

Sinonasal Hemangiopericytoma: A Case Report

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Abstract

Hemangiopericytoma is a vascular tumor that is rarely seen in the nose and paranasal sinuses. Biological behavior of these tumors is not completely known, and their natural history is not predictable. This case report presents a patient with a sinonasal hemangiopericytoma. Diagnosis and management of these uncommon tumors is discussed.

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Keywords • Nasal sinonasal • hemangiopericytoma

Introduction

Mesenchymal soft tissue tumors are rare in head and neck. The ratio of all sarcoma tumors arising from this region is 5–10%. In addition, most of these lesions are not completely specific to head and neck.¹ Hemangiopericytoma (HPC) is an uncommon vascular tumor that involves most frequently in retroperitoneum, pelvis and skeletal muscles of preferably lower extremities. It is extremely rare in the head and neck region.^{2,3}

Case Description

The case was a 23 year old female with one year history of left nasal airway obstruction, rhinorrhea and frequent epistaxis. She had no ophthalmologic symptoms and was otherwise asymptomatic.

Physical examination revealed the obstruction of left nasal fossa due to medialization of lateral nasal wall. There was no sign of facial edema, and globe positions and movements were normal. Contrast enhanced computer tomography (CECT) confirmed a unilateral noncalcified soft tissue mass. Vascular nature was apparent considering the enhancement of the mass (figure 1). The biopsy was consistent with a vascular tumor.

Subsequent Magnetic Resistance Angiography (MRA) revealed a vascular mass supplied predominantly by the left internal maxillary artery (figure 2). Embolization was carried out, and the next day a left medial maxillectomy was performed. The mass was excised with an adequate margin, which was confirmed to be free of tumor by frozen sections.

The pathological examination of the mass showed a circumscribed cellular tumor comprised of tightly packed small cells surrounded by an intact reticulin sheath (figure 3). The pathology finding was compatible with the diagnosis of hemangiopericytoma.

Postoperative clinical course was uneventful. Since, the excised tumor margins were completely free of tumor cells, post operative radiotherapy was not used. The patient remained free of the disease for 5 years after the operation.

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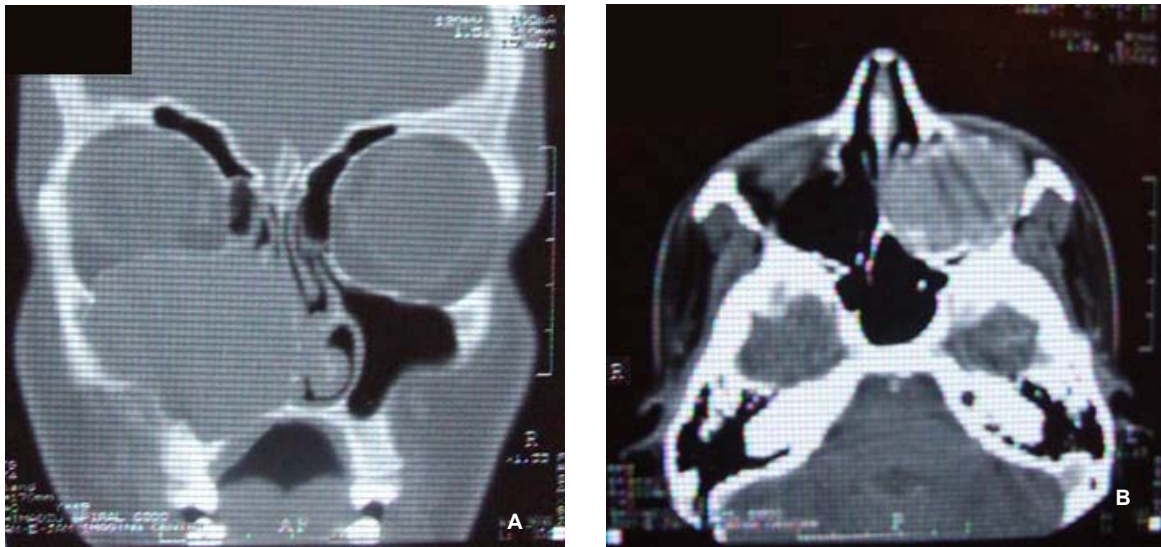


Figure 1: A. Coronal and B. Axial facial CT scan. A large mass has filled and expanded the left maxillary sinus.

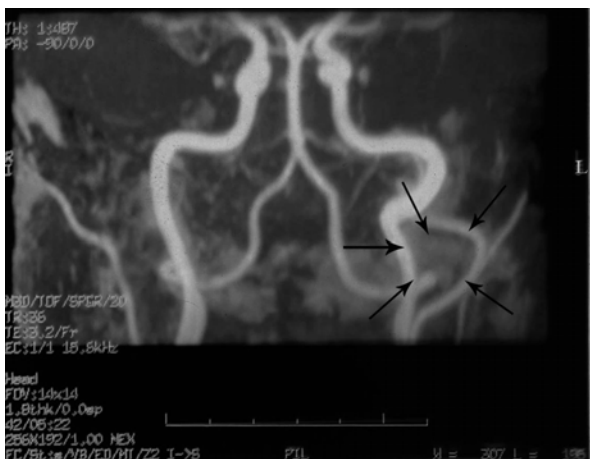


Figure 2: Magnetic Resistance Angiography of the mass inside left maxillary sinus. The arrows show a vascular mass supplied by internal maxillary artery.

