OPEN ACCESS – CASE REPORT

Glomangiopericytoma: A Rare Sinonasal Neoplasm

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ABSTRACT

Glomangiopericytoma is a rare neoplasm of the nasal and paranasal sinuses comprising less than 1% of all tumors of the said region. We report of a 59-year-old hypertensive male who presented with epistaxis. CT scan findings showed a mass in the right nasal cavity with extension into the ethmoid and sphenoid sinuses. Histopathologic diagnosis was glomangiopericytoma confirmed with immunohistochemistry studies. Prognosis is favorable with complete resection of tumor and long-term monitoring.

Key words: glomangiopericytoma, paranasal sinus neoplasms, intranasal neoplasms, sinonasal hemangiopericytoma-like tumor

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INTRODUCTION

Glomangiopericytoma is a rare vascular tumor of the nasal cavity and paranasal sinuses. It comprises less than 0.5% of all sinonasal neoplasms with a characteristic and prominent perivascular growth pattern.[1] Gross appearance is similar to the more common nasal inflammatory polyps. Immunomorphologic features will differentiate this tumor from other intranasal neoplasms and soft tissue hemangiopericytomas arising from other sites.[2]

CASE

We report a case of a 59-year-old male, known hypertensive and diabetic, who had a history of on and off epistaxis several months prior to admission. Successive epistaxis led to a consult with an otorhinolaryngologist who noted a right intranasal mass. CT scan of the paranasal sinuses revealed a polypoid soft tissue mass in the right posterior nasal cavity extending into the posterior ethmoid sinus superiorly and into the right side of the sphenoid sinus posteriorly. The mass measured 2.6 x 2.5 x 1.5 cm and exhibited fairly homogenous contrast enhancement. There was no lytic nor sclerotic changes observed in the adjacent osseous structures. The right superior and middle turbinates were obscured by the said mass (Figure 1). An uneventful nasal endoscopy was eventually done to control the bleeding. Resection of the tumor was done several weeks later with no reported complications.

Histopathology of the mass reveals a cellular tumor composed of variedly sized blood vessels, some showing branching, staghorn appearance. The overlying respiratory epithelium was unremarkable and uninvolved. The tumor cells were composed of spindle shaped cells with ovoid nucleus, eosinophilic cytoplasm and inconspicuous nucleoli (Figure 2). Immunohistochemistry studies revealed diffuse positivity with smooth muscle actin and negative staining with CD45, pan-cytokeratin, CD31, CD34 and S-100 (Figure 3). Based on the abovementioned features, this case was signed out as glomangiopericytoma.

