

Two Case Reports of Sinonasal Glomangiopericytoma: A Very Uncommon Neoplasm

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Abstract: *Sinonasal glomangiopericytoma (formerly called hemangiopericytoma) is a rare vascular tumour, arising from Zimmerman's pericytes surrounding capillaries. It is an indolent tumour affecting older adults, with low malignant potential and excellent prognosis following complete resection, which is usually curative. Long-term follow up is essential part of the management, as recurrence can occur many years after treatment. In the past two decades, about 250 cases were reported. We represent here two case reports: both are for male patients in their sixties, presented with right sided sinonasal glomangiopericytoma. These case reports will be accompanied by a brief literature review.*

Keywords: Glomangiopericytoma, hemangiopericytoma, sinonasal tumor, endoscopic resection

1. Introduction

HPC is a very uncommon tumor of mesenchymal origin, forming less than 1% of all blood vessel neoplasms[1]. 15% of HPC occur in the head and neck region, of them, 1-5% occur in the nose and paranasal sinuses. It is derived from Zimmerman's pericytes, which are modified smooth muscle cells in the periphery of blood vessels[2].

In the sinonasal region, the tumour usually is benign with low tendency of metastasis. The recurrence rate can be up to 25%[3].

2. Case 1

60-year-old male, hypertensive patient admitted to the emergency department with acute onset of dyspnea and chest pain. Cardiac Investigation showed inferior wall ST segment myocardial infarction. He underwent an emergency coronary artery bypass graft. Several weeks after the operation, he mentioned that he has been complaining of chronic right nasal obstruction with occasional episodes of epistaxis. Nasal endoscopy showed a huge right nasal fossa mass. CT scan of the paranasal sinuses (images 1,2 and 3) showed heterogeneous mass in the right nasal fossa, measuring 37mm in height, 22 mm in width and 68 mm in depth. There was no intraocular or intracranial extension. The nasal septum was pushed to the opposite side. CT chest, abdomen and pelvis showed no further lesions.

MRI showed isointense lesion on T1, isointense on T2 with strong enhancement on T1 after gadolinium injection. Biopsy was done, which reveal the diagnosis of glomangiopericytoma. Surgery was done under neuronavigation guidance. The mass was adherent to the middle turbinate and lateral nasal wall. The lesion was completely removed through medial maxillectomy, using Denker approach, and the middle turbinate was sacrificed. The anterior skull base was drilled using diamond burr to ensure complete removal. Blood loss was 800 cc, and the patients was stable and didn't require blood transfusion. Histopathology report showed a benign HPC that was

positive for STAT6, CD99, BCL2, CD34, FXIIIa and CD31 on immunohistochemistry. The patient was regularly followed up to 18 months without any symptoms and signs of recurrence.

Image 1,2 and 3: coronal, sagittal and axial cuts (respectively) of CT paranasal sinuses showing tumor filling the right nasal cavity.

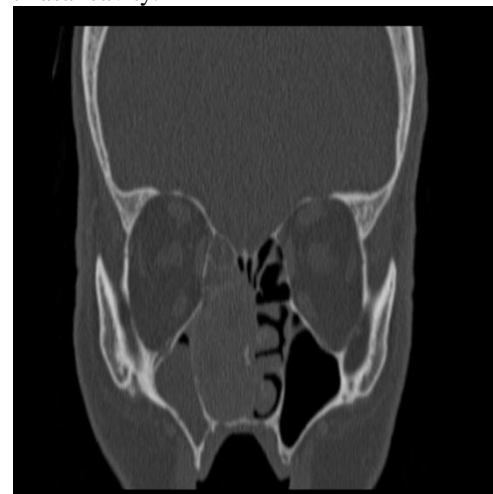


Image 1

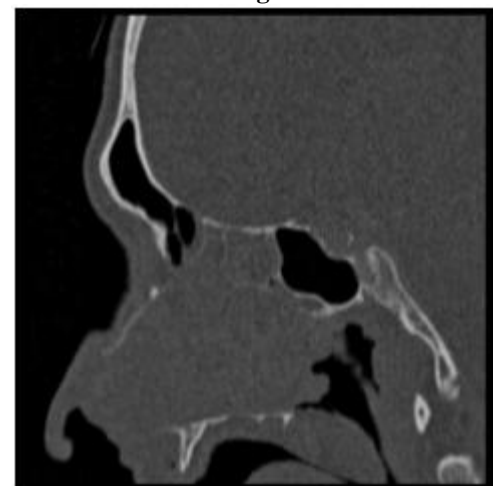


Image 2

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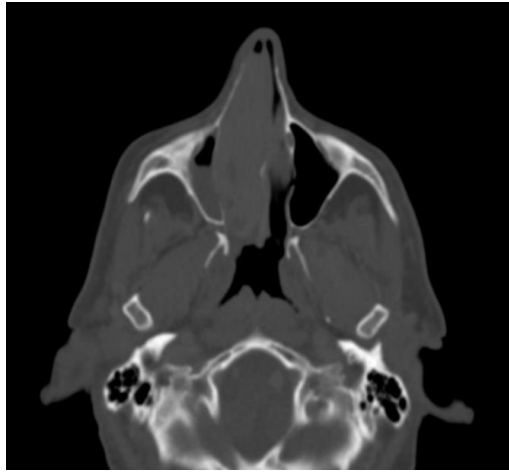


