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Glomangiopericytoma of Nasal Cavity: A Rare Sinonasal Perivascular Tumor

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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 behavior, and histologic features. Glomangiopericytoma is a borderline low malignancy tumor with a good prognosis after complete surgical resection. A 42-year-old man presented with progressive nasal obstruction and frequent nasal bleeding. Rhinoscopic examination and imaging studies showed a mass occupying the right nasal cavity without bone destruction. The tumor was removed successfully by transnasal endoscopic excision. The pathological examination with immunohistochemistry confirmed the diagnosis of glomangiopericytoma. Histological characteristics, therapeutic principles and prognosis of this tumor are discussed in this article.

Key words: glomangiopericytoma, hemangiopericytoma, sinonasal type hemangiopericytoma, nasal cavity, benign tumor, histology

INTRODUCTION

Glomangiopericytoma is defined as a sinonasal tumor with perivascular myoid phenotype, which accounts for less than 0.5% of all sinonasal tumors. Glomangiopericytoma 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. This classification includes the tumor described as sinonasal type hemangiopericytoma, and hemangiopericytoma-like tumor.[2] 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 septum of right nasal cavity, which was treated by transnasal endoscopic surgery.

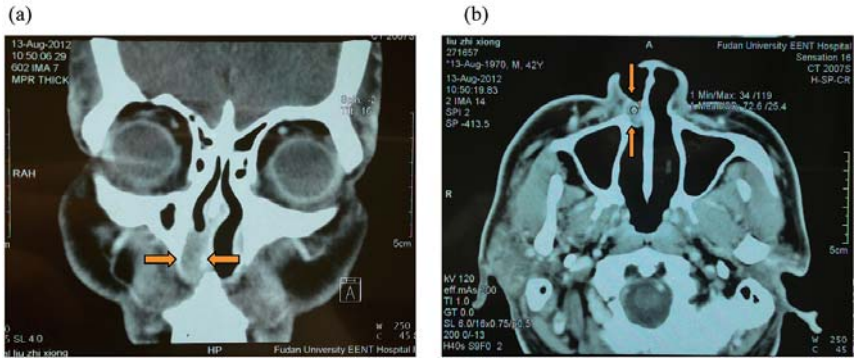

Fig. 1 Coronal (a) and axial (b) computed tomography scans showing soft tissue density (arrows) in the anterior part of the right nasal cavity.

CASE REPORT

A 42-year-old man presented with an one-month history of progressive right nasal obstruction and frequent epistaxis .He had a history of hepatitis B. Rhinoscopy revealed a friable grayish pink polypoid mass, that bled easily with minimal manipulation occupying the right anterior nasal cavity. The skin of the external nose was free of the tumor. There was no enlargement of the cervical lymph nodes. Computed tomography showed soft tissue density about 2 cm×1.8 cm in the anterior part of the right nasal cavity without bony destruction or invasion to the surrounding tissue (Fig.1a,1b). The patient claimed that the tumor size increased gradually. The tumor bled very easily, so biopsy was not performed. With these clinical characteristics and radiographic findings, we sup-