DISCUSSION

Glomangiopericytoma is a rare mesenchymal tumor arising almost exclusively from the nasal cavity or paranasal sinuses. It comprises less than 0.5% of all sinonasal tumors.[1-3] There is a slight female preponderance with a female to male ratio of 3:1. Most patients experience nasal obstruction and epistaxis,[3] similar to that experienced by our patient. Etiology is still not clear; however, predisposing factors include trauma, corticosteroid use, hypertension and pregnancy. Our patient denied previous trauma and corticosteroid use but has a history of hypertension. Hemangiopericytomas were first described by Stout and Murray in 1942 as a soft tissue tumor with a distinct branching proliferation of vascular channels and perivascular hyalinization of small blood vessels.[4] Over the years, the concept of hemangiopericytomas evolved as these tumors were noted to occur in the nasal and paranasal sinuses in 5% of cases. These nasal and sinonasal hemangiopericytomas also behaved indolently compared to its soft tissue counterparts, had distinct morphologic features and were noted to show true pericytic differentiation. As such, in 1976, Compangno and Hyam termed these lesions as hemangiopericytoma-like intranasal tumors.[4,5] In 2005, the World Health Organization (WHO) proposed the term glomangiopericytoma to reflect the findings of several studies that show this tumor's similarity and close relationship with glomus tumors.[5] Glomangiopericytomas are indolent tumors with overall excellent survival (>90% 5-year survival) when complete excision is achieved. Recurrences are usually a result of inadequate resection. The lesions appear fleshy pink to red, hemorrhagic and polypoid on gross examination. Histologically, the characteristic appearance is that of cells arranged in various architecture, separated by vascular channels in staghorn or "antler-like" configuration. Mitosis is rare, necrosis and hemorrhage are uncommon and nuclear pleomorphism is mild to absent.[4,5] A malignant glomangiopericytoma should be suspected in the presence of large tumor size (>5 cm), bony invasion, pleomorphic nuclei, increased mitotic activity (>4/10 high power fields) and increased proliferation activity (>10% Ki-67 proliferation index).[6] Other benign spindle cell neoplasms may be confused with glomangiopericytomas and some close differential diagnosis include: angiofibroma, vascular leiomyoma, lobular capillary hemangioma and solitary fibrous tumors. Angiofibromas are seen almost exclusively in adolescent males. Histologically, they present with large, ectatic vessels, abundant stromal collagen and bland stellate shaped cells. This tumor will stain positive with androgen receptor 22 and β-catenin. Sinonasal leiomyomas are uncommon and can present with staghorn-type vascularity with ovoid nuclei, and prominent fascicular growth pattern. Strong positivity with desmin can help differentiate this tumor from glomangiopericytoma. Lobular capillary hemangioma can also present grossly as an intranasal mass. This tumor is also vascular with a central feeder vessel and gives rise to smaller, slit-like vascular channels. Immunohistochemistry studies show positivity with CD31 and CD34, highlighting the tumor cells' endothelial differentiation. Sinonasal solitary fibrous tumors are also uncommon tumors with variable vascularity and haphazardly arranged cells. Absence of perivascular hyalinization and presence of coarse collagen bundles distinguish this from glomangiopericytoma. The tumor cells of solitary fibrous

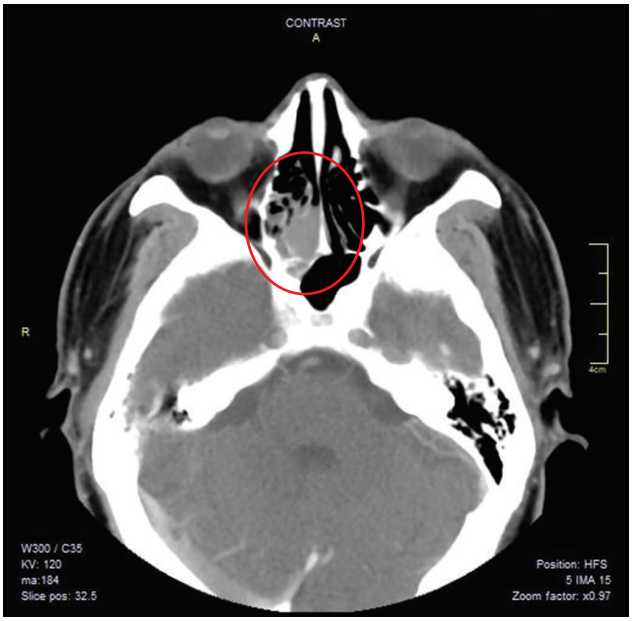

Figure 1. CT scan showed a 2.6 x 2.5 x 1.5 cm polypoid soft tissue mass in the right posterior nasal cavity extending into the posterior ethmoid sinus and into the right side of the sphenoid sinus.

