

Bangladesh J Otorhinolaryngol 2015; 21(1): 47-50

Case Report

Haemangiopericytoma of the Larynx: A Rare Clinical Entity

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Abstract

Haemangiopericytomas are rare vascular neoplasms of the head and neck. Laryngeal involvement is even extremely rare, with only 11 previously reported cases in the literature. We present an unusual case of a 45 years old man with obstructing left glottic mass. Tracheostomy was done for the stridor and FOL showed huge reddish vascular mass which was attached along the whole length of the undersurface of the left vocal cord. Microlaryngoscopic excision of the whole mass was done, with biopsy consistent with a haemangiopericytoma. Following operation tracheostomy tube was put off and he got his voice back. The patient is currently 2 years post op follow-up with no recurrence. Otolaryngologists need to be aware of this rare tumour that can be treated successfully with surgical resection. Close long term follow up is needed since recurrence can present many years after initial treatment.

Key words: Haemangiopericytoma, glottic mass;

Introduction

Haemangiopericytomas are rare vascular neoplasms 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 11 previously reported cases in the

larynx^{1,4-7} Haemangiopericytomas have a propensity for local recurrence, unpredictable behavior, and the potential for distant metastasis through haematogenous spread and hence the tumour must be extirpated as radically as possible while protecting the organ system.^{8,9} As it is a very rare tumour, the clinical course, treatment, outcomes and prognosis have not well delineated. We describe an unusual case of 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 45 year old man attended to the out patient department of Rajshahi Medical College Hospital with persistent hoarseness for 3 years and mild stridor for 15 days and eventually came to the hospital with severe stridor for 2 days. He did not have any sore

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throat or dysphagia. He had a direct laryngoscopy and biopsy done at the same hospital for hoarseness of voice 2 years back but did not have any diagnosis. Tracheostomy was done for the stridor (figure-1). The patient was uncomfortable for indirect laryngoscopic examination and fiberoptic laryngoscopy showed a huge reddish vascular mass which was attached along the whole length of the undersurface of the left vocal cord (figure-2). Microlaryngoscopic excision of the whole mass was done which was found to be a firm mass (figure-3 & 4). Biopsy was consistent with a haemangiopericytoma, an extremely rare form of laryngeal lesion (figure-5). Following operation tracheostomy tube was put off (figure-6) and he got his voice back. The patient is currently 2 years post- op with no evidence of disease and no dysphonia or dysphagia. After searching the internet it was recorded to be no 12 haemangiopericytoma arising in larynx and first reported case in Bangladesh.

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 11 previously reported cases in the literature.^{1,4-7} 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 complete excision of the glottic mass. He is currently 2 years 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.

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