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Hemangiopericytoma Of The Larynx In Childhood : A Rare Clinical Entity

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ABSTRACT Hemangiopericytoma are rare vascular neoplasms of head and neck. Laryngeal involvement is even extremely rare, with only 12 previously reported cases in the literature. Ours is the first case in the literature and in India to be present in childhood. We present with an unusual case of a 6 years old boy with tracheostomy done at GMCH for the stridor and diagnosed as case of supraglottic mass with glottic extension. Biopsy and Immunohistochemistry was done which was Vimentin, CD34, CD31 positive s/o malignant hemangiopericytoma. After full clinic-radiological evaluation and discussion with multidisciplinary team, in a view of locally advanced lesion preoperative EBRT was given, but disease progressed after EBRT. Hence total laryngectomy with permanent tracheostomy was done with HPE report consistent with a malignant haemangiopericytoma. Patient is currently 9 months post opt follow-up doing well with no recurrence. Otolaryngologists and surgical oncologist need to be aware of this rare tumor that can be treated successfully with surgical resection. Close and long term follow up is needed.

KEYWORDS : Hemangiopericytoma, Supraglottis and Glottic mass,Immunohistochemistry External beam radiotherapy(EBRT), Total Laryngectomy.

Introduction

Haemangiopericytomas are rare vascular neoplasm's of the head and neck, representing 1.3% of vascular tumours.[1] They arise from the pericytes of the Zimmerman, which are pericapillary spindle cells that provide mechanical support and regulate luminal diameter. [2] Since first described by Stout and Murray in 1942 ,there are only approximately 200 cases of haemangiopericytomas described in the literature.[3] 15-25% of haemangiopericytomas arise in the head and neck, with only 12 previously reported cases in the larynx.[1,4–7,12] Haemangiopericytomas have a propensity for local recurrence, unpredictable behavior, and the potential for distant metastasis through haematogenous spread and hence the tumor must be extirpated as radically as possible while protecting the organ system.[8,9] As it is a very rare tumor, the clinical course, treatment, outcomes and prognosis have not well delineated. We describe an unusual case of supraglottic and glottic haemangiopericytoma managed successfully with complete surgical excision. We will review our case regarding clinical features and management and a review of the literature.

CASE REPORT

A 6 years old boy presented to our OPD with tracheostomy done in government hospital for severe stridor with no significant past medical history. Patient was referred for further management at our institute. In our institute we have evaluated the patient with, CT scan of neck which was s/o well defined slightly enhancing heterogeneous soft tissue mass (42mm x 32mm x 43mm) involving supraglottis with involvement of b/l AE folds and obliterates both pyriform fossa, lesion infiltrates the both lobes of thyroid gland with significant luminal narrowing.(fig 1) Direct laryngoscopic examination was done which was s/o sub-mucosal mass lesion involving supraglottis ,b/l AE fold ,b/l PF with post cricoid extension, posterior pharyngeal wall normal. Biopsy and IHC confirmed it to be malignant hemangiopericytoma. After clinic-radiological evaluation and discussion with multidisciplinary team, in a view of locally advanced disease radiotherapy was advised. EBRT 30 Gy in 15 fraction was given from 8/1/16 and concluded on 2/2/2016. Post radiotherapy CT neck was done s/o increase in size of mass with involvement of b/l AE fold, b/l pyriform fossa, b/l false and true cord, lesion extends into post cricoids region, b/l thyroid lobe involvement with suspicious erosion of thyroid cartilage, s/o no response to radiotherapy, hence decision for surgical treatment was taken. Total laryngectomy with permanent tracheostomy done (figure 2). Pathology confirmed a 5.0×4.5×2.5 cm growth involving post cricoid region with infiltration into left lobe of thyroid gland with negative all surgical margins. No mitosis was noted. Specimen IHC was positive for Vimentin, CD34, CD31(positive in blood vessel),CD 99(cytoplasmic positivity), consistent with malignant hemangiopericytoma(fig .3).