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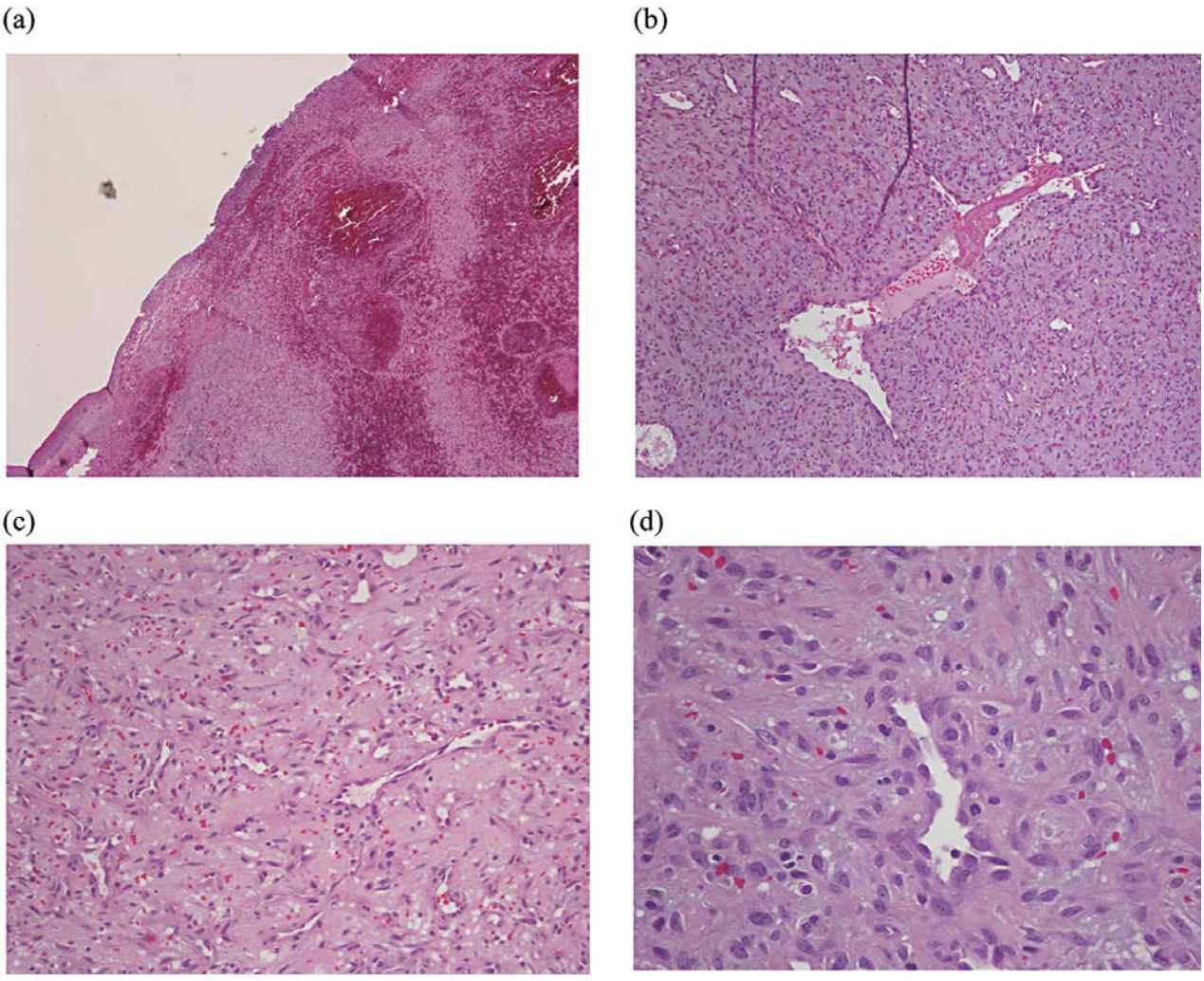

Fig. 2 Photomicrographs of the tumor specimen. (a) Tumoral proliferation of spindle-shaped cells arranged in fascicles and covered by an intact respiratory epithelium (hematoxylin and eosin stain, x40). (b) Staghorn vessels with spindle cell proliferation. Presence of perivascular hyalinization (hematoxylin and eosin stain, x100) (c) Aggregation of thin-walled vessels separated by edematous stroma with spindle cell proliferation (hematoxylin and eosin stain, x200) (d) Uniform, elongated to oval tumor cells, with oval to spindle-shaped nuclei with mild nuclear pleomorphism and occasional mitosis (hematoxylin and eosin stain, ×400).

posed the tumor to be benign hypervascular tumor, such as hemangioma, although the possibility of metastatic cancer was not excluded. We considered that the tumor was resectable because of the small size, limited expansion and the easily accessible location of the tumor.

Endoscopic surgery was performed. The tumor was pedunculated with a large stem at the anterior part of the nasal septum. The stem was cut and the tumor was removed completely. During operation, the bleeding was insignificant and controlled without difficulty by compression with pledgets soaked in diluted epinephrine.

Histological examination revealed tumoral proliferation of spindle-shaped cells arranged in fascicles and covered by an intact respiratory epithelium. Numerous staghorn vessels were found and surrounded by spindle-shaped cells. Presence of perivascular hyalinization was noted (Fig.2a,2b). Aggregation of thin-walled vessels was separated by edematous stroma with spindle cell proliferation (Fig 2c). Tumor cells was uniform, elongated to oval, with oval to spindle-shaped nuclei with mild nuclear pleomorphism and occasional mitosis (Fig.2d). Necrosis and cytological atypia were absent. Immunohistochemical staining confirmed endothelial cells stained with the antibodies to actin, CD34, and D2-