Image 3



Image 5

3. Case 2

64-year-old male patient, who had a complete endoscopic resection of glomangiopericytoma of the right ethmoid sinus 14 years ago, presented with moderate to severe headache over the period of few weeks, following a common cold. Nasal endoscopy showed a small red, firm lesion of the area of right posterior ethmoid, in contact with lamina papyracea. Also, there was a second huge lesion of the sphenoid sinus. Paranasal sinus CT scan showed huge heterogeneous lesion of the sphenoid sinus, extending into right cavernous sinus, measuring 42 x 50 x 43 mm, with also intracranial extension as well. T1 weighted images on MRI showed a cystic isointense lesion with strong enhancement after gadolinium administration. The mass was hypointense on T2 weighted images. The cavernous portion of the right internal carotid artery was completely surrounded by the tumour. Biopsy confirmed recurrence of the disease. Due to the difficult access to lesion and its proximity to vital structures (eye, cavernous sinus, internal carotid artery and brain), debulking endoscopic resection was done. Pathology report showed no mitosis or cellular atypia. The patient underwent complementary treatment by radiotherapy, the lesion got shrunken and the patient clinically stable.

Image 4 and 5: axial and coronal (respectively) of T1-weighted MRI after gadolinium injection showing the lesions in the nasal cavity with intracranial extension.

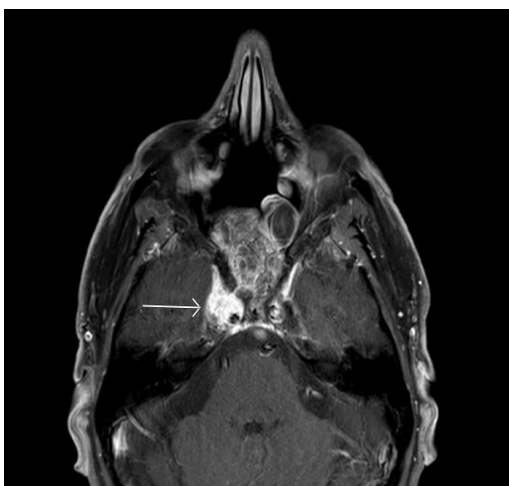


Image 4

4. Discussion

HPC was first described at 1942 by Stout and Murray[4]. It is derived from mesenchymal cells with pericytes differentiation. In 2005, the WHO classification of head and neck tumours stated that sinonasal HPC should be named glomangiopericytoma, due to its close similarities in behaviour and histopathology to glomus tumours[5].

About 5% of HPC occurs in the nose and paranasal sinuses. It affects adults, usually in 6th or 7th decade of life, although 10% of cases occur in children[6]. It has no sex predominance[7].

Multiple risk factors have been proposed, including hypertension, trauma and corticosteroids use, but none of them have been accepted[8].

Clinically, the tumour appears firm, well circumscribed and usually well-vascularized, rendering biopsy not advisable except in selected cases. Symptoms include long standing unilateral nasal obstruction, epistaxis, hyposmia, and in more aggressive forms, patients might present with pain and diplopia due to local invasion and bony destruction.

Diagnosis is made by endoscopic examination and by imaging. CT scan shows homogenous well circumscribed soft tissue mass with bony destruction and strong enhancement after intravenous contrast administration. On T1-weighted MRI, the lesion is usually isointense that strongly enhances after gadolinium administration. It appears low intense to isointense on T2-weighted images.

Histologically, the tumour can be classified as benign, low-, intermediate- or high-grade malignancy, depending on cellular pleomorphism, mitosis and cellularity. Microscopic features included diffuse growth with fascicular, solid or focally whorled pattern of spindle or oval tumour cells that arrange themselves around prominent, small, thin walled submucosal blood vessels. Vessels are prominent with staghorn appearance [9] and perivascular hyalinization.

Treatment modality of choice is wide surgical excision; with open approaches were the standard way of excision.

Nowadays, endoscopic resection is becoming more popular way of management, as it is associated with more accurate assessment of the site of origin and less blood loss, especially with preoperative embolization[10]. Chemotherapy, radiation and also immunotherapy are used for malignant, inoperable or palliative cases. Prognosis is favorable, with 5-year survival rate is more than 90%. Factors in favor of poor prognosis include tumor size of more than 6,5 cm and histological findings of necrosis, nuclear atypia and high number of mitosis[11]. Reported recurrence rate varies from 0.9 to 50%, with some cases happen up to 25 years after primary resection, making long term follow up an essential part of managing such lesions.

5. Conclusion

Glomangiopericytoma is a very rare tumor of the nose and paranasal sinuses. It arises from capillary pericytes, and it is usually benign but can be malignant as well, depending on the number of mitotic figures, hemorrhage, necrosis and increased cellularity. Diagnosis is made by clinical and radiological assessment. Surgery can be curative, with wide locale excision using endoscopic method becoming the standard way of treatment. Long term follow up is essential to detect recurrence.

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