Haemangiopericytoma of the Oropharynx

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SUMMARY

Haemangiopericytoma (HPC) is a rare vascular tumor arising from the pericytes of Zimmermann and is characterized by its unpredictable and variable biological behavior. These tumors are common in the trunk and the extremities and less than 16% of the cases occur in the head & neck region[1, 2]. Its occurrence in the oral cavity and pharynx is uncommon. We report an extremely rare case of haemangiopericytoma of the oropharynx arising from the tonsillo-lingual sulcus as a pedunculated mass, which was successfully excised per-orally after a preliminary tracheostomy. No recurrence has been noted on a follow-up of more than six years.

KEY WORDS:
Haemangiopericytoma, Oropharynx, Histopathology

INTRODUCTION

Haemangiopericytomas are uncommon tumors of vascular origin formed by the proliferation of the pericytes of Zimmermann[3, 4]. The pericytes lie external to the endothelial cells of the capillaries and with its contractile properties they are capable of changing the size of the vascular lumen. As these cells are seen in all capillaries, haemangiopericytoma can occur in any part of the body. Most of the cases of haemangiopericytoma occur in the trunks and the lower extremities and only in 7 to 16% of the cases they are seen in the head and neck region[1, 2]. Majority of head and neck HPCs occur in the naso-sinus region. Rare cases of HPC arising in the oral cavity, skull base, infra-temporal fossa, parapharyngeal space, middle ear and orbit have been reported but its occurrence in the pharynx is extremely rare. We report one such case of HPC of the oropharynx presenting as a large pedunculated mass arising from the tonsillo-lingual sulcus.

CASE REPORT

A 34-year-old Chinese female presented to us in April 2001 with foreign body sensation in the throat of six months duration associated with dysphagia. She found her symptoms gradually worsening and developed a change in voice and noisy breathing especially during sleep. She noticed a mobile mass in her throat and eventually had mastered a technique to protrude the mass into the oral cavity. She gave no history of trauma, pain or blood stained saliva. She had a characteristic 'hot-potato voice' and clinical examination revealed a pedunculated, reddish, globular, smooth surfaced, mass of about 4 x 3 centimeters arising from the left tonsillo-lingual sulcus (Figure 1). The mass was soft in consistency, non-tender, non-pulsatile and did not bleed on touch. Indirect laryngoscopy could not be performed due to the sheer size of the mass. She had a mild inspiratory stridor and clinically no neck nodes were palpable. CT imaging showed a highly vascular tumor in the oropharynx with well-defined margins (Figure 2). With a provisional diagnosis of a benign vascular tumor, excision of the mass was contemplated. After a preliminary tracheostomy, the mass was exposed using a Doyen's mouth gag. Applying forward traction on the mass, the pedicle of the tumor was identified and clamps were applied on the pharyngeal mucosa around the pedicle with adequate tissue clearance. The operated site was ligated using 1.0 silk and there was virtually no blood loss. Post-operative period was un-eventful and she was decannulated on the third post-operative day. The biopsy was reported as haemangiopericytoma of the near aggressive type as it showed mitotic activity of up to 3 per 10 hpf and the resected margin was free of the tumor. Due to the near aggressive nature of the tumor a regular follow up post operatively was suggested by the pathologists. On her last follow-up, 6 years post-operatively, she was asymptomatic with no clinical evidence of recurrence.

DISCUSSION

Stout and Murray first described Hemangiopericytoma in 1942 while studying the glomus tumour. In 1949, they established HPC as a discrete pathological entity by reviewing all the published reports and adding 25 new cases. Majority of the HPC's occur in the trunk and the lower extremities and around 7 to 16% of them occur in the Head and neck region[1,2]. There are reports of HPC arising from the nasopharynx, maxilla, mandible, oral cavity, infratemporal fossa, orbit and the temporal bone. No case of HPC from the oropharynx has been reported as yet.

HPC is a vascular tumor arising from the pericytes of capillaries. Zimmermann[3] described the pericytes, as round or spindle shaped cells with long branching cytoplasmic processes that cover the outer wall of capillaries. These cells eventually transform into mature smooth muscle cells and are thus capable of regulating the size of the vascular lumen. As pericytes are present in the capillaries, HPC can occur anywhere in the body. The sub mucosal location and the intraluminal growth of these vascular tumors make them prone for injury resulting in superficial ulceration and hemorrhage.