DISCUSSION

Hemangiopericytomas are considered vascular tumors with variable malignant potential, manifested clinically by distant metastases, typically to the lung, liver and bony skeleton. [7] While there are low rates of regional recurrence due to the hematogenous spread of hemangiopericytomas, the rates of local recurrence and distant metastases are significant. Local failure rates have been reported at 40%, while distant metastases occur in 30-33% of patients in most recent head and neck cases series. [2,8] Overall, the rate of distant metastases has been reported between 18 and 57% in the literature, and can occur up to 11 years after initial diagnosis and treatment. [8] Hemangiopericytomas can be classified as benign, borderline, and malignant based on histologic grade, with higher grade tumors correlating with higher rates of distant metastases and decreased survival.[7] In McMaster's case series, 6 of the 16 borderline tumors metastasized (37.5%), and 6 exhibited local recurrence after excision. Of the 32 malignant tumors, 25 (78%) developed distant metastases. In addition, long-term follow up was recommended because metastases developed in 11% of patients with malignant tumors and 7% with borderline tumors after a 5 year disease free interval. 10 The prognostic value of histologic findings was also corroborated by Enzinger's case series. Higher grade tumors with > 4 mitotic figures/ 10 high power field, presence of necrosis, and tumor size greater than 6.5 cm had poorer overall 10 year survival.[7] Despite these prognostic factors, the clinical outcome and optimal management of hemangiopericytomas of the larynx are still unknown due to the paucity of cases reported in the literature. [11] In addition, much of the above data is based on small case series. While sinonasal hemangiopericytoma has been well described, laryngeal involvement is much more rare, with only 12 previously reported cases in the literature. [1,4–7,12] Most occurred in supraglottis. Eight of the 12 cases were treated with partial or complete surgical excision , ranging from partial supraglottic laryngectomy to total laryngectomy. While the mainstay of treatment for hemangiopericytomas is surgical excision, the indications for adjunctive treatment are unknown and controversial. In the largest most recent head and neck case series, 4 out of 12 patients received postoperative external beam radiation to a median dose of 60 Gy for positive surgical margins, high grade histology, or recurrent lesions. [2] While there are several reports of adjunctive radiation and chemotherapy in a few case series, there are no large scale studies looking at outcomes of postoperative adjunctive treatment. [3] Our patient was successfully treated with total laryngectomy. He is currently 9 months post-operative follow-

up without any evidence of disease, dysphonia or dysphagia.

CONCLUSION

Hemangiopericytoma is an extremely rare vascular neoplasm with a propensity for local recurrence, unpredictable behavior, and the potential for distant metastasis. Due to the paucity of laryngeal cases reported in the literature, the clinical outcome, prognosis, and indications for postoperative adjunctive treatment are unknown. Otolaryngologists need to be aware of this rare tumor that can be treated successfully with surgical resection. Close long-term follow up is needed since recurrence can present many years after initial treatment.

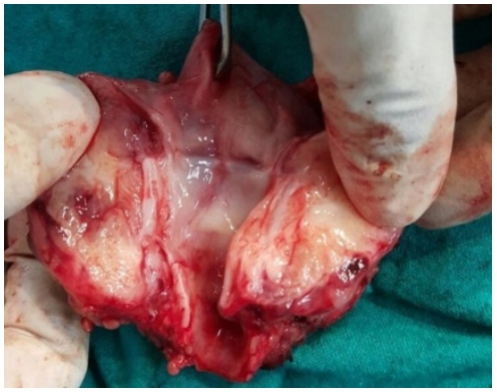
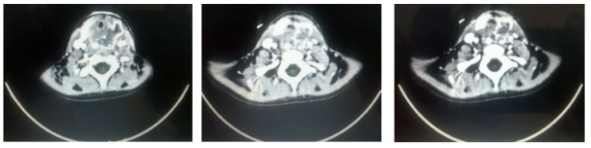


Fig. 2 specimen photo

Fig. 1



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