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ORIGINAL ARTICLE

Hemangiopericytoma: A rare sinonasal tumor

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KEYWORDS

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Abstract *Introduction:* Hemangiopericytomas are rare, vascular neoplasms that are derived from Zimmerman's capillary pericytes. They are found most commonly in the retroperitoneum/pelvis and lower extremities and are known to have malignant biological behavior. Of the paranasal sinuses, ethmoid and sphenoid sinuses are most commonly involved.

Case Report: We report an uncommon case of a 77-year-old man, who presented with history of nasal block, nasal discharge and epistaxis since 10 years. On examination, there was a fleshy polypoidal mass seen filling the entire right nasal cavity. CT scan of the osteomeatal complex revealed a soft tissue density lesion occupying the entire right nasal cavity, projecting through the posterior choanae into the nasopharynx. An endoscopic excision of the entire mass was done. Histopathological examination and immunohistochemistry confirmed the diagnosis of Hemangiopericytoma.

Discussion: Hemangiopericytoma is a rare vascular tumor. These lesions are more frequently reported in the seventh decade of life, and the usual presentation includes epistaxis and nasal obstruction. While corticosteroid use, hypertension and pregnancy have been proposed as etiological factors in the development of HPC, this is not widely accepted. It exhibits low malignant potential and distant metastasis is exceedingly rare.

Conclusion: Hemangiopericytomas of the nasal cavity and paranasal sinuses exhibit low malignant potential and distant metastasis. The treatment of choice is wide surgical resection which is the only curative modality.

1. Introduction

Hemangiopericytomas (HPC) are rare, vascular neoplasms, derived from Zimmerman's capillary pericytes, which surround all capillaries, and which account for 1% of all vascular tumors.[1] Initially described by Stout and Murray, they are found most commonly in retroperitoneum/pelvis and lower extremities and are known to have malignant behavior. 7.5–25% of all HPCs are found in the head and neck region, with a marked tendency to occur in nasal cavity and paranasal

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sinuses.[1,2] Of the paranasal sinuses, ethmoid and sphenoid sinuses are most commonly involved. HPCs at these sites do not have the same risk of malignancy and metastasis as HPCs elsewhere.

Due to variation in histological and clinical features of sinonasal type hemangiopericytomas, compared with their soft tissue counterparts, it is now widely accepted that these tumors represent an independent clinical entity, referred to as glomangiopericytoma, due to its similarity to glomus tumors.[3]

Effective management requires wide surgical excision with clear resection margins, as these tumors are relatively radioresistant. Recent advances in endoscopic surgery have led to the development of techniques that now permits endoscopic resection.

2. Case report

A 77 year old man presented to our service with history of nasal block and nasal discharge since 10 years. He also had occasional episodes of epistaxis and headache. He was a known case of diabetes and hypertension on long term aspirin treatment.

On examination, there was a fleshy polypoidal mass seen filling the entire right nasal cavity, displacing the septum to the left side. There was a minimal bleeding on touch. Postnasal examination revealed the mass extending into the nasopharynx through the right choanae.

A CT scan of the osteomeatal complex was performed, which revealed a soft tissue density lesion occupying the entire right nasal cavity, projecting through the posterior choanae into the nasopharynx, deviating the septum to the left and medial wall of the maxillary sinus to the right. The lesion was superiorly displacing the ethmoid sinus laterally and extending to the cribriform plate (Fig. 1).

The patient was posted for surgery and an endoscopic excision of the lesion was done. The lesion was initially mobilized

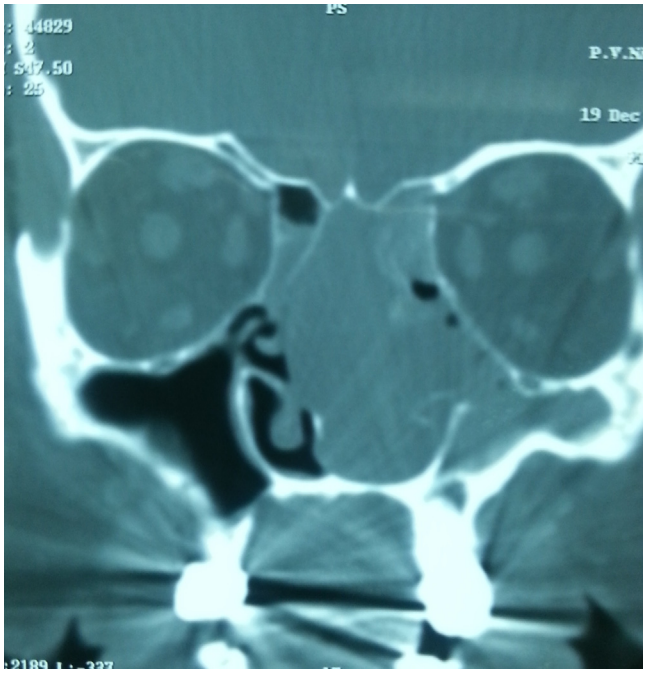

Figure 1 CT scan showing mass filling the entire right nasal cavity and displacing the septum and medial wall of maxillary sinus.

