CASE REPORT

Sino-Nasal Hemangiopericytoma - A Rare Tumor

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Summary

Hemangiopericytoma is a very rare angiogenic tumor. In the nasal cavity, it can be considered malignant. It occurs in various parts of the body but those in the nasal cavity account for only 5% of total cases[1]. Less than 200 cases have been reported worldwide involving the nose and paranasal sinuses. Due to its rarity a proper line of management has not been established to tackle this tumour. This article highlights two cases of hemangiopericytoma (HPC), one in an adult and the other in a child, presenting as an intranasal mass.

Key Words: Hemangiopericytoma, Sino-nasal vascular tumors, Endoscopic tumor resection

Introduction

Epistaxis and/or nasal obstruction are the most common symptoms among patients who present to ENT clinic. It causes much anxiety to the patient and clinicians alike. It is imperative to exclude an intranasal mass, whether benign or malignant. We present two cases of hemangiopericytoma (HPC) that presented as an intra nasal mass. HPC is a rare tumor of variable malignant potential that is thought to originate from the vascular pericytes of Zimmerman[1]. Although they represent less than 1% of all vascular neoplasms, 15% to 30% of these tumors are found in the head and neck, most commonly in the nasal cavity and paranasal sinuses.

Case 1

A 12 year-old Chinese girl presented with intermittent epistaxis with gradually worsening left nasal obstruction over a 2 month period. On examination, there was a reddish granular mass in the left nasal cavity which was attached to the nasal septum. The mass bled on touch, therefore biopsy was deferred. A contrast enhanced CT scan of the paranasal sinuses revealed a heterogenously enhancing vascularised soft tissue mass in the anterior part of the left nasal cavity measuring 1.1cm by 0.6cm by 1.3cm. There was no evidence to suggest an extension into adjacent structures or bony erosion. A wide excision biopsy under general anaesthesia was performed. Intraoperatively, the base of the tumor was vascular in nature but was easily detachable. Postoperative recovery was uneventful. No tumor recurrence was noted fifteen months post surgery.

Case 2

A 57 year old Chinese lady on treatment for hypertension presented with a history of intermittent nasal obstruction for six years. Examination revealed a pulsatile right nasal mass arising from the posterior part of the nasal septum, anterior ethmoids and sphenoid. Serial CT scans of the paranasal sinuses revealed a soft tissue mass almost completely filling the right nasal cavity causing compression / displacement of the medial wall of the right ethmoid sinus and the nasal septum. It also revealed evidence of bony erosion

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involving the inferior wall of the right sphenoid sinus (Fig 3). An endonasal endoscopic removal of the tumour with spheno-ethmoidectomy and a wide middle meatal antrostomy was performed under general anaesthesia. She recovered well postoperatively and was tumor free after eleven months post surgery.

The histology in Case 1 and Case 2 was reported as HPC (Fig 1 and 2).

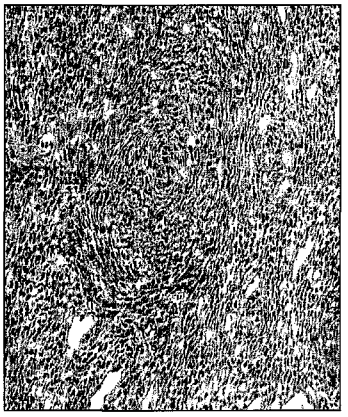

Fig. 1: (Case 2) shows a cellular tumor, arranged in a whorling pattern, consisting of monotonous cells and indistinct eosinophilic cytoplasm. Mitotic activity is generally absent. Blood vessels range from small capillary-sized vessels to sinusoidal spaces with little intervening collagen.

Fig. 2: Immunohistochemistry showing positive staining for smooth muscle actin in tumor cell.

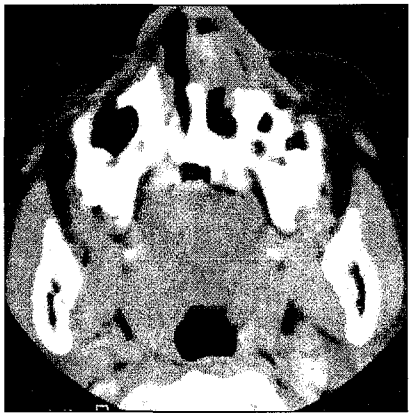

Fig. 3: Axial CT scan of Case 1 showing a localized tumor arising from the septum and obstructing the nasal cavity.

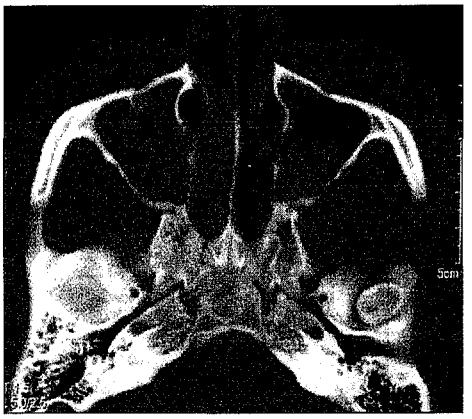

Fig. 4: Axial CT of the paranasal sinuses of Case 2 shows a soft tissue mass completely filling the right nasal cavity and causing compression/displacement of the medial wall of the right ethmoid sinus and the nasal septum.