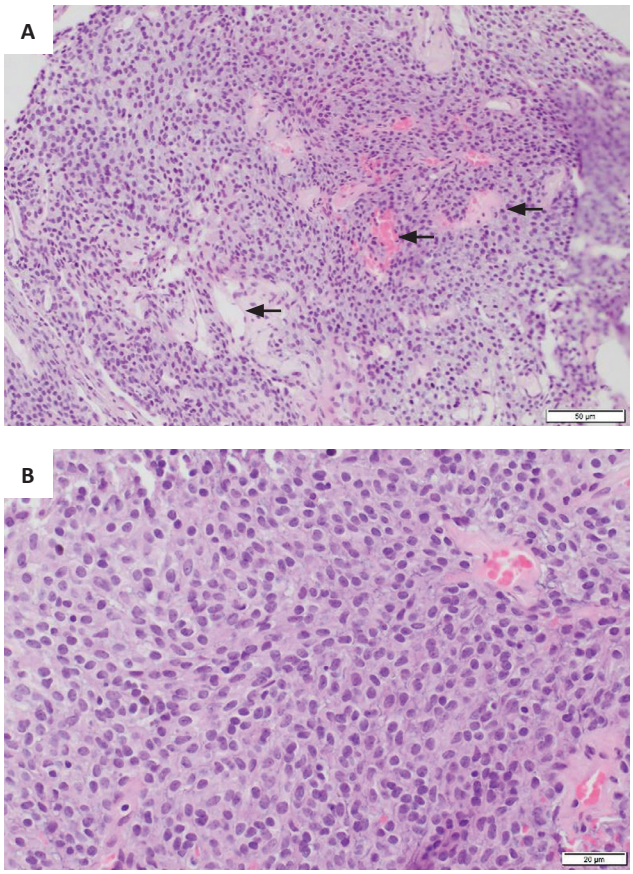

Figure 2. (A) Cellular tumor composed of variedly sized blood vessels (H&E, 20x). **(B)** The tumor cells composed of spindle shaped cells with ovoid nucleus, eosinophilic cytoplasm and inconspicuous nucleoli (H&E, 40x).

