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Case Report

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A rare sinonasal tumour with orbital manifestation: glomangiopericytoma

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ABSTRACT

Sinonasal glomangiopericytoma is a tumor showing similarities to a glomus tumour but is less aggressive and has a lower malignancy potential. It was initially reported by Stout and Murray in 1942. Our patient was a 19 years old lady who presented to us with unilateral epistaxis during her pregnancy. Further investigations including a nasopharyngeal biopsy showed signs of an aggressive behaving sinonasal glomangiopericytoma. She underwent left medial maxillectomy via lateral rhinotomy approach to achieve complete excision of the tumor. This was vital as past literatures suggest that incomplete excision of tumor is the most pertinent factor to tumor recurrence. Distant metastasis is rare. Complete excision with regular long term follow up is the treatment of choice. The role of chemo and radiotherapy remains controversial.

Keywords: Glomangiopericytoma, Sinonasal tumour

INTRODUCTION

Glomangiopericytoma is a rare tumor of low malignant potential. It was first reported as hemangiopericytoma by Stout and Murray in 1942.[1] Since then, its definition has been in much debate. In 2005, World Health Organization identified it as a unique sinonasal low malignancy lesion and proposed the term 'glomangiopericytoma' and 'myopericytoma' because of its similarity with glomus tumors.[2]

We present a case of aggressive behaving sinonasal glomangiopericytoma (malignant glomangiopericytoma) treated with surgical resection in Hospital Sultanah Bahiyah, Alor Setar.

CASE REPORT

A 19 years old, Myanmar lady came to our clinic at 28 weeks of pregnancy complaining of left nostril epistaxis for past 1 year with alternating nasal obstruction. Otherwise, she has no known medical illness with no complaints of pain or visual disturbance. Nasal endoscopy revealed a fleshy irregular mass occupying the whole of left nasal cavity, most likely arising from the roof of nasal cavity with contact bleeding. Patient returned to our clinic 14 weeks later after delivery, with slight left eye proptosis but vision and extraocular movement still normal. CT scan revealed a left nasal mass of 5.8 cm×3.8 cm×4.1 cm with the involvement of left ethmoid, frontal and maxillary sinuses. The mass had no clear plane of demarcation with the left middle and inferior turbinate with erosion to the left lamina papyracea and extending to extraconal space of left orbit, indenting onto left medial rectus muscle. It also caused erosion to medial wall of maxillary sinus. In view of financial constraints, no MRI was done. However, clinically patient had no signs of cerebral infiltration. Biopsy result suggestive of aggressive behaving sinonasal glomangiopericytoma.

In view of extend of the tumor, the patient underwent left medial maxillectomy via lateral rhinotomy approach. Intraoperatively, the tumor was seen occupying the entire left nasal cavity extending superiorly to the olfactory groove, inferiorly to the nasal wall, laterally into the entirely maxillary cavity and posteriorly to the choanae. The left lamina papyracea was absent. Left frontal, maxillary and ethmoid sinus explored, tumor removed in piecemeal. Intranasal tamponing was done with bismuth iodine paraffin paste impregnated ribbon gauze and kept for 5 days. No postoperative complications were noted.

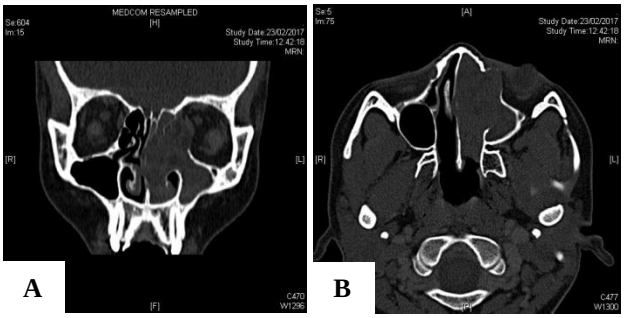

Figure 1 (A and B): An axial and coronal CT scan view showing glomangiopericytoma filling the left nasal cavity extending into the left maxillary sinus and left extraconal space abutting on the medial rectus muscle.

