Malignant Paraganglioma of The Nose

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Citation

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Abstract

Paragangliomas of the head and neck are uncommon tumors in head and neck area. The most frequent form is the carotid body tumor, while paragangliomas of the nasal cavity, paranasal sinuses and nasopharynx are rare especially malignant ones.

INTRODUCTION

Paraganglioma are uncommon tumors of head and neck and that usually arise from carotid bodies, jugulotympanic ganglia, ganglion nodosum of the vagus, and microscopic paraganglia aggregates dispersed in the mouth, nose, nasopharynx, larynx, and orbit. These are generally benign, slow growing tumors arising from neural crest cells in association with segmental and anatomic ganglion. Paragangliomas arising in the sinonasal tract especially the malignant are rare.

CASE REPORT

A 24-year-old male reported to ENT OPD with bilateral nasal obstruction, nasal deformity and left sided proptosis of 5 months duration. He had undergone excision of similar nasal mass 6 months back. Pathology at that time revealed the tumor to be a hemangiopericytoma. The patient was well postoperative only for a month after which he developed above complaints. Physical examination showed a soft tissue mass in both the nasal cavities with widening of nasal bridge and left-sided proptosis (Fig 1). In the oral cavity there was a bulge in middle of the hard palate. He had no cervical lymphadenopathy or cranial nerve deficits and vision was normal. Computerized tomography of the paranasal sinus revealed soft tissue mass involving bilateral nasal cavities, maxillary, ethmoid and sphenoid sinuses with erosion of septum and hard palate (Fig 1 and 2).

Figure 1

Figure 1: Clinical photograph of patient showing nasal mass.

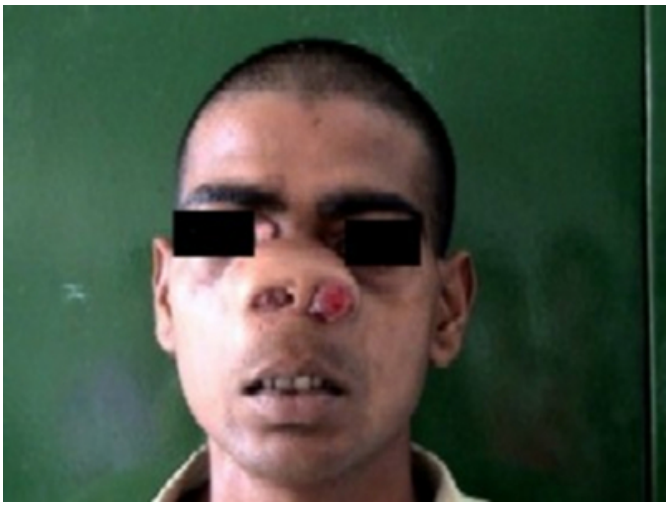


Figure 2

Figure 2: CT scan axial cut

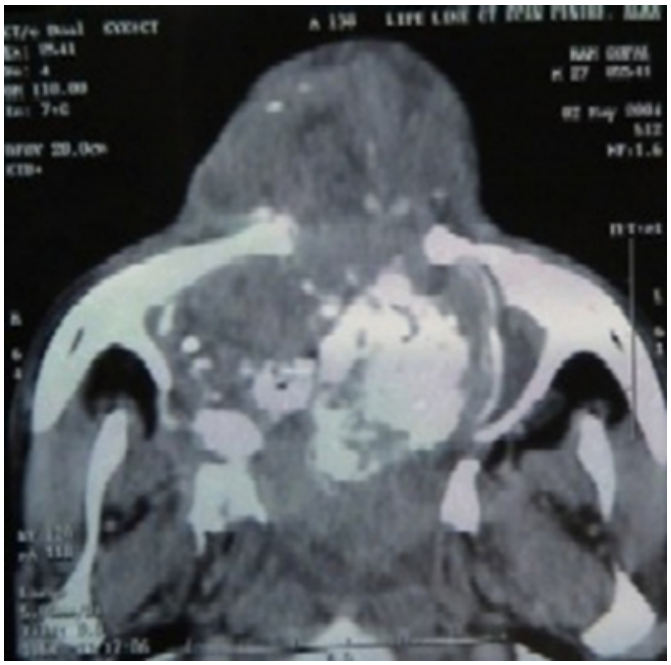
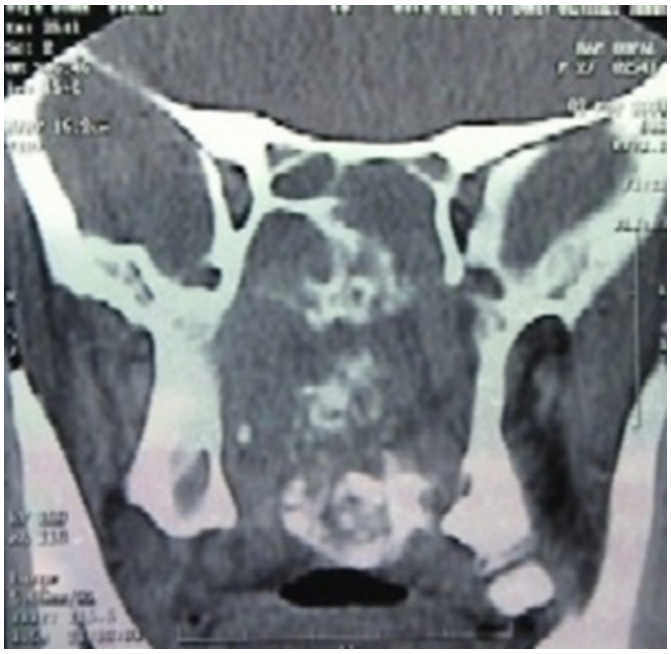