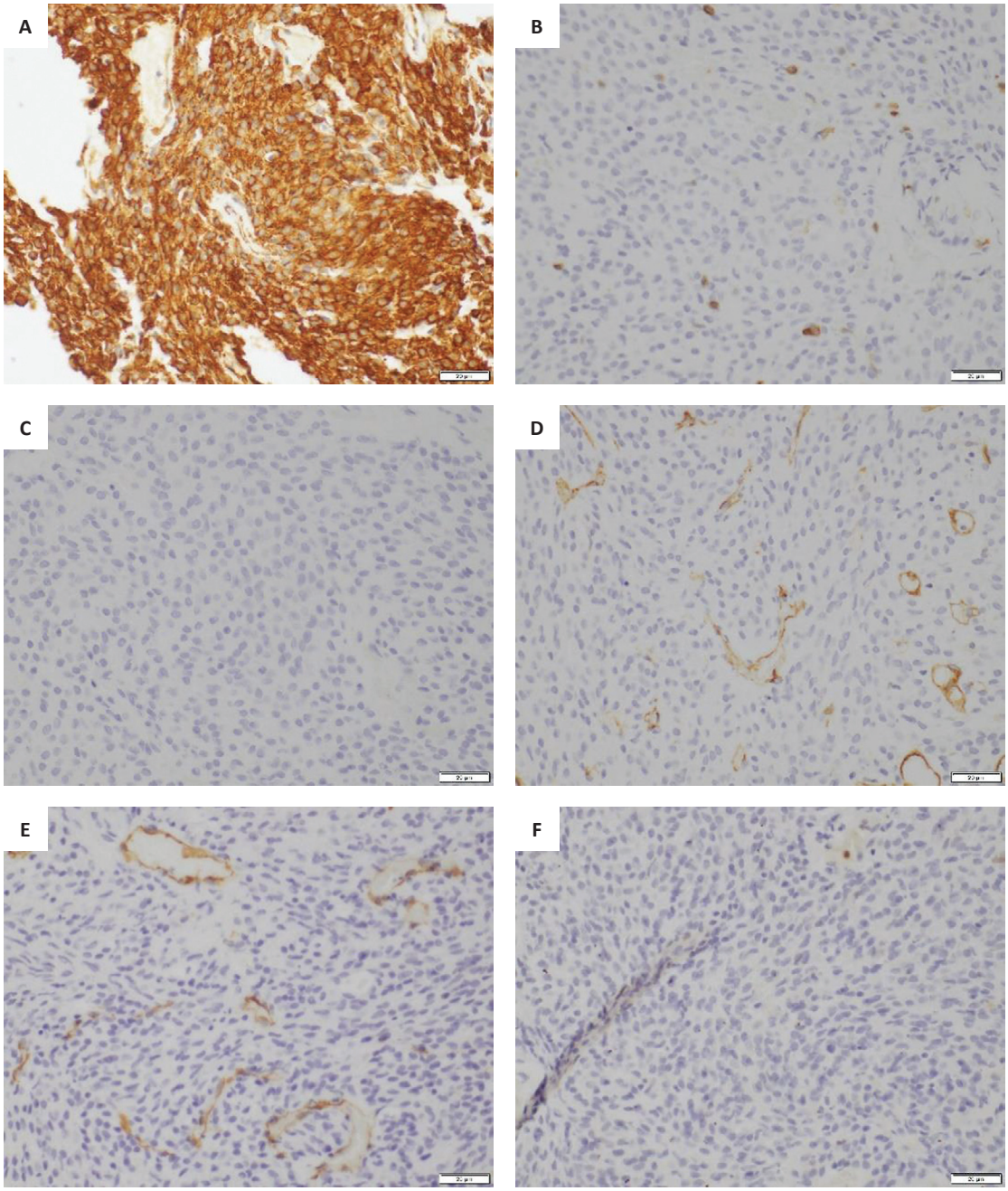

Figure 1. Immunohistochemistry profile of the tumor. **(A)** Smooth muscle actin positive tumor cells (HRP, 40x); **(B)** CD45 negative (HRP, 40x); **(C)** Pan-cytokeratin negative (HRP, 40x); **(D)** CD31 negative (HRP, 40x); **(E)** CD34 negative (HRP, 40x); **(F)** S-100 negative (HRP, 40x).

tumor stain positively with CD34 and variably with smooth muscle actin.[4,6] Immunohistochemistry studies can aid in the differentiation of glomangiopericytoma from the abovementioned benign tumors. Glomangiopericytomas show diffuse reactivity with smooth muscle actin and vimentin and negativity with CD45, CD31, desmin, S-100, STAT-6 and NSE. The tumorigenesis and molecular genetics of glomangiopericytoma are not well-established, however, studies have shown that mutation activation of β-catenin with the associated cyclin D1 overexpression are central events in the pathogenesis of glomangiopericytoma.[7,8] β-catenin is a cadherin-associated membrane protein that is involved in the regulation of cell-to-cell adhesion, a terminal component of the Wnt-signaling pathway. Accumulation of β-catenin results in nuclear translocation, with the nuclear expression of β-catenin demonstrated to up-regulate cyclin D1, leading to its oncogenic activation.[8] Several studies have shown the utility of β-catenin and cyclin D1 in the diagnosis of glomangiopericytoma. Most studies are in agreement that show strong nuclear expression with β-catenin in virtually all glomangiopericytoma.[8-10] Similarly, cyclin D1 was also noted to exhibit nuclear positivity in most (70-100%) of the tumor cells.[8,9] Complete surgical resection is the standard treatment in glomangiopericytoma with radiotherapy reserved only for cases that are inoperable or metastatic.[11] Lymphatic and hematogenous spread of malignant glomangiopericytoma have been reported in 5% of cases and were seen to involve distant organs such

as the lungs, liver and bone.[5] Our patient eventually underwent complete resection with no untoward post-surgery events reported. However, at present, he is already lost to follow up. The patient could have benefitted from a regular follow-up since recurrence may occur even with a long disease-free interval.[5] Recurrence has been reported in 15.1% of cases and is most commonly due to inadequate resection. The WHO in the 2017 Classification of Tumors, describes this tumor as an indolent tumor with the prognosis being favorable with an excellent survival rate.[6,12]

CONCLUSION

Glomangiopericytoma is a rare, indolent neoplasm of the sinonasal region. Surgery remains the treatment of choice and is curative when completely resected. Reporting of cases will help in increasing knowledge, aid in establishment of diagnosis and create definitive guidelines for the treatment for glomangiopericytoma.

ETHICAL CONSIDERATION

The authors have tried to reach out to the patient through his cell phone number and email address, but they have not received any response from him. They also tried to contact him through his attending physician, but the patient has already been lost to follow up. They have also alerted the hospital's radiology and laboratory departments in case the patient goes for a subsequent examination but up until the present, the patient has not returned to the hospital. The authors have exercised due diligence in trying to reach the patient for his consent, but they have not been successful in doing so.

STATEMENT OF AUTHORSHIP

Both authors certified fulfillment of ICMJE authorship criteria.

AUTHOR DISCLOSURE

Both authors declared no conflict of interest.

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