CASE REPORT

Haemangiopericytoma—base of tongue

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Summary Haemangiopericytoma is a rare vascular neoplasm arising from pericytes in the walls of the capillaries and venules. This tumor is reported to occur mostly in lower extremity and retro peritoneum with rare occurrence in the head and neck region. Generally they are slow growing, locally infiltrative and aggressive, with various rates of pleomorphism and malignant potential. Clinical presentations of these tumors are characteristically non-specific and diagnostic modalities are of little help. Surgery remains the gold standard of the treatment with additional benefits if adjuvant RT is taken. Here we report a case of haemangiopericytoma arising in the base of the tongue with a brief review of the literature. To our knowledge this is the first case in this location to be reported in the English literature. © 2005 Published by Elsevier Ltd.

KEYWORDS
Oral cancer; Haemangiopericytoma; Base of tongue tumor; Soft tissue sarcoma

Introduction

Haemangiopericytoma (HPC) is an uncommon neoplasm first described by Stout and Murray in 1942. It arises from pericytes1–7 which are slender elongated cells located in the walls of capillaries and venules. These cells are external to the endothelial cells and are separated by basement membrane. They are considered undifferentiated cells that can differentiate into other cell types including smooth muscles.3,4 This tumor has been reported to occur mostly in the lower extremity and retro peritoneum.1–7 About 15–20% of cases have been reported in the head and neck region8 including orbit, nasal cavity, paranasal sinuses, nasopharynx, buccal mucosa, lip, floor of the mouth, gum, mandible, salivary gland and larynx.2,3,5,9–12

Here we are presenting the first case to be reported of haemangiopericytoma of base tongue in
a young adult male. In our review of literature we found no reference regarding haemangiopericytoma of base tongue in adults.

Case report

A healthy male aged 32 years presented with complaints of throat pain, change in voice and gradually increasing dysphagia. Past history was not suggestive of any previous medical illness or surgery. On examination there was no palpable mass in the neck and no cervical lymphadenopathy. All blood investigations were within normal limits, the chest skiagram was normal. Barium swallow showed mass in the oropharynx (Fig. 1). On indirect laryngoscopic examination there was an exophytic friable, fleshy growth in front of the epiglottis. There was no clinical or radiological evidence of distant metastases from the tumor. On direct laryngoscopic examination under general anaesthesia there was a large exophytic friable vascular growth arising from the central part of the BOT (base of tongue) rest of oropharynx, hypopharynx and glottis were normal. Histological picture showed sheets of spindle cells having scanty cytoplasm and plump nucleus. Tumor also showed prominent vessels and mitoses seen were 3–4/10 HPF. Overall the picture was of spindle cell sarcoma highly suggestive of haemangiopericytoma.

Subsequently for management the patient underwent wide excision of BOT tumor by transhyoid pharyngotomy through vallecula, by a transverse neck incision at the level of hyoid bone under GA with nasotracheal intubation (Fig. 2). There was a pedunculated exophytic friable mass arising from BOT measuring 6 cm x 5 cm. Wide excision of growth was carried out with margin of 1 cm of normal BOT. Margins of resections were confirmed by intra operative frozen section and were found to be negative for tumor cells. Resulting defect in BOT was closed primarily by absorbable sutures. Patient had uneventful post-operative recovery and was discharged on the 10th day.

On gross examination of the specimen tumor was 6 cm x 5 cm x 1.5 cm in size, cut surface was white, firm and some areas showed papillary appearances and scattered areas of necrosis and haemorrhage. Microscopic examination showed spindle to oval shaped cells with scanty cytoplasm and plump nucleus (Fig. 3). Tumor showed extensive staghorn vascular pattern (Fig. 4), mitoses were 4–5/10 HPF. There were focal myxoid changes and haemorrhagic areas. PAS and reticulin staining showed prominent vascular pattern. Cells were negative for Manson Trichrom stain. On immunohistochemistry tumor cells showed vimentin and actin positivity and were negative for Factor VIII.
The patient is on regular follow up with no evidence of recurrence.

Discussion

Haemangiopericytoma is an uncommon mesenchymal vascular neoplasm that is reported with rare occurrence in head and neck region. Nasal cavity and paranasal sinuses are the most common sites of presentation while oropharynx remains the rarest. This is a tumor of unknown etiology although various factors like previous trauma, steroid use and genetic factors have been suggested to cause this.

