

Nasopharyngeal Hemangiopericytoma in a Sudanese child: A Literature Review and Case Report

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Hemangiopericytoma (HPC) is an unusual vascular tumor derived from pericytes, first described in 1942 by Stout and Murray^{1,2}. These cells can be found surrounding all capillaries and are thought to be immature smooth muscle derived from mesenchyme. Pericytes add structural support to the capillaries and have a contractile capability playing a role in capillary blood flow regulation, modulating both flux and permeability^{1,3,4}. Approximately 1% of all vasoformative neoplasms are HPC, with 15 to 25% presenting in the head and neck^{4,5}. Of those occurring in the head and neck, approximately 50% involve the paranasal sinuses and nasal cavity. Other sites of predilection are the soft tissues of the scalp, face, and neck, parotid gland, orbit and rarely the salivary glands and larynx. There is no gender predilection and the age of incidence is usually between 50 to 70 years in 90% of cases with only 10% occurring in children. Since HPC is derived from capillary pericytes of Zimmerman`s, its theoretical site of origin can be virtually anywhere in the body. Indeed, HPC has been reported in a wide variety of tissues outside the head and neck, most frequently occurring in skeletal muscle and the retroperitoneum^{4,6}. The site of lesion has a significant bearing on clinical behavior and perhaps on histological character as well.

The gross picture of HPC is always a solitary solid mass with smooth surface and color ranging from grayish white to reddish brown. Areas of hemorrhage, necrosis and cystic degeneration are common. It may or may not be encapsulated.

Sinonasal HPC usually consists of well-differentiated tumors with low potential for local recurrence or metastasis. HPC usually evolves with slow and non-painful growth that progresses towards nasal obstruction and epistaxis, which are the most common symptoms^{3,7}. When this tumor only invades the nasal fossa and paranasal sinus, the prognosis is more favorable than in the case of meningeal or other patterns of invasion. The treatment of choice is surgical resection. Regarding etiology, a past history of trauma, prolonged steroid use and hypertension are

said to have some correlation, but such correlations have not been formally demonstrated⁸.

This is a case report and a literature review of a nasopharyngeal hemangiopericytoma in a Sudanese girl who presented to the Department of ENT/H&N Surgery at Wad-Medani teaching hospital. This is the first case, to our knowledge, to be reported in the Arab region and the Sudan.

Case Report

A 12 years old female student presented to the ENT referral clinic, Wad-Medani teaching hospital, on the 2nd of February 2007. Her main complaint was nasal blockage and recurrent nose bleeds for three months. The condition started by progressive painless nasal obstruction, snoring and mouth breathing. The condition was not associated with any other systemic manifestations or family history of a similar condition.

On Examination she looked ill, afebrile, pale and not cyanosed or jaundiced. Anterior and posterior rhinoscopy revealed a

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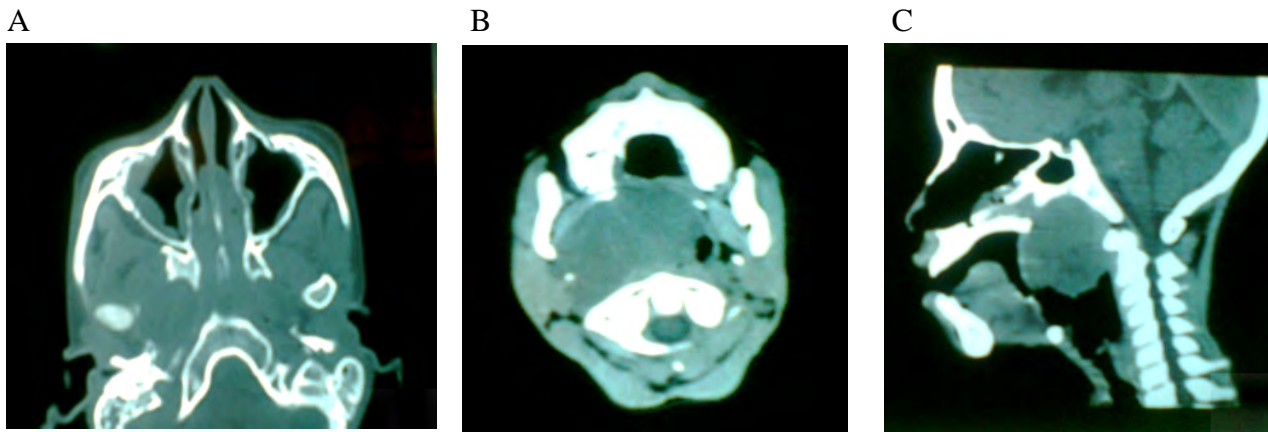
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mass completely occluding the nasopharynx. The soft palate was obviously pushed forward & downwards and no other abnormality was detected. Otoscopy showed normal right tympanic membrane with positive Rinnie's test and a retracted left tympanic membrane with negative Rinnie's test. No abnormality was detected on indirect laryngoscopy or neck examination. No abnormality was detected on systemic examination including the central nervous system.