These tumors are unpredictable neoplasms with some tumors exhibiting rapid growth while the others exhibit minimal changes over years of observation. McMaster[2] categorized NHC histologically into Benign (low-grade), Borderline

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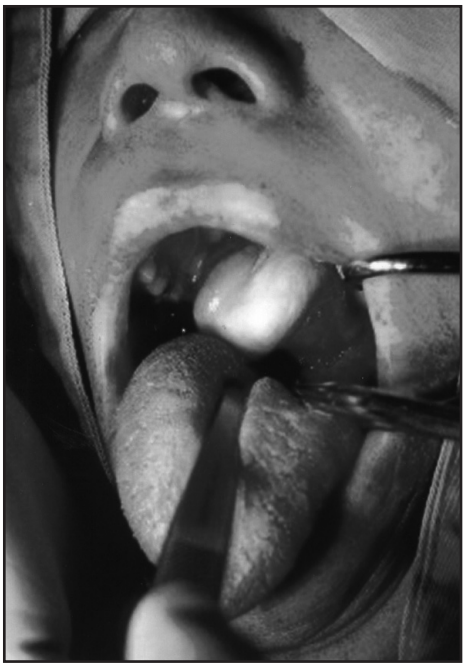

Fig. 1: Mass of about 4X3 centimeters arising from the left tonsillo-lingual sulcus

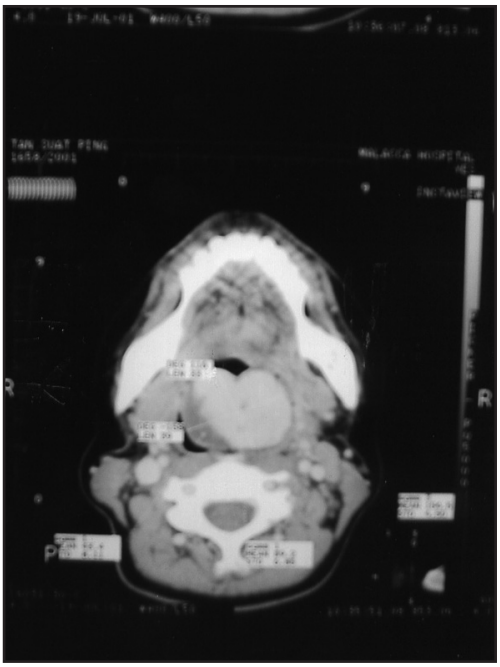

Fig. 2: CT imaging showed a highly vascular tumor in the oropharynx with well-defined margins

(Intermediate) or Malignant (High Grade). They predicted tumors to have a malignant property if they had a slight degree of cellular anaplasia or one mitotic figure per 10 high power field or having a moderate degree of cellular hyperplasia and one mitotic figure per 20 high power fields. Enzinger and Smith [1] emphasized that 4 or more mitotic figures per 10 high power fields were indicative of a rapidly growing malignancy capable of recurrence and metastasis. Pathological appearance of the resected HPC is therefore predictive of later metastatic potential. Modern radiological investigations like CT Scan, Ultrasound, Doppler sonography, arteriography can give information about the location and also the extent of tumor invasion into the surrounding tissue. Most of the authors advocate Surgery as the mainstay of treatment. The role of radiotherapy and chemotherapy as adjuvant treatment is controversial in the malignant form of hemangiopericytomas. However, radiotherapy may have a role in cases of incomplete removal of malignant hemangiopericytomas. A benign form of hemangiopericytoma does not exclude the possibility of a recurrence. These tumors have been known to recur years and even decades later.

Management of airway during and after surgery in these situations of oropharyngeal tumor is challenging. The airway can be secured various ways normally like orotracheal intubation, laryngeal mask, fibreoptic intubation and tracheostomy. In our patient CT imaging showed a highly vascular tumor in the oropharynx with well-defined margins. Indirect laryngoscopy could not be performed due to the sheer size of the mass. She had a mild inspiratory stridor. Further more the fear of traumatic bleeding and possible aspiration we have not attempted orotracheal intubation or fibreoptic intubation. Hence preliminary tracheostomy was performed.

Hemangiopericytoma of the oropharynx is extremely rare. Wide surgical excision is the treatment of choice. Long-term follow up is required in view of the high incidence of local recurrence and the very late appearance of metastasis.

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