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Case Report

Imaging features of a rare case of Sinonasal Glomangiopericytoma

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Abstract

Glomangiopericytoma (GPC) is a rare unique sinonasal mesenchymal neoplasm arising from the pericytes surrounding the capillaries. It constitutes < 0.5 % of all sinonasal neoplasms.

It is categorised as a borderline or low-malignant potential tumour by the World Health Organisation (WHO). Only a few cases of Glomangiopericytoma of sinonasal cavity have been reported so far. We report a case of a 48-year-old female who presented with features of nasal obstruction and epistaxis. She underwent contrast enhanced CT of paranasal sinuses which showed an avidly enhancing mass involving predominantly the right sinonasal cavity. It is important to recognize this highly vascular lesion and differentiate it from other malignant and inflammatory lesions affecting the sinonasal cavity as it can result in frequent episodes of epistaxis. Complete surgical resection and follow up is essential due to its high recurrence rate.

Keywords: Glomangiopericytoma, sinonasal cavity, neoplasm, malignant, rare, nasal obstruction, epistaxis, avidly enhancing.

Introduction

Glomangiopericytoma is a rare highly vascular neoplasm. It was formerly called as "Sinonasal hemangiopericytoma" because of its hemangiopericytomatous pattern.[1] It differs from soft tissue hemangiopericytomas in terms of its location, biologic behaviour and histologic features, hence, Sinonasal hemangiopericytomas represent a distinct entity.[2] Etiology remains unknown. It usually arises in the nasal cavity and may extend to the paranasal sinuses. Peak incidence is during the sixth or seventh decade with a slight female predominance. It typically presents as a unilateral nasal mass with obstruction and epistaxis.[3] It has excellent prognosis after complete surgical resection. Metastases is rare, however characterized by frequent recurrences. Hence, long term follow up is necessary.[4]

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Case History

A 48-year-old female presented with complaints of nasal obstruction, facial pain and head ache, on and off since 1 year and epistaxis on and off since 3 months, increased since 1 day. She is a known case of hypertension. No other predisposing factors. No history of nasal trauma/surgery.

On examination, the external framework of her nose was normal.

After control of epistaxis, nasal endoscopic examination was performed which revealed a reddish smooth mass in her right nasal cavity which was friable and bled easily on touch.

Patient was then referred to our department for CECT PNS study.

Imaging features

On CECT PNS study: An ill-defined avidly enhancing mass was noted epicentered in the right nasal cavity. Medially, the mass was causing erosion of the bony nasal septum and extending into the left nasal cavity. Superiorly, it was eroding the walls of ethmoid sinuses with extension into the sinus. Posteriorly, it was eroding into the wall of the sphenoid sinus with extension into right sphenoid loculus and inferiorly, into the choana. None of the nasal turbinates were visualized separately. There was no evidence of any intracranial / infraorbital extension (Shown in Figure 1- 13).

CT Scan (Plain Study, Tissue Window):

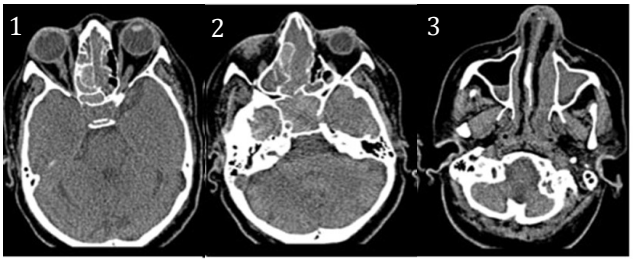

Figure 1,2 & 3: CT Scan plain study, soft tissue window, axial sections extending from the level of bilateral ethmoid sinuses upto bilateral nasal cavities & nasopharynx, shows an ill-defined soft tissue density lesion in bilateral ethmoid sinuses, nasal cavities, posteriorly extending into the sphenoid sinus. Also noted soft tissue densities in bilateral maxillary sinuses.

