ExTRANASOPHARYNGEAL ANGIOFIBROMA OF THE LARYNX

ABSTRACT

Objective: To present a rare case of laryngeal extranasopharyngeal angiofibroma, discussing its diagnosis, treatment and differences from the more typical juvenile angiofibroma.

Methods:

Design: Case Report
Setting: Tertiary Government Hospital
Patient: One

Results: A 51-year-old male with a two-year history of hoarseness developed difficulty of breathing. Direct laryngoscopy showed a 2x2x1cm glistening, multinodular, pedunculated, firm, pink mass attached to the posterior half of the right true vocal fold obstructing the glottic opening and extending superiorly to the ventricle. Microlaryngeal excision was done. Histopathology showing numerous vascular channels surrounded by dense paucicellular fibrous tissue was consistent with angiofibroma.

Conclusion: Primary extranasopharyngeal angiofibroma is rare, with only four previously reported cases occurring in the larynx. We presented what may possibly be the first locally reported case. Although histopathologically similar to the more common juvenile nasopharyngeal angiofibroma, this was atypically seen in the larynx of an older adult patient. Direct laryngoscopy provided excellent exposure for identification as well as complete surgical resection. Unlike the nasopharyngeal type, no massive bleeding was encountered. Prognosis for this extranasopharyngeal angiofibroma is excellent as recurrence is noted to be rare, however, long term follow-up is recommended.

Keywords: extranasopharyngeal angiofibroma, laryngeal angiofibroma

Angiofibromas typically occur in the nasopharynx. Although histologically benign, they show locally aggressive growth with bone destruction, extension to adjacent areas and possible fatal complications such as intracranial invasion or hemorrhage. Nasopharyngeal angiofibromas usually occur almost exclusively in young males and they originate at the junction of the posterolateral wall and roof of the nose at the superior margin of the sphenopalatine foramen, the approximate location of the embryological buccopharyngeal membrane and not in the nasopharynx as commonly thought. Head and neck angiofibromas arising from outside the nasopharynx are rare.
In a review of 65 cases of extranasopharyngeal angiofibromas (ENA), Windfuhr and Remmert found only four in the larynx. Clinical characteristics of ENAs do not conform with those of nasopharyngeal angiofibromas and, for this reason, these tumors must be regarded as a separate entity. A better understanding of this atypical entity obviates the need for unnecessary invasive procedures typically reserved for nasopharyngeal angiofibroma.

We present a case of extranasopharyngeal angiofibroma affecting the glottic region. Differences between ENA and the more typical nasopharyngeal angiofibroma will be discussed. Diagnosis and treatment of laryngeal angiofibroma will be elaborated upon.

**CASE REPORT**

A 51-year-old male was admitted due to progressive difficulty of breathing. He had been hoarse for two years, described as having a coarse, strained voice which gradually developed into a weak, strained voice. He was a 20-pack-year smoker and occasionally drank alcoholic beverages. Indirect laryngoscopy revealed a multinodular, non-necrotic, pink mass covering the true vocal folds and filling up the ventricles. Emergency tracheotomy was done.

Direct laryngoscopy showed a 2x2x1cm glistening, multinodular, pedunculated, firm, pink mass attached to the posterior half of the right true vocal fold obstructing the glottic introitus and extending superiorly to the ventricle (Figure 1). Microscopic evaluation revealed numerous vascular channels surrounded by dense paucicellular fibrous tissue (Figure 3). The cells in the fibrous tissue were cytologically bland and spindle shaped. The nuclei lacked hyperchromasia and had small nucleoli. The vascular channels were slit-like or dilated, and varied in number, configuration and thickness. Findings were consistent with angiofibroma.

While follow-up videolaryngoscopy at one week was essentially normal, a repeat three weeks post-operatively showed nodular scar tissue on the right vocal fold where the base of the excised tumor pedicle had been located (Figure 4).