Discussion

First described by Zimmerman, "pericyte" refers to the amoeboid cells in the perivascular space. Stout and Murray in 1942 coined the term "hemangiopericytoma" to describe a tumor composed of tightly arranged spindle cells around a central lumen lined by endothelial cells[1]. Histology can be extremely variable. Their etiology is unknown but, trauma has been implicated as a factor. Majority are slow growing, benign tumors that are well-differentiated and locally infiltrative, with malignant potential.

Pathology

Grossly, HPC are red or grey yellow, soft, fragile hypervascular lesion[2]. It has been suggested they are the only tumours that truly show pericyte differentiation among the hemangiopericytomas arising in adulthood.

Most sinonasal hemangiopericytomas are well-circumscribed and have a uniformly bland appearance. The tumor cells are round to oval shaped, with indistinct eosinophilic cytoplasm. Nucleoli are inconspicuous and mitotic activity is generally absent. Blood vessels range from small capillary-sized vessels to sinusoidal spaces with stag horn shape. Vimentin and actin are the only consistent positive stains in these tumors.

Behavior is generally indolent though very rare metastatic tumors have been described. Sino-nasal hemangiopericytoma are classified as malignant neoplasm but clinically may manifest as either benign or malignant[3]. Late recurrence is seen in up to 50% of cases with a 10% incidence of metastasis[4]. They are mainly non lethal, as reported by El-Naggar et al of 14 cases that were followed up to 120 months[5].

Clinical Features

A benign course is more common in the nose and paranasal sinuses. Most patients with sinonasal HPC present with epistaxis and / or nasal obstruction months or years prior to diagnosis. Less frequent symptoms include local pain, impaired visual acuity, headache, local swelling, epiphora and diplopia. In this case report, one patient presented with intermittent nasal obstruction and the other with intermittent epistaxis and obstruction.

Management

Differential diagnosis of a bleeding nasal mass includes angiofibroma, pyogenic granuloma, leiomyoma and malignant fibrous histiocytoma. However, final diagnosis is still based on histology. HPC affect all ages, majority presenting between the 20th and 70th decade (median age 46 years[2]). Incidence in childhood has been reported to be about 10%. Indeed, very few cases of pediatric head and neck HPC have been documented. The youngest documented age is two and a half years. Gender or race predominance is not well demonstrated.

Intranasal biopsies may be performed, beware the risk of hemorrhage. CT and MRI are useful to assess exact site and extent of the tumor. The degree of tumor vascularity, feeding vessels or arteriovenous shunting can be clearly demonstrated on angiography. The role of angiography in pre-operative embolisation is debatable. Some studies have suggested pre-operative embolisation as a routine to reduce tumor vascularity and size therefore reducing the intraoperative hemorrhage. In both our cases pre-operative embolisation was deferred and intra operative bleeding was minimal. Literature review suggests that embolisation is not essential for HPC excision.

The treatment of choice is wide surgical resection which is the only curative modality. There has been increasing usage of endonasal endoscopic resection techniques for sinonasal HPC. Its slow expansile growth producing smooth instead of infiltrative tumour borders may allow for endoscopic resection with negative margins. Endoscope allows a magnified view and therefore accurate assessment of the site of origin and relation to surrounding structures, not afforded by open techniques. Also, it preserves the nasal physiologic function and avoids a large external incision. Factors not favorable to an endoscopic approach include septal deviation, tumours which are large or highly vascular, intracranial extension, orbit or the pterygopalatine fossa involvement[3]. Therefore, patients have to be stringently assessed and selected to maximize the advantages of the endoscopic approach. Endoscopic techniques should always be used with conversion to open or external approaches kept as a possible option should complications arise. (e.g. torrential hemorrhage or inadequate margins.)

Other approaches used include lateral rhinotomy, Caldwell-Luc or transfacial approach. These procedures increase morbidity and prolong hospitalization. Our patients have an average hospital stay of 4 days. Other modalities of treatment described in the literature included radiotherapy and/or chemotherapy which have doubtful benefit.

Recurrence rate has been reported at about 20%[2], and are more commonly delayed, with most occurring greater than 5 years after initial surgery. Early recurrences are often without symptoms. Certain gross features associated with malignancy are tumor greater than 6.5cm, anatomic sites in the retroperitoneum, extremities, trunk and recurrent lesions. The clinical course of the tumor has been correlated to the tumour grading. Necrosis, nuclear atypia, high mitotic counts are associated with poorer outcome[3]. Definite confirmation of malignancy is based on histopathologic evidence as there are no pathognomonic signs or symptoms. Even if malignant, the tumor seldom metastasizes. The common sites being the lungs, liver, bone and regional lymph nodes. Also, the histologic features and diagnosis do not always correlate with the clinical behavior of the lesion. Therefore, long-term surveillance cannot be overemphasized as all HPC have malignant potential.

As the diagnosis of HPC is made easier by the advent of special immunohistochemistry staining techniques and electron microscopy, HPC will be less an enigmatic tumor than it used to be and removal less problematic as advanced endoscopic techniques continues to evolve. Endoscopic techniques promise great surgical advantage for both the surgeon and the patient provided proper patient selection and pre-operative planning is done in expert hands. The benign or malignant nature of a hemangiopericytoma must be monitored through close and careful follow-up. A lifelong follow-up is mandatory.

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