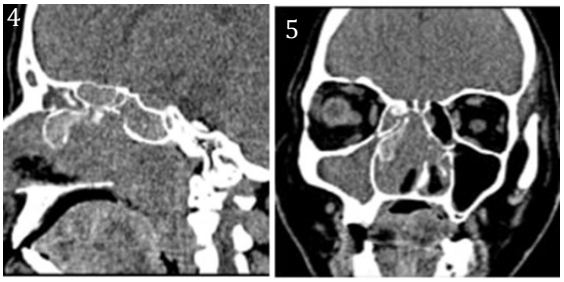

Figure 4 & 5: CT scan plain study, soft tissue window, sagittal and coronal reformatted images, shows an ill-defined soft tissue density lesion in bilateral nasal cavities, posteriorly extending into the choana and sphenoid sinuses, superiorly extending into the ethmoid sinuses. There is also complete opacification of right maxillary sinus and obliteration of right maxillary sinus ostium.

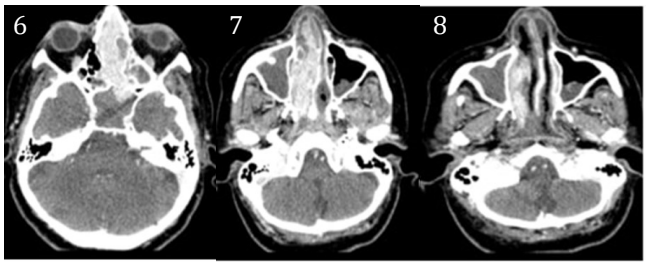

Figure 6, 7 & 8 : CT scan , contrast study , axial sections from the level of ethmoid and sphenoid sinuses upto the nasal cavity and nasopharynx, shows an ill-defined avidly enhancing mass in bilateral ethmoid sinuses, predominantly involving the right nasal cavity and posteriorly extending to the sphenoid sinus.

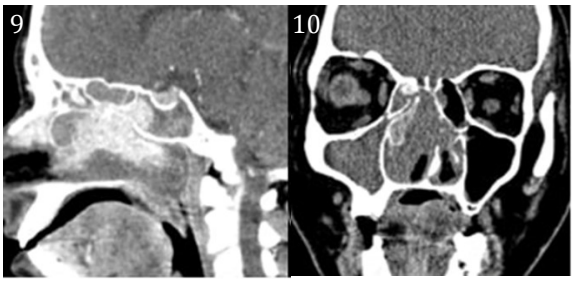

Figure 9 & 10 : CT scan, contrast study, sagittal and coronal reformatted images, shows an ill-defined avidly enhancing mass in the nasal cavity, posteriorly extending into the choana and sphenoid sinuses, superiorly extending into the ethmoid sinuses. Non enhancing soft tissue density areas are also noted in the nasal cavities and right maxillary sinus – likely mucosal thickening.

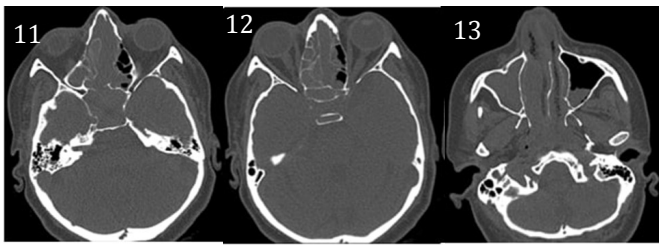

Figure 11,12&13: CT scan, bone window, axial section shows that the mass is causing erosion of he anterior wall of sphenoid sinus, few of the walls of ethmoid sinuses and part of the boney nasal septum. The remainder of the nasal septum appears deviated towards the left side.

Operative details

Patient underwent infrastructure maxillectomy and endoscopic debridement of the mass. Pre-operative embolization was carried out by ligation of the external carotid artery. No intraoperative or post-operative complications were noted.

A red coloured unilocular mass with smooth surface was observed intraoperatively predominantly occupying the right nasal cavity and was seen extending to the anterior & posterior ethmoid air cells, superiorly involving the sphenoid sinus and inferiorly extending to involve the nasal choana. Mucopus was noted in the maxillary antrum (Shown in Figure 14- 16).