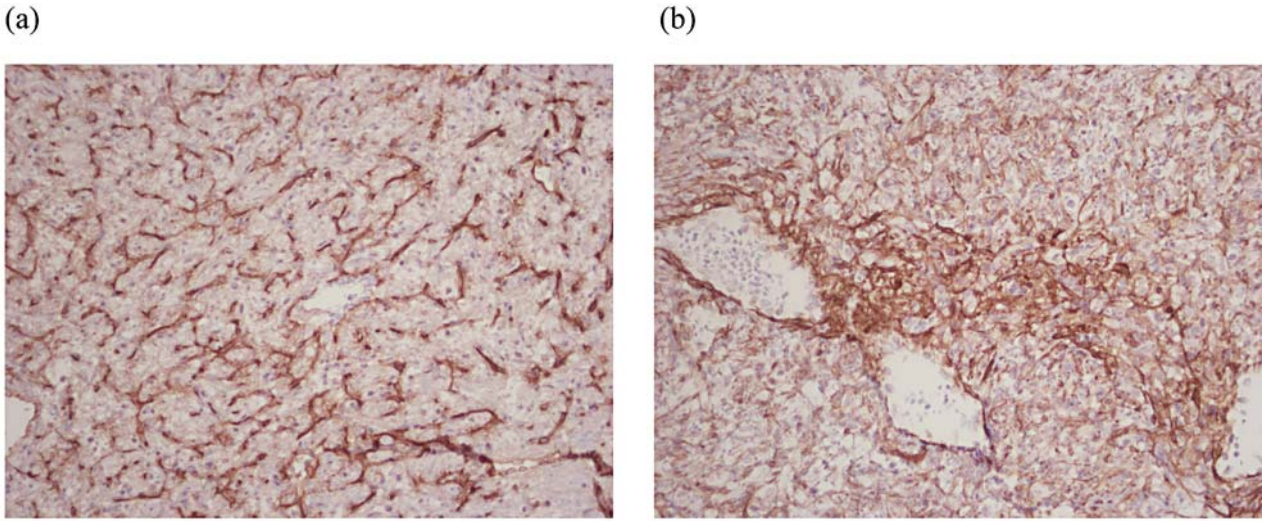

Fig. 3 Immunohistochemical staining showing(a) endothelial cells react to CD34 (×200). (b)tumor cells are strongly positive to muscle specific actin (×200).

40. The tumor cells were strongly positive to actin, and negative to CD34 and D2-40. And the blood vessels wall was positive to CD34 (Fig.3a,3b). These histopathologic findings led to a diagnosis of glomangiopericytoma.

DISCUSSION

Glomangiopericytoma is also called sinonasal hemangiopericytoma. It is categorized to borderline and low-malignant-potential soft tissue tumors of the nose and paranasal sinuses and was defined by the World Health Organization in 2005 as a sinonasal tumor demonstrating a perivascular myoid phenotype.[2] The concept of hemangiopericytoma was first described in 1942 by Stout and Murray[1] as a soft tissue tumor with the characteristic of vascular proliferation including branching vessels and small vessel perivascular hyalinization with otherwise marked morphologic heterogeneity. And this tumor arising from the sinonasal tract was first described by Compagno in 1976 as "hemangiopericytoma-like".[4]

Glomangiopericytoma was previously known as hemangiopericytoma, which was defined as the tumor arising from pericytes which can occur in any location with capillaries. This lesion was thought to fall in the spectrum between glomus tumors and capillary hemangiomas and hence the term hemangiopericytoma was chosen.[5] However, the clinical and histologic characteristics of sinonasal hemangiopericytoma differ from those of hemangiopericytoma developing elsewhere in the body. Therefore, glomangiopericytoma, also called sinonasal hemangiopericytoma, is considered as a distinct entity.[6, 7]

Glomangiopericytoma is a rare tumor of nasal cavity while it accounts for less than 0.5% of all sinonasal neoplasms.[2,6] People in a broad range of age may be affected, with a peak incidence in the seventh decade of life and a slight female predominance.[8,9] The majority of patients presents with unilateral nasal obstruction and/or recurrent epistaxis while the complaints of difficulty in breathing, visual disturbance, pain and headache are less frequent.[6,10] Clinically, glomangiopericytoma is polypoid, beefy red to grayish pink, soft, fleshy to friable, and edematous to hemorrhagic in appearance with easily bleeding on touching.[2,9] Regional lymph node involvement is rare.[11]

The etiology is still unclear, although past trauma, hypertension, pregnancy and use of corticosteroids may be the causes of the tumor which was described earlier by Angouridakis N *et al*.[3] However, the patient in this study doesn't have the past histories of trauma, hypertension and steroid use.