Figure 3

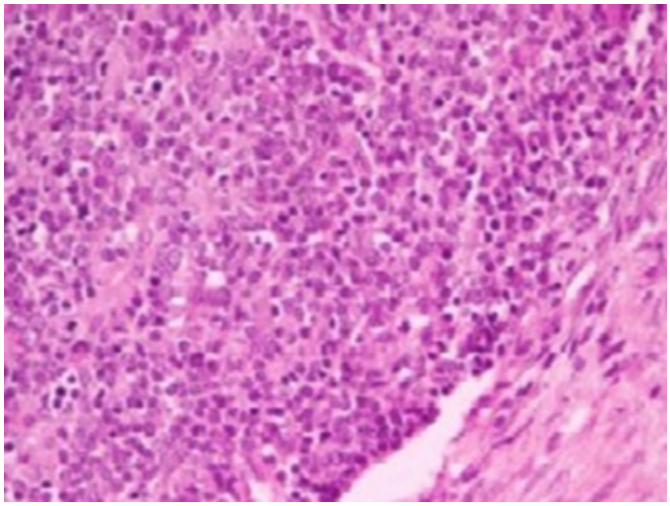
Figure 3: CT scan coronal cut



Patient was taken up for bilateral medial maxillectomy with excision of palatal mass. Histological examination of the mass showed it to be malignant paraganglioma. Section showed a cellular tumour composed of cells present in small sheets and characteristic Zellbellan pattern. Cells had moderate amount of eosinophilic cytoplasm and round nucleus with mild pleomorphism (Fig 4).

Figure 4

Figure 4: Histopathology of the nasal mass showing cells with moderate eosinophilic cytoplasm and round nucleus with mild pleomorphism. (H&E 400X).



There were focal areas of necrosis and high mitosis. Immunihistochemistry revealed Neuron specific enolase positivity in tumour cells. S-100 positive sustantecular cells were also found. Based on morphology, immunohistochemical findings and necrosis and increased mitosis a diagnosis of paraganglioma was made, possibly malignant. Patient received 30 grays of radiotherapy postoperative and is symptom free for last 8 months.

DISCUSSION

Paragangliomas arise from paraganglionic tissues of autonomic nervous system. Paraganglia are of neuroectodermal origin with cells containing catecholamines granules (type 1, chief cells) surrounded by Schwann like supporting cells (type 2, sustenacular cells)[8]. These cells have a widespread distribution through out the body and have been found in the lungs, heart, retroperitonium and urinary bladder. In head and neck the most common sites are carotid body, jugular bulb, along glossopharyngeal(especially its tympanic branch) and vagus(nodose ganglion) nerves[1,8]. These tumors account for 0.012% of all cancers of human body. Paragangliomas of nose and paranasal sinuses are very rare. The exact sites of origin of nasal paragangloimas are not definitely known. Several authors however have suggested that paraganglionic tissue is present in the pterygopalatine fossa[1,10] in agreement with the finding that paraganglia are usually in close association with arteries and cranial nerves[11]. However on review of the recent literature, most of the cases were described in the region of middle turbinate of ethmoid sinus.

Paragangliomas characteristically are composed of pale to clear staining cells with round nuclei forming well defined nests called Zellballen separated by a capillary network rich in reticulin. These cells contain cytoplasmic granules that have been demonsrated to be neurusecretory by electron microscopy[1,10,12]. Around 10% of cases may be malignant. Malignant parganglioma may exhibit several features that are not seen in their benign counterparts i.e. central necrosis of the Zellballen, invasion of the lymphatic and vascular spaces and the presence of mitotic figures[9]. Also some authors found that aggressive tumors contain fewer sustentacular cells than benign ones[14].

In a review of the head and neck paragangliomas approximately sixty percent were carotid body tumors, eighteen percent vagal body and eleven percent jugulotympanic[9] Very few cases of paragangilomas have been reported in the literature. Bolkov and Schecking reported the only malignant case that arose from ethmoid sinuses. Majority of these nasal tumors arise from the lateral nasal wall (turbinates). In our case also the tumor arose from the lateral nasal wall and then extended to the maxillary and ethmoid sinuses.

Treatment of these lesions, both benign and malignant, has consisted of surgical resection. Because of the location and nature of the tumor, they are difficult to resect and tend to recur locally. This had made the control of these lesions difficult and, for this reason, some authors have advocated full-dose radiation therapy(4500-6000rad).The results of radiation therapy have been variable, but it appears that radiation therapy slows tumor growth and rate of recurrence in most case[13] .However it does not completely destroy the lesion.

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