Post-surgery, intranasal tamponing with antibiotic ointment was done.

Patient was advised to be under regular follow up due to high incidence of recurrence of the tumour.

The entire resected vascular mass along with the anterior maxilla was sent for histopathological analysis.

INTRA-OPERATIVE IMAGES (Figure 14, 15 & 16):

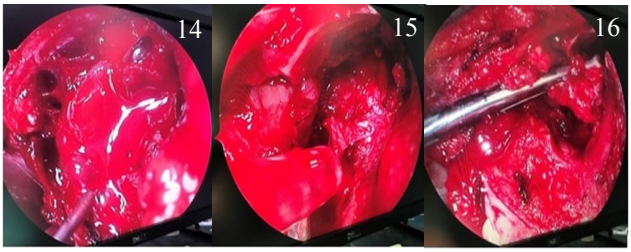

Figure 14, 15 & 16: Intra-operative nasal endoscopic images shows a highly vascular, red coloured, unilocular mass within the nasal cavity.

Histopathology

Histopathology report with haematoxylin and eosin staining of the specimen showed multiple tissue bits showing respiratory epithelium. Subepithelium showed multiple vascular channels with surrounding stromal cells which were round to oval, with minimal nuclear atypia and absent mitotic figures. Stroma appeared highly cellular with no area of necrosis. Section from the bone showed respiratory epithelial lining with areas of squamous metaplasia. Subepithelium showed fibrocollagenous tissue and bony trabeculae with no atypical cells.

Above features were consistent with Glomangiopericytoma of the right sinonasal cavity.

HEMATOXYLIN AND EOSIN-STAINED IMAGES (Figure 17 & 18):

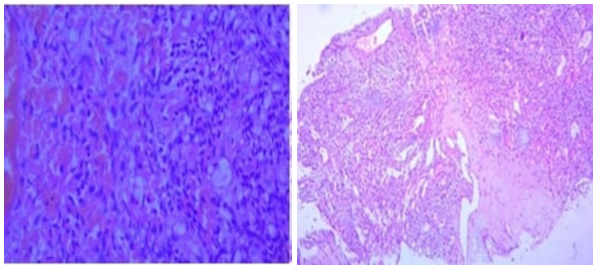

Figure 17 & 18: Hematoxylin and eosin-stained images shows multiple tissue bits showing respiratory epithelium. Subepithelium shows multiple vascular channels with surrounding stromal cells which are round to oval, with minimal nuclear atypia and absent mitotic figures.

Discussion

Sinonasal neoplasms are rare, accounting for ~3% of the head and neck malignancies and ~3.6 % of upper aero-digestive tract malignancies.[5] CT and MRI are important imaging modalities used in evaluation of patients with Sinonasal neoplasms.

Sinonasal tumours can be either epithelial or non-epithelial tumours. Non-epithelial tumours include neuroectodermal and nervous system tumours, mesenchymal tumours, osseous and cartilaginous tumours, lymphoreticular and fibro osseous tumours.[3]

GPC of sinonasal cavity is a highly vascular mesenchymal tumour and constitutes < 0.5 % of all sinonasal neoplasms. Other highly vascular tumours involving the sinonasal cavities include juvenile nasopharyngeal angiofibroma, hemangioma and glomus tumour.

The concept of hemangiopericytoma/GPC was first described in the year 1942 by Stout and Murray as soft tissue tumours with characteristic vascular proliferation including branching vessels and small vessel perivascular hyalinization. This lesion was thought to fall in the spectrum between glomus tutors and capillary hemangiomas, and hence the term hemangiopericytoma was chosen, which is now known as GPC.[6]

Other synonyms of GPCs include Sinonasal-type hemangiopericytoma, hemangiopericytoma-like tumour and hemangiopericytoma of Sinonasal origin.