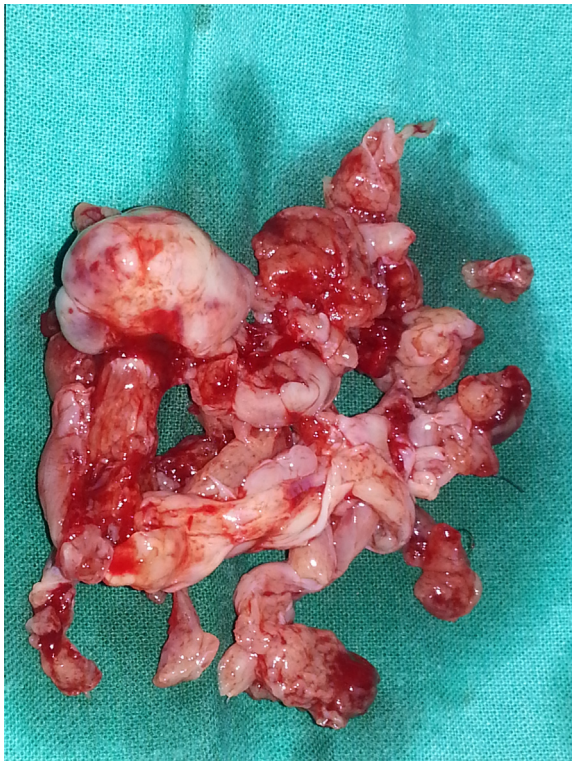

Figure 2 Post operative specimen removed from the right nasal cavity.

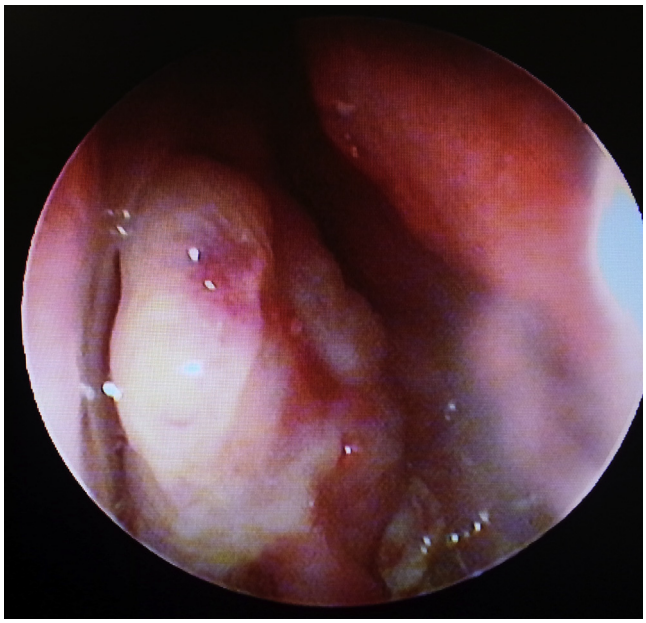

Figure 3 Follow Up sinoscopy showing nasal cavity to be free of growth.

from the nasopharynx into the nasal cavity and debulking was done. Following this sinoscope was passed. The lesion was seen to be arising from the sphenoethmoidal region. Endoscopic excision of the mass was carried out. The right maxillary sinus ostium was widened and sinus was filled with purulent discharge. Mass was completely removed from the ethmoidal and sphenoid region (Figs. 2 and 3).

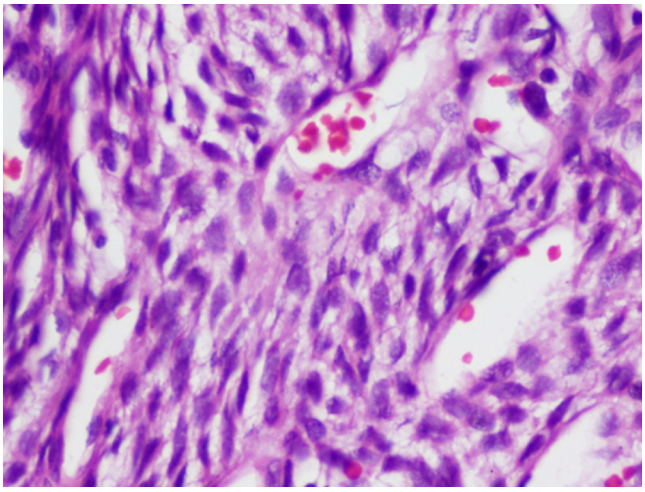

Figure 4 Spindle shaped tumor cells are seen in sheets and whorls around vascular channel. (H and E-45×).