Complete blood count showed normocytic normochromic RBCs, neutrophilia with moderate toxic granulation and reactive lymphocytes, slightly increased

platelets with few giant forms, and ESR of 80mm\1st hour. The specific gravity of her urine was 1.030 and microscopy revealed three crosses of urate crystals.

C-T scan non contrast axial, coronal and sagittal sections of the *paranasal sinuses, nasopharynx & oropharynx* showed a 7.5 cm. x 5.7 cm. oro-nasopharyngeal soft tissue mass severely compromising the oropharyngeal airway and completely obstructing the nasal cavity. There was also thickening of the mucous membrane lining the paranasal sinuses, but no evidence of bony destruction or erosion. (Figures 1: A, B and C).



Figures 1: *C-T scan* non contrast axial (A & B) and sagittal (C) sections of the *paranasal sinuses, nasopharynx & oropharynx* showed a huge oro-nasopharyngeal soft tissue mass severely compromising the oropharyngeal airway and completely obstructing the nasal cavity.

Examination under anesthesia revealed a large friable, spongy, polypoidal mass filling the oropharynx and nasopharynx with wide base attached to the supero-posterior wall of the nasopharynx. The mass was removed by transoral digital inoculation with only mild bleeding which was controlled by pressure packing. The tumour base was curetted by adenoid curette to remove any remaining tissues. Anterior and posterior nasal packs were applied. The mass specimen was sent for histopathology which was reported as aggregates of fragmented biopsy showing prominent stag horn vascular channels and fanning out diffuse sheets of bland spindle cells. Features were keeping with

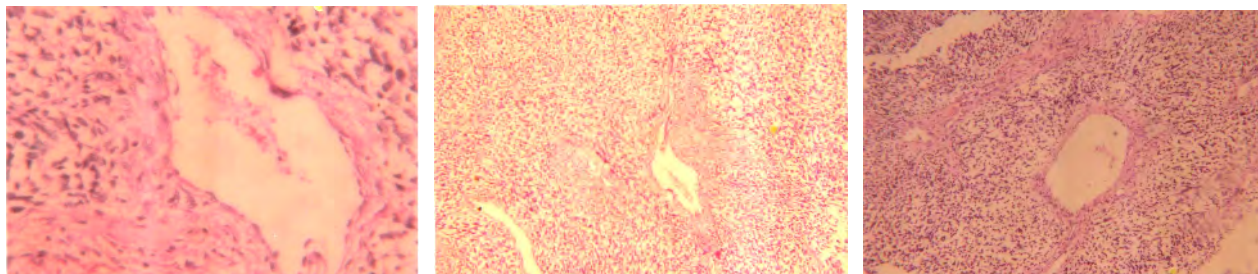
Hemangiopericytoma of the nasal cavity. Margins could not be assessed in a fragmented specimen. (Figures 2: A, B and C).

Discussion

Despite that 90% HPC cases arise in the age group 50-70 years; this case represents a young girl presenting with HPC in an unusual age. The sinonasal tract is the commonest (15 to 30%) site of origin in the head and neck region^{9, 10}. In this case the site of origin was the nasopharynx. It showed typical manifestations of a slowly growing, painless mass presenting with nasal obstruction and epistaxis. Although there was no cervical lymphadenopathy at presentation, the

unilateral conductive hearing loss and soft palate paralysis raised the suspicion of a nasopharyngeal tumor necessitating biopsy. Moreover, in this case the tumour had invaded beyond the nasopharyngeal cavity

and paranasal sinus, expanding and compressing the soft tissue of the oropharynx. This picture usually carries an unfavorable prognosis.



A: Hematoxylin & eosin stain; original magnification x40.

B: Hematoxylin & eosin stain; original magnification x10.

C: Hematoxylin & eosin stain; original magnification x10.

Figure 2: A, B and C: The mass specimen histopathology showing prominent stag horn vascular channels and fanning out diffuse sheets of bland spindle cells; features keeping with Hemangiopericytoma.

The case was managed by surgical excision followed by radiotherapy. This is the treatment of choice for an unresectable tumor or a tumour with high-grade histological findings or recurrent tumour^{9, 10}. However, radiotherapy is the only adjuvant therapy (HPC considered relatively resistant to radiotherapy). The HPC has a recurrence rate of 18% and a metastatic rate of 2.5% mainly to the lung and liver and about 3% of patients die of the disease. Unfortunately our case showed all these features.

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