Etiology of GPCs remains unknown; however past trauma, hypertension, pregnancy and use of corticosteroids are considered as predisposing factors. Peak incidence is during the sixth or seventh decade with a slight female predominance. Most common symptoms include epistaxis and/or nasal obstruction.[3]

Studies have proven that most cases of sinonasal glomangiopericytoma originated most commonly from the nasal septum followed by the paranasal sinuses.[7]

Clinically, these tumours are variable in size (average size being ~ 3.1 cm, ranging between 1-8 cm)[8] and firm, soft, red or fleshy, haemorrhagic, friable, polypoid masses.[3]

GPC of Sinonasal origin are clinically and pathologically different from soft tissue GPCs.

Gross appearance of these lesions is not generally useful. If intact, cut surface may be solid, soft, fleshy or friable, or haemorrhagic and/or edematous areas may be present.

On microscopy, these tumors usually have fascicular growth, however, storiform or whorled growth patterns can also be seen.[6] They are usually covered with normal respiratory epithelium.[7] Squamous metaplasia[8] and ulcerations on the surface epithelium are rarely present. Thin free zone of basement membrane and neoplastic tumour cells are detected.[9] Unlike glomus tumours, cells have a syncytial appearance, such as having lack of obvious cell borders and eosinophilic cytoplasm. The cells are oval, pale-staining chromatin and have one or more small nucleoli. Most tumour cells have inflammatory cells and extravasated red blood cells. Mild cytologic atypia and mitotic figures may be seen, but necrosis is not found .[11] A small percentage of tumours have giant cells, clear cells or show myxoid degeneration.[8] Numerous thin-walled, branching staghorn vessels with perivascular hyalinization can be seen.[7]

Immunohistochemical profile shows that the tumour cells stain for vimentin, smooth muscle actin, muscle specific action, Bcl-2 and beta-catenin. Occasional variable staining for CD34, S-100, CD99 and CD 31 has been reported.[11] Negative staining of desmin and keratin. Additional negative staining for Bcl-2, CD99 and CD117 are also found. Few of the cases showed immunoreactivity for cyclin D1, progesterone receptor, CD 146 , WT1, mTOR and EGFR. Studies have suggested mutational activation of beta-catenin and associated cyclin D1 overexpression may be central events in the pathogenesis of glomangiopericytoma and nuclear accumulation of beta-catenin can be a diagnostic marker for glomangiopericytoma.[7] large tumours invade bone and show deep nuclear pleomorphism, necrosis and high mitotic proliferation and Ki67 proliferation index is more than 0.10%.[12]

On imaging studies like on a CT scan, the mass appears as a well-defined or an ill-defined lobulated, avidly enhancing soft-tissue mass with erosive bony remodeling.

On MRI, the mass appears as T1 hypointense, T2 hyperintense with multiple flow voids showing high mean ADC values and shows avid post contrast enhancement and wash-in and wash-out pattern on dynamic contrast-enhanced MRI study.

Tumour is known for frequent recurrences; however, metastases is rare. Hence, long term follow up is necessary.

Treatment includes complete surgical resection with preoperative embolization as the tumour is highly vascular. Best surgical approach is via the trans nasal endoscopic route, unlike our case where an open approach was followed (infrastructure maxillectomy and endoscopic debridement of the mass). It is associated with 5-year survival greater than 90% after complete surgical resection. Local recurrence has been reported in 7% to 40% of cases because of incomplete surgical resection; however, aggressive behaviour is rare.[5]

GPCs can be considered as a differential diagnosis for any highly vascular sinonasal neoplasm. Differential diagnosis with respect to our case includes Sinonasal hemangioma, intranasal glomus tumour/glomangioma and rare possibility of Sinonasal angiofibroma can also be considered.

Conclusion

Even though sinonasal glomangiopericytoma is categorised as borderline low malignant tumour by the WHO, a minority can still recur or can even be fatal due to the possibility of uncontrolled epistaxis. In our case, even though the mass was completely resected, patient was asked to be on regular follow up due to high chances of recurrence of the disease.

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