**Discussion**

Primary extranasopharyngeal angiofibroma is very rare. The most common site for extranasopharyngeal angiofibroma is the maxillary sinus. The ethmoid and sphenoid sinuses, nasal septum, middle and inferior turbinate, conjunctiva, molar and retromolar region and larynx are other sites where extranasopharyngeal angiofibromas have been reported. Our patient may be the first locally reported case of laryngeal angiofibroma.

Angiofibroma presenting with at least one of the following criteria such as origin or location other than nasopharynx, presenting complaints other than nasal obstruction or epistaxis, age younger than seven or older than 25, female sex and multifocality are considered...
As benign tumors of the larynx are reported to be less than 1% of all laryngeal neoplasms, it is imperative to identify these tumors since treatment and prognosis vary greatly. Diagnostic modalities include computed tomography scan, magnetic resonance imaging and angiography. Angiobroma of the larynx is diagnosed with direct laryngoscopy and biopsy. While biopsy of nasopharyngeal angiofibromas may not be done, it should be done in cases of laryngeal tumors to ensure that malignancies will not be missed.

Surgical excision is the treatment of choice for angiofibromas. Angiofibromas have characteristic endothelial-lined vascular spaces, with little or no smooth muscle layers and are devoid of an internal elastic lamina, which preclude vasoconstriction and contribute to brisk episodes of bleeding when traumatized. Interestingly, the feeding arteries within the stalk of the tumor mass contain a complete muscle wall which has normal contractile ability and results in little bleeding after tumor excision from its pedicle. Selective angiography has been recommended for nasopharyngeal angiofibroma as it has the ability to identify the feeding vessels and allows the option of pre-operative embolization for vascular control. In our case, as the mass was relatively small and easily excised at its base, minimal bleeding was encountered. It appears that angiography may not be necessary in managing laryngeal angiofibroma.

The clinical presentation of our patient (Table 1) was virtually similar to four previously reported cases. In two of the four, endolaryngeal endoscopic or microendolaryngeal surgical excision was performed with successful results. The other two cases involved partial laryngopharyngectomy and transcervical resection, respectively.

Due to the confined space in the nasopharynx, inadequate surgical exposure and subsequent incomplete resection probably account for the higher recurrence rate in nasopharyngeal angiofibromas. Although tumor recurrence is rare for laryngeal angiofibromas and is not expected in our case, long-term follow-up is recommended.

Table 1. Comparison of Reported Cases of Laryngeal Extranasopharyngeal Angiofibroma

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age</th>
<th>Sex</th>
<th>Site</th>
<th>Symptom/s</th>
<th>Onset</th>
<th>Therapy</th>
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<tbody>
<tr>
<td>1</td>
<td>64</td>
<td>F</td>
<td>Left aryepiglottic area</td>
<td>Dysphagia and dyspnea</td>
<td>2 ½ months</td>
<td>Tracheostomy, partial laryngopharyngectomy</td>
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<tr>
<td>2</td>
<td>29</td>
<td>M</td>
<td>Right vocal fold, subglottic</td>
<td>Hoarseness</td>
<td>7 months</td>
<td>Microlaryngeal surgery</td>
</tr>
<tr>
<td>3</td>
<td>30</td>
<td>F</td>
<td>Interarytenoid region</td>
<td>Asymptomatic</td>
<td>On O RL exam</td>
<td>Tracheostomy, laryngofissure and microlaryngeal surgery</td>
</tr>
<tr>
<td>4</td>
<td>25</td>
<td>M</td>
<td>Larynx (site not specified), pharynx</td>
<td>Dysphagia and voice changes</td>
<td>6 months</td>
<td>Transcervical resection</td>
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</table>

<table>
<thead>
<tr>
<th>Case</th>
<th>Onset</th>
<th>Site</th>
<th>Symptom/s</th>
<th>Therapy</th>
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<tbody>
<tr>
<td>Our Case</td>
<td>2 years</td>
<td>Right vocal fold</td>
<td>Hoarseness and dyspnea</td>
<td>Tracheostomy, microlaryngeal surgery</td>
</tr>
</tbody>
</table>

REFERENCES