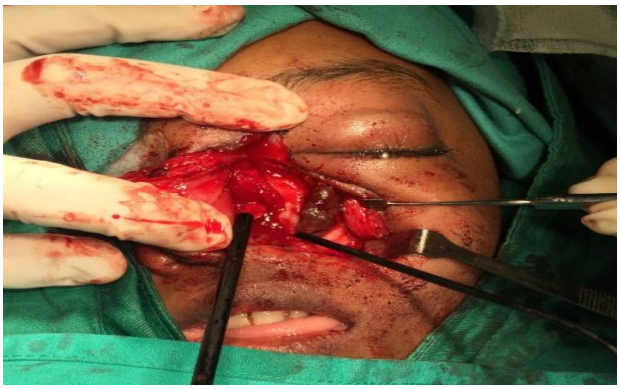

Figure 3: Intraoperative, tumor occupying entire left maxillary sinus.

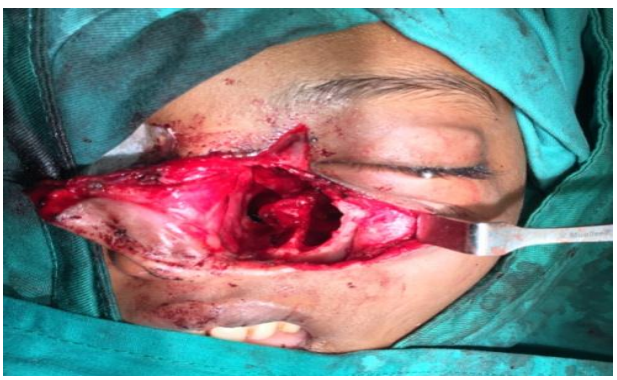

Figure 4: Post-operative tumor excision.

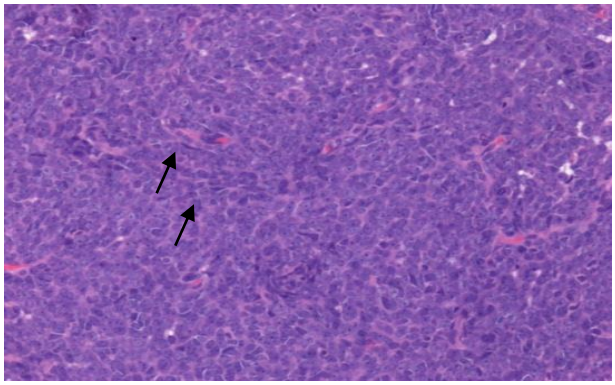

Figure 5: Tumor cells display oval to elongated nuclei, indistinct cell border with scattered mitosis (6/10 h.p.f.). Many blood vessels are seen in between the cells. No abnormal mitosis noted.

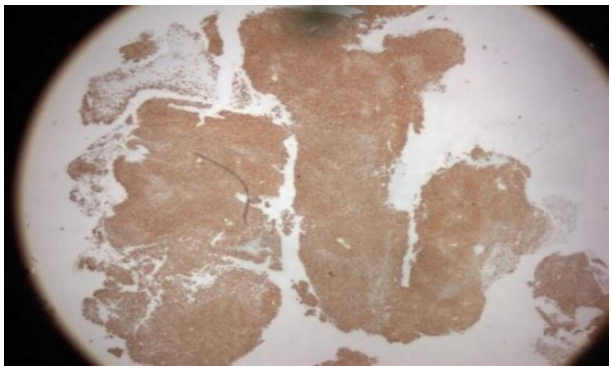

Figure 6: Tumor cells stain positive to vimentin.

DISCUSSION

Glomangiopericytoma, also known as sinonasal hemangiopericytoma is a rare type of tumor of mesenchymal origin. The disease location, clinical characteristics, histopathological features and mortality rates of glomangiopericytoma differs from conventional soft-tissue hemangiopericytoma.[2,3] Glomangiopericytoma arise almost exclusively from the nasal cavity or paranasal sinuses and is characterized histologically by a pattern of prominent perivascular growth. Unlike soft-tissue hemangiopericytoma, it has borderline low malignant potential and only accumulates to <0.5% of all sinonasal neoplasms.[4,5]