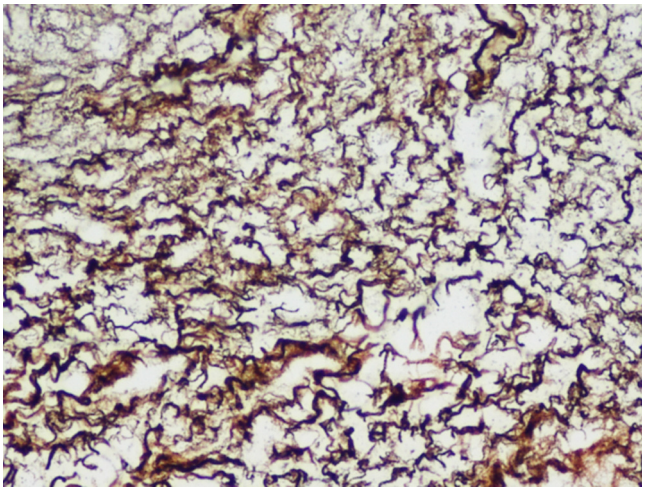

Figure 5 IHC–smooth muscle actin is strongly positive in tumor cells.

Histopathological examination of the biopsy specimen revealed tumor cells seen arranged in interlacing fascicles separated by dilated and irregularly shaped branching blood vessels. Cells had oval vesicular to elongated hyperchromatic nuclei with indistinct cytoplasmic borders. Mitosis was <2/10 HPF. Reticulin stain showed tumor cells proliferating around the blood vessels. IHC markers were done for confirmation of the diagnosis. SMA (smooth muscle actin) was strongly positive, while CD34 was focally positive. S100 and CD99 were negative. Diagnosis of Hemangiopericytoma was confirmed (Figs. 4 and 5).

3. Discussion

Hemangiopericytoma is a rare vascular tumor occurring anywhere in the body, but more commonly involves the soft tissues of trunk and lower extremities. Approximately 15% of all soft tissue HPCs occur in the head and neck region, mainly in the nasal cavity and paranasal sinuses. These lesions are more frequently reported in the seventh decade of life, and the usual presentation includes epistaxis and nasal obstruction.

While corticosteroid use, hypertension and pregnancy have been proposed as etiological factors in the development of HPC, this is not widely accepted.[4]

Diagnosis depends firstly on thorough endoscopic evaluation and secondly on imaging techniques including CT and MRI. The characteristic findings on CT include a soft tissue mass with enhancement following the administration of intravenous contrast, and gadolinium contrast in case of MRI scanning.[5] The role of routine angiography in the management of HPC is not clearly defined. However its use is advocated for pre-operative planning and embolization, which in turn significantly reduces the risk of intraoperative hemorrhage.[6,7]

From a histological viewpoint, an accepted definition and classification of HPC do not exist. Several authors have addressed the histologic grading of HPC as benign, borderline, low grade malignant and overtly malignant.[8] The histological features of HPC consist of uniform, spindle-shaped cells with indistinct cytoplasm and large nuclei, distributed around vascular channels, typically exhibiting a stag horn branching pattern. The vascular channels are often more readily defined following reticulin staining.

However, since its initial description the characteristic histological features in HPCs have been found to be non-specific and encountered in up to 15% of unrelated soft tissue neoplasms. In the 2005 World Health Organization classification of head and neck tumors, it was recommended that sinonasal-type HPC be referred to as glomangiopericytoma, in light of the similarity with glomus tumor.[9]

Immunohistochemically, the HPC cells are invariably positive for Vimentin, SMA but very rarely and only focally for smooth muscle markers such as desmin. A sub-population of tumor cells is also immunoreactive for factor XIIIa and histocompatibility antigen HLA-DR. CD34 usually stains only endothelial cell component, although sometimes there is a diffusion effect in the immediate vicinity.[10]

The treatment of choice is wide surgical resection which is the only curative modality. There has been an increasing usage of endoscopic resection techniques for sinonasal HPC because of its slow expansile growth which produces smooth instead of infiltrative tumor borders. Endoscope allows a magnified view and therefore accurate assessment of the site of origin and relation to surrounding structures. Also, it preserves the nasal physiologic function and avoids a large external incision. Factors that not favorable include septal deviation, tumors which are large or highly vascular, intracranial extension, orbit or the pterygopalatine fossa involvement. Therefore, patients have to be stringently assessed and selected.

The reported recurrence rate of HPC is quite varied, ranging from 7% to 20%, with an average time of recurrence of 6–7 years.[2] While an incomplete primary excision has been identified as the primary factor in recurrent disease, severe nuclear pleomorphism, osseous invasion, large tumor size (more than 5 cm) and a high mitotic to proliferation rate also appear to significantly increase the risk of recurrence.

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