Histological examination is important for the diagnosis. Hematoxylin - eosin staining shows a subepithelial well-delineated but unencapsulated cellular tumor, surrounded by the normal respiratory epithelium[8], 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 round to spindle-shaped nuclei and lightly eosinophilic

cytoplasm.[2,3,5,6,8,10] Compared to soft-tissue hemangiopericytoma, limited mitotic activity and atypia, and less hemorrhage and necrosis are seen in glomangiopericytoma.[6,7] Immunohistochemically, glomangiopericytoma can also be distinguished from soft-tissue hemangiopericytoma by the characteristics of diffuse reactivity for actins, factor XIIIA and vimentin without strong diffuse staining for CD34.[2,6,12]

Glomangiopericytomas may be confused with a variety of spindle cells and vascular neoplasms occurring in the sinonasal tract. It is relatively more difficult to distinguish glomangiopericytomas from other benign/borderline vascular-rich spindle cell lesions, most commonly including lobular capillary hemangioma (pyogenic granuloma), solitary fibrous tumor, leiomyoma, and angiofibroma than from sarcomas, which are usually clearly malignant.[5]

Lobular capillary hemangiomas (LCHs) are often confused with glomangiopericytomas, while both are cellular tumors with a prominent vascular components. In contrast to glomangiopericytomas, LCHs have a distinct lobular architecture and frequently contain staghorn-type vessels in the interlobular stroma. The tumor cells lining slitlike vascular spaces have more irregular nuclear contours and mitoses. Immunohistochemically, the neoplastic cells of LCHs show vascular endothelial differentiation with CD31 and CD34 positive stains.[13]

Similar to glomangiopericytomas, solitary fibrous tumors (SFTs) consist of a cellular spindle cell proliferation effacing submucosal structures with staghorn-type vessels. In contrast to glomangiopericytomas, SFTs have a prominent collagenous stroma with spindle cells intimately associated with ropy collagen bundles. The tumor cells show more nuclear overlap and contour abnormalities. Immunohistochemically, the neoplastic cells of SFTs stain positively for CD34, with less consistent actin staining.[14]

Sinonasal leiomyomas with staghorn-type and capillary-sized vessels associated with spindle cell proliferation mimic glomangiopericytomas. Prominent fascicular growth is the characteristic of leiomyoma and the tumor cells have fibrillary eosinophilic cytoplasm. In comparison to glomangiopericytomas, the nuclei are more elongated with coarser chromatin and occasional perinuclear vacuoles. Immunohistochemically, Desmin staining is typically strongly positive in these tumor cells, which helps to separate them from glomangiopericytomas.[15]

Angiofibromas have abundant stromal collagen, making them eosinophilic at low-power magnification, and have a prominent vascular stroma. The tumor cells are usually stellate shaped and evenly spaced within the collagen-rich stroma.[16] In contrast, glomangiopericytomas contain spindle cells, lack of collagenous stroma, and are much more cellular. Immunohistochemically, the nuclei of angiofibroma cells are positively stained for androgen receptor[17] and β-catenin[18], which is helpful to distinguish them from glomangiopericytomas.

Glomangiopericytoma is categorized as a borderline low malignancy tumor by the WHO classification in 2005, with an outstanding overall survival rate which can be achieved by complete excision.[2,6,19] The local recurrence rate is reported as 16.8%, but the recurrences may be a consequence of incomplete excision and considered as a residual disease.[6,9] In our present case, the tumor lesion was completely resected by transnasal endoscopic excision which leads to low potential of recurrence; even so, the patient is still highly recommended to be regularly followed up with systemic examination.

In conclusion, glomangiopericytoma is an uncommon, indolent, unique sinonasal neoplasm with a perivascular myoid phenotype that differs from traditional soft tissue "hemangiopericytoma" in location, biologic behavior, and morphology. Glomangiopericytoma is categorized as a borderline low malignancy tumor. Complete transnasal endoscopic excision is the choice of treatment. Regular post-operative follow-up is recommended for early finding of tumor recurrence.

DISCLOSURE

All authors declare that this study has no conflict of interest.

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