Past studies suggest that glomangiopericytoma can occur in all ages with a peak at sixth to seventh decade and slight preponderance in the female group.[6] The exact etiology is unknown but several factors has been proposed such as post trauma, hypertension, pregnancy and the use of corticosteroids.[7]

Clinically, patients may present with unilateral nasal obstruction and recurrent epistaxis, to a lesser extend visual disturbance and pain. A soft, fleshy, polypoidal beefy red to greyish mass can be found endoscopically and is usually friable to touch. Lymph node involvement

is rare.[3,4,7] Generally, CT and MRI imaging characteristically shows a non-calcified soft- tissue mass in the nasal fossa or paranasal sinuses which may sometimes be mistaken for an inflammatory polyp.[5,8] An imaging will also provide details of the extend of the tumor and reveal an aggressive pattern of spread that contributes to the decision of disease management.

The diagnosis of glomangiopericytoma is based on histopathology with the characteristic findings of richly vascularized tumor with stag horn branching pattern and prominent perivascular hyalinization.[5,9] Immuno-histochemical studies showed that the tumor cells were strongly positive to vimentin, α-smooth muscle actin and negative to CD34. A sub-population of tumor cells are also immunoreactive for factor XIIIa and histocompatibility antigen HLA-DR.[5]

Despite the distinctive architectural pattern and characteristic histological features, difficulties exist in attempting to predict the biological behavior of the tumor. Kowalski et al suggest that proliferation index with immunoperoxidase stain for Ki-67 of >10% may be indicative of a more aggressive subset.[5,10]

Aggressive behaving glomangiopericytoma is uncommon and usually exhibit large size tumor (>5 cm), bone invasion, profound nuclear pleomorphism, increase mitotic activity (>4/10 h.p.f), necrosis and >10% proliferative index.[4,5,10]

In our patient, the only predisposing factor was pregnancy. However, features of bone invasion on CT imaging with large tumor size, histopathological result showing increase mitotic activity (6/10 h.p.f) and Ki67 proliferation index of 40% leads us to a conclusion that this patient had sinonasal glomangiopericytoma with aggressive behaving features.

Complete excision of tumor is the treatment of choice as it is an indolent tumor with excellent prognosis after surgical resection. In cases where the tumor is well vascularized, preoperative angiography and embolization has been suggested to minimize blood loss and facilitate surgical tumor resection.[11] In the modern age, endoscopic surgery is preferred as it offers magnified view with minimal blood loss and allowing patients to preserve their natural physiology of the nose.[11-13]

In this patient, preoperative angiography and embolization was not done in view of financial constrains as patient is a foreigner. Endoscopic approach alone was deemed insufficient as CT imaging displayed evidence of mass indenting onto the nasal septum with orbital extension. Hence, combination of endoscopic plus open resection was done to achieve complete excision of the tumor with better intraoperative hemostasis control.

Case studies have suggested that incomplete excision was the most important predictor of recurrence.[12,13]

Few studies reports the use of radiotherapy and chemotherapy in cases of inoperable tumors, metastasis or palliative therapy. Limited study data showed that patients receiving radiotherapy following an incomplete surgical resection has lower rate of recurrence.[12] However, small number of data limits the assessment of its treatment efficacy.[14] The question of whether and when to initiate adjuvant therapy is not quite clear in the current literature, nor are prognostic factors widely mentioned.

Although glomangiopericytoma has a borderline low malignancy nature and distant metastasis is exceedingly rare, recurrence rate of 26.7% have been reported.[15] In view of the high recurrence rate, long term follow up is vital in early detection of potential recurrence and possible metastasis.

CONCLUSION

Glomangiopericytoma are rare tumors with characteristics of perivascular myoid phenotype. A high index of suspicion is needed for diagnosis of these tumors as multiple case studies describe patients being misdiagnosed and undergoing polypectomy before reaching the correct diagnosis. Orbital involvement of such tumor is exceedingly rare and their incidence is estimated to be 0.8% to 3% of primary orbital tumors.[16] It most commonly have an indolent growth pattern but a small cluster may exhibit aggressive behavior or metastasis. Complete excision is the treatment of choice. Long term follow up is required as recurrence has been reported to occur up to 17 years following initial presentation.[12] The role of chemo and radiotherapy remains controversial.

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