HPC may occur at any age, more commonly in 4th or 6th decade with no racial or sexual preponderance. Clinical presentation of HPC are characteristically non-specific. It mostly presents as slow growing painless mass associated with only local symptoms. Pain usually occurs in advanced cases and most often caused by pressure over neurovascular structures. Hypoglycemia has been reported in few cases of HPC in which blood sugar level came back to normal after resection of tumor.

Rontegenographic features are not specific for HPC and consists of radiopaque soft tissue mass displacing neighbouring structures. Focal calcification has been also reported. Daniels et al., after reviewing radiological literature evaluating HPC from many anatomic sites reported CT, MRI and angiographic features of HPC. In cases of HPC, consistent CT findings were hypervascularity with enhancement and well circumscribed margins. Both of these findings are non-specific. MRI findings are of a well circumscribed vascular tumor suggestive of HPC and further clarifies the boundaries and plane of tumor extension. Angiography shows a characteristic filling showing a dense well circumscribed stain secondary to accumulation of contrast in tumor capillary network. Overall angiographic picture of HPC is of a richly vascular tumor with dilation of the regional arteries in the arterial phase, diffuse capillary blush and dilation of draining veins in the late capillary phase. Angiogram does not differentiate HPC from other richly vascular soft tissue neoplasm but helps in narrowing down the diagnostic possibilities. According to one report absence of early venous filling in HPC is highly noteworthy and it differentiates HPC from other malignant soft tissue and benign vascular tumors. Diagnostic imaging studies are employed in cases of HPC for anatomic delineation, narrowing down the list of most probable diagnoses, defining the local extent as well as relationship to vital structures and identification of distant metastasis. MRI and angiographic findings are therefore useful in planning and execution of effective surgical approaches.

HPC has a uniquely variable range of malignant potential. Tumor may be classified as ‘benign’, 'borderline malignant' and 'malignant’ on the basis of histological findings. An association has been suggested between hypercellularity, mitotic activity anaplasia, necrosis and hemorrhage with more malignant HPC. Enxinger and coworker have reported that tumor having 0–4 mitoses per 10 HPF have 77% 10 year survival while tumors having >4 mitoses per 10 HPF have 29% 10 year survival. They have also reported that tumors having <6.5 cm diameter have 92% 10 year survival.

HPC have high rate of local recurrence and distant metastases to lung and skeleton as most frequent sites. Enxinger and coworker have considered lymphnode metastases to be rare but others have reported metastases to lymphnode also. Metastatic rates for borderline and malignant variety has been reported to be 45%. Blackwinkle and Diddans in comprehensive review of 224 cases reported that haemangiopericytoma is highly malignant over a life time in about 50% of cases. Latency period of HPC recurrence have been reported upto 16 years to 33 years, although peak incidence of recurrence is between 1 and 5 years and varies with particular organ system involved. They have reported recurrence rate of 80% in tumors of CNS and 50.5% for tumors of musculoskeletal system.

As this tumor is not of a common occurrence with the biological behaviour quite unpredictable,
management of this tumor has been a subject of debate. Surgery is considered the standard therapy whenever tumor is technically resectable. Consensus in the literature is to treat all resectable HPC with wide excision.1–6,10,14,15,17,18 Local recurrence and solitary metastasis have also been widely excised with prolonged survival.14 Pre-operative tumor embolization has been suggested to be a useful procedure having value in helping to reduce blood loss during surgery which allows aggressive resection of large tumors especially in retroperitoneum and visceral sites.3,14,15

Initial report regarding use of RT as primary or adjuvant therapy were not very encouraging however, recent reports have documented tumor response to doses >45 Gy2–4,14,18 thereby suggesting RT is useful in unresectable or recurrent tumor for purpose of palliation.1,17,18 Mira et al. have suggested that bulky resectable tumors should be excised whenever possible prior to radiation therapy.18 Chemotherapy has not been found to be useful as adjuvant or primary therapy.1–3,14 Partial or short term remission of metastatic tumor have been reported with doxorubicin as single agent or in combination therapy.2,3,14,19 However, long term remission or survival is not reported.

At present, surgery i.e. wide excision is the standard treatment. Additional benefits may be gained by the use of adjuvant RT. Treatment outcome however depends on tumor size, the number of mitotic figures per 10 HPF and the histological grade with newer technologies coming up, early detection and effective treatment options have now become possible. However, close regular follow up of the patients is required for long duration for early detection and effective management of recurrence.

References