumor [5]. There is a lack of literature regarding management of the neck in patients presenting with nasal LMS; however, as spread tends to be loco regional, there is likely limited benefit from neck dissection or radiation to the neck.

Conclusion

Sinonasal leiomyosarcoma is a rare entity. When encountered, surgical excision should be considered the primary treatment modality, with reservation of chemotherapy for advanced disease.

Conflict of interest

There are no conflicts of interest for the above authors.

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