Glomangiopericytoma of nasal cavity

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ABSTRACT
Glomangiopericytoma/sinonasal type hemangiopericytoma is a rare sinonasal neoplasm arising from the pericytes surrounding capillaries and accounts for less than 0.5% of all sinonasal tumors. This tumor differs from conventional soft tissue hemangiopericytoma in location, biologic behaviour and histologic features. Glomangiopericytoma is a borderline low malignancy tumor with a good prognosis after complete surgical resection. We report a case of 60-year-old woman who presented with progressive nasal obstruction and frequent nasal bleeding and was diagnosed as glomangiopericytoma on histopathological and immunohistochemistry findings. Histological characteristics, differential diagnosis and prognosis of this tumor are discussed in this article. This case has been reported because of its rarity and an array of differential diagnosis.

KEY WORDS: Glomangiopericytoma, nasal cavity, sinonasal type hemangiopericytoma

INTRODUCTION
Glomangiopericytoma is defined as a sinonasal tumor with perivascular myoid phenotype, which was first reported as hemangiopericytoma in 1942 by Stout and Murray[1] as a soft tissue tumor with characteristic vascular proliferation including branching vessels and small vessel perivascular hyalinization. The World Health Organization (WHO) classified this tumor as glomangiopericytoma in 2005.[2] It is rare and accounts for <0.5% of all sinonasal neoplasms.[3] The etiology is not clear although past trauma, hypertension, pregnancy and use of corticosteroids may be involved.[3] We present a case of glomangiopericytoma arising from the septum of the right nasal cavity, which was treated by transnasal endoscopic surgery.

CASE REPORT
A 60-year-old female presented to ENT Department Surat Municipal Institute of Medical Education and Research Hospital with 3 months history of progressive right nasal blockage, nasal bleeding and difficulty in breathing and was clinically diagnosed as a nasal polyp. Rhinoscopy was done which revealed a friable grayish pink polypoidal mass, that bleed on manipulation which was occupying right nasal cavity.

Computed tomography (CT) scan paranasal sinus view showed large ill-defined heterogeneous enhancing lesion involving the right nasal cavity widely infiltrative (right ethmoid air cells, right half of nasopharynx, right maxillary ostium, right nasal turbinates and probably right half of uncinate process and right half of sphenoid bone) in surrounding spaces; with lytic-sclerotic changes [Figure 1]. Diagnosis of vascular lesion-hemangioma/hemangiopericytoma/other neoplastic – inflammatory pathology were given on CT scan.

The biopsy was taken and sent to Histopathology Department. Microscopic examination showed squamous epithelial lining; subepithelial tissue showed the lobular arrangement of spindle-shaped cells admixed with vascular channels of variable size with areas of hemorrhage and secondary inflammatory infiltrate. The differential diagnosis of hemangiopericytoma and solitary fibrous tumor (SFT) was made, and immunohistochemistry (IHC) advised. After a week, excision biopsy specimen was received. Received tissue was in multiple pieces, grey-brown in color and collectively measured 5 cm × 5 cm × 1.5 cm. Histopathological examination of sections
The majority of patients present with regional lymph node incidence in the seventh decade of life and a slight female predominance. The majority of patients present with unilateral nasal obstruction and/or recurrent epistaxis. Clinically, glomangiopericytoma is polyloid, beefy red to grayish pink, soft, fleshy to friable and edematous to hemorrhagic in appearance which bleeds easily on touching. Regional lymph node involvement is rare. The etiology is still unclear, although past trauma, hypertension, pregnancy and use of corticosteroids may be the causative factors as described by Angouridakis et al.

Microscopically this tumor consistently displays myoid appearance without cellular pleomorphism. Hematoxylin and Eosin staining shows a subepithelial well-delineated but unencapsulated cellular tumor, surrounded by the normal respiratory epithelium, characterized by diffuse growth of closely packed cells, forming short fascicles and sometimes exhibiting storiform, whorled or palisaded pattern, interspersed with numerous thin-walled and branching staghorn vessels. The neoplastic cells are uniform and elongated to oval, with a round to spindle-shaped nuclei and lightly eosinophilic cytoplasm. Compared to soft tissue hemangiopericytoma; limited mitotic activity, atypia and less amount of hemorrhage and necrosis are seen in glomangiopericytoma. Also cells are usually arranged in certain patterns such as sheets, short fascicles, and long rows. Immunohistochemically, glomangiopericytoma can be distinguished from soft tissue hemangiopericytoma by the characteristic diffuse reactivity for actin, factor XIIIa and vimentin without strong diffuse staining for CD34.

Differential diagnosis of glomangiopericytomas are with lobular capillary hemangioma (LCH), SFT, leiomyoma, and angiofibroma; than from sarcomas, which are usually clearly malignant. In contrast to glomangiopericytomas, LCH has a distinct lobular architecture often with an identifiable large, central feeder vessel.
CONCLUSION

Glomangiopericytoma is an uncommon, indolent, unique sinonasal neoplasm of older adults with a perivascular myoid phenotype that differs from traditional soft tissue “hemangiopericytoma” in location, biologic behavior and morphology. Characteristic histologic features include a diffuse, subepithelial, cellular proliferation of bland, spindled cells and a distinctive vascular network composed of variably sized vascular channels, some with perivascular hyalinization. Entities such as LCH, SFT, sinonasal leiomyoma, and angiofibroma are commonly confused with this lesion. Complete transnasal endoscopic excision is the choice of treatment. The regular postoperative follow-up is recommended for early finding of tumor recurrence.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES
