Hemangiopericytoma of the larynx: a rare clinical entity

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Abstract

Hemangiopericytomas are rare vascular neoplasms of the head and neck. Laryngeal involvement is even more rare, with only 10 previously reported cases in the literature. We present an unusual case of a 46 year old man with an incidental finding of an obstructing submucosal supraglottic mass, with biopsy consistent with a hemangiopericytoma. The patient underwent preoperative angiobiolization, left partial pharyngectomy and supraglottic laryngectomy, and level IV lymph node excisional biopsy. Pathology confirmed a hemangiopericytoma with negative surgical margins, and lymph nodes negative for tumor. The patient is currently 9 months post-op with no evidence of disease, and no dysphonia or dysphagia.

Introduction

Hemangiopericytomas are rare vascular neoplasms of the head and neck, representing 1.3% of vascular tumors. They arise from the pericytes of Zimmerman, which are pericapillary spindle cells that provide mechanical support and regulate luminal diameter. Since first described by Stout and Murray in 1942, there are only approximately 200 cases of hemangiopericytomas described in the literature. 15-25% of hemangiopericytomas arise in the head and neck, with only 10 previously reported cases in the larynx. Hemangiopericytomas have a propensity for local recurrence, unpredictable behavior, and the potential for distant metastasis through hematogenous spread. Due to the rarity of this tumor, the clinical course, treatment outcomes, prognostic factors, and need for adjunctive treatment have not well delineated. We describe an unusual case of a supraglottic hemangiopericytoma managed successfully with a partial pharyngectomy and partial supraglottic laryngectomy through a lateral pharyngotomy. We will review the clinical features of hemangiopericytomas, as well as their management and a review of the literature.

Case Report

The patient is a 46 year old man with no significant past medical history who was incidentally found to have an obstructing submucosal supraglottic mass while being evaluated for complaints of nasal obstruction. The patient denied any dysphagia or obstructive symptoms, but did report a change in voice. Flexible fiberoptic laryngoscopy revealed an ovoid submucosal mass involving the left aryepiglottic (AE) fold, which was obstructing view of the glottis. The true vocal cords appeared mobile and uninvolved when the mass was bypassed with the laryngoscope, however the view was limited. Preoperative Computed Tomography (CT) of the neck noted a 3.7 x 2.6 x 4.3 cm contrast enhancing lesion of the left supraglottis and pyriform sinus with laryngeal deviation to the right and no evidence of cartilage erosion (Figure 1).

The patient was taken to the operating room for awake tracheostomy and direct laryngoscopy with biopsy, since awake fiberoptic intubation could not be performed due to the degree of supraglottic obstruction. A large submucosal mass was noted at the region of the left AE fold, with obliteration of the left pyriform sinus and no glottic involvement (Figure 2). Biopsy of this lesion resulted in extensive bleeding, which was controlled with Surgicel, Floseal, thrombin soaked pledges, and suction cautereization. Biopsy was consistent with a hemangiopericytoma. PET/CT noted an intensely FDG avid mass (SUV 8.2) involving the left epiglottis, AE fold, and lateral/posterior pharyngeal wall, with mild uptake in level IV lymph nodes (SUV 3.1), but no distant metastases (Figure 3).

The patient underwent preoperative angiobiolization of the feeding left superior thyroid artery, left partial pharyngectomy & supraglottic laryngectomy, and level IV lymph node excisional biopsy. The mass was exposed through a lateral pharyngotomy and was found to be centered in the left pyriform sinus with some involvement of the medial wall and AE fold, with an intact arytenoid cartilage. Blunt dissection of the well encapsulated tumor was then performed, leaving the pharyngeal mucosa intact, with minimal blood loss.

Pathology confirmed a 8.3 cm hemangiopericytoma with negative surgical margins, and lymph nodes negative for tumor. No mitosis was noted, and the tumor showed evidence of embolization with areas of necrosis. Hematoxylin and eosin (H&E) stain revealed the characteristic CD34+ pericytes around small to medium sized blood vessels, with reticulin stain highlighting pericytic collagen (Figure 4).

The patient is currently 9 months post-op with no evidence of disease, and no dysphonia or dysphagia.

Discussion

Hemangiopericytomas are considered vascular tumors with variable malignant potential, manifested clinically by distant metastasis, typically to the lung, liver and bony skeleton. While there are low rates of regional recurrence due to the hemogenous 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 case series. 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.

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. In McMasters 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 followup was recommended because metastases developed in 11% of patients with malignant tumors and 7% with borderline tumors after a 5 year disease free interval. 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.

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. In addition, much of the above data is based on small case series. While sinonasal hemangiopericytomas has been well described, laryngeal involvement is much more rare, with only 10 previously reported cases in the literature. Most occurred in the supraglottis. Seven of the 10 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 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. 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.

Our patient was successfully treated with an open left partial pharyngectomy and supraglottic laryngectomy through a lateral pharyngotomy for a low grade hemangiopericytoma. He is currently 9 months post-operative without any evidence of disease, dysphonia or dysphagia.

Conclusions

Hemangiopericytomas 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 followup is needed since recurrence can present many years after initial treatment.

References