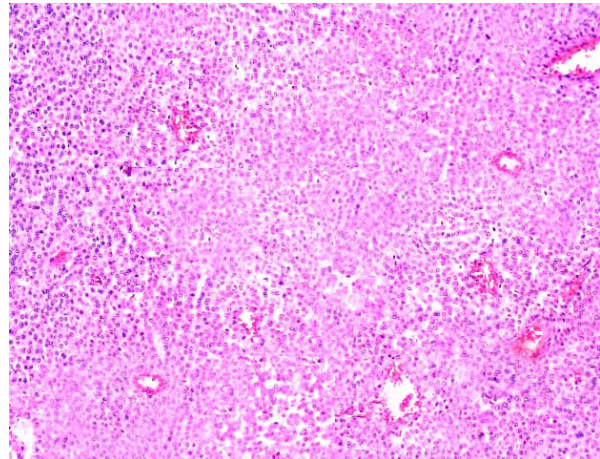


Figure 3: Hematoxylin-Eosin stained (x1000) microscopic picture of the mass showing tightly packed proliferated spindle cells and several vascular structures, compatible with the diagnosis of hemangiopericytoma

Discussion

The origin of HPC is the Zimmerman's capillary pericytes, which are commonly found in pelvic cavity, retroperitoneal area and lower extremities. Approximately 15% of all HPCs occur in the head and neck regions. Thompson and colleagues studied the clinical and histopathologic presentation of head and neck HPCs, and stated that the tumor was different from the ones arising from other sites of body, so they were considered a distinct entity.⁴

Hemangiopericytoma is seen in all age groups, but they are more common in the third to fifth decades of life. The tumor has no known risk factors such as sex, race or hereditary. Moreover, the etiopathogenesis of the tumors is not known well.⁵

Sinonasal HPC is commonly presented with nasal obstruction and/or epistaxis. In advanced stages, the patient may develop local pain, headache and visual disturbances. In physical examination, the mass may be seen as a tan, gray or white unilateral mass within the nasal fossa that can be mistaken for a nasal polyp. In palpation the mass may be rubbery, firm or soft.⁵ In imaging, especially contrast enhanced CT scan, the mass is frequently seen as a unilateral soft tissue density that remarkably enhances after the administration of intravenous contrast media.^{5,6}

The existence of any lesion that is presented as a unilateral mass in the nasal fossa and paranasal sinuses is important in the differential diagnosis of HPC, especially the lesions that have similar clinical and imaging

characteristics. Some of such lesions include nasal polyps, nasal and sinuses tumors such as inverted papilloma, angiofibroma, different malignancies and fibro osseous lesions.⁵

The diagnosis of HPC is based on histopathological findings, which demonstrate the tumor as tightly packed proliferated spindle cells surrounded by an intact reticulin sheaths. Their cytoplasm are sparse, and their nuclei are vesicular. The cells have ovoid to elongated nuclei. The arrangement of tumor cells produces interlacing bundles with many vessels intervening between them occasionally interconnected with stag horn pattern (figure 3).⁵⁻⁷

The appropriate treatment for sinonasal HPC is surgical removal with adequate margins. Angiography and embolization can reduce intra-operative bleeding, especially in large tumors. Radiotherapy has not been considered as a useful modality, because these tumors are radioresistant.^{5,7} With this such an approach to treatment, the tumor has been reported to have a local recurrence rate of 17% and a survival rate of 88% over 5 years.⁴ However, a recurrence rate of 35–45% have also been reported.⁶⁻⁸

Even histologically benign looking tumors may metastasize, therefore, the natural history of the tumor is not predictable and discrimination between benign and malignant tumors is clinical rather than histological.^{4,8} Based on histopathological features of HPC from other regions of the body, it was suggested that nuclear atypia, high mitotic rate, increased cellularity and cellular pleomorphism, presence of necrosis and large tumor size might be associated with aggressive behavior, and might predict less favorable outcomes.⁹ However, it is not clear whether or not such a suggestion is applicable to sinonasal HPC.

Local recurrence and distant metastasis have been seen years after primary diagnosis. A metastatic chance of up to 15% has been reported.⁴ Hemtogenic metastasis involving lung, liver and bones have been described. They are commonly associated with a local recurrence.^{7,9} On the other hand, the lymphatic metastasis and involvement of regional lymph nodes have not been reported. Considering the likelihood of late occurrence of a local recurrence

or a distant metastasis, it is important to follow patients with sinonasal HPC for life.¹⁰

Conclusion

Sinonasal HPC is rare and difficult to diagnose. A complete pre-operative work-up, a perfect surgery and a careful post-operative follow-up is necessary to achieve best results.

Conflict of Interest: None declared

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