Laryngeal Amyloidosis: A Rare Entity causing Hoarseness of Voice

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ABSTRACT

A 65-year-old man with persistent hoarseness of voice of 1-year duration was found to have a lesion in both vocal cords based on flexible laryngoscopy. He was diagnosed as having amyloidosis of the glottis on biopsy. He did not have any associated comorbidities. He underwent laser excision of the lesion following which his voice improved. There were no signs of recurrence during the 6-month follow-up period.

Keywords: Amyloidosis, Larynx, Laser excision, Laser surgery, Phonosurgery.

INTRODUCTION

Amyloidosis is the extracellular deposition of the fibrous protein amyloid in one or more body sites. Amyloidosis may broadly be classified as either primary or secondary. Primary amyloidosis is idiopathic, whereas the secondary form is associated with a chronic inflammatory or infectious process.1 Localized amyloidosis of the head and neck is a rare disease, and the larynx is the most frequent site of involvement.1 Of all the benign tumors of the larynx, amyloidosis accounts for only about 1% of such lesions.2 Diagnosis is made after excluding evidence of systemic amyloidosis and systemic diseases associated with secondary amyloidosis.1,3 Compared with other sites in the head and neck region, amyloidosis of the larynx is unlikely to be related to a systemic illness.4,5 In the larynx, the ventricles are most frequently involved, followed by the subglottis, the aryepiglottic folds, and the true vocal cords. Localized amyloid has been found in other head and neck sites, including the nasopharynx, salivary glands, paranasal sinuses, nose, eye, oral cavity, oropharynx, and tracheobronchial tree.6 Patients typically present with hoarseness of voice, although they may present with cough, globus, hemoptysis, stridor, or dyspnea if the lesion crosses the midline.

Microscopic examination typically shows a diffuse submucosal globular deposition of a largely acellular eosinophilic material that exhibits apple-green birefringence under polarized light when stained with Congo red. There may also be an associated sparse, mixed chronic inflammatory infiltrate consisting of mature plasma cells and lymphocytes.5

In this article, we present a case of a 65-year-old male who was diagnosed with and treated for laryngeal amyloidosis along with the review of literature.

CASE REPORT

A 65-year-old man presented with progressive change in voice since 1 year. He did not give any history of difficulty breathing or swallowing, throat pain, or referred otalgia. He did not complaint of any swelling in the neck. There was no similar history prior to this, and there was no history of previous surgery. He was on oral hypoglycemics for type 2 diabetes mellitus for the last 10 years.

Examination of the oral cavity and neck did not show any remarkable findings. Indirect laryngoscopy showed a raised irregular mass on the left false and true cord more toward the midline. Anterior and inferior extent could not be identified. Airway appeared adequate.

A computerized tomography scan with contrast was done of the neck, which showed a nodular enhancing lesion involving the left true and false cords, paraglottic fat, and laryngeal ventricle (Fig. 1).

A flexible laryngoscopy was done which showed an irregular lesion involving the left false cord, left ventricle, superior surface of left true cord, anterior commissure, and extending to the anterior one-third of right false cord. Subglottis appeared normal, and airway appeared adequate.

The patient underwent microlaryngoscopy with multiple biopsies from the lesion. The flexible laryngoscopy findings were confirmed intraoperatively. Biopsies showed fibrous connective tissue lined by respiratory epithelium displaying focal squamous metaplasia. The subepithelial tissue showed acellular pale eosinophilic amorphous deposits resembling amyloid focally, extending to the epithelium and on the wall of blood vessels consistent with amyloidosis.
The lesion was negative for fungus and tuberculosis. He was evaluated by the Department of Medicine for systemic amyloidosis which did not show any significant findings.

The patient was reposted under general anesthesia in September 2014 for laser excision of the lesion. The mucosa-covered lesion was completely excised using Lumenis Acupulse CO₂ laser attached to the microscope (Fig. 2). This tissue was sent for biopsy which also came back as amyloidosis (Fig. 3).

The patient was kept in follow-up for the last 6 months on OPD basis and, repeat flexible laryngoscopy showed no signs of recurrence of glottic amyloidosis (Fig. 4).

**DISCUSSION**

The exact cause or origin of laryngeal amyloidosis is not known.

Two theories have been proposed to explain localized amyloidosis of the larynx. One inculcates a plasma cell reaction to inflammatory antigens and is supported by pathologic studies showing mixed polyclonal plasma cells interspersed with the amyloid tissue. Another more likely scenario points to the body’s inability to clear light chains produced by plasma cells located in the mucosal-associated lymphoid tissue.⁵

Treatment of laryngeal amyloidosis ranges from observation to hemilaryngectomy. If symptoms are present, endoscopic surgical excision of any amyloid tumor with maintenance of functional anatomy is the goal of treatment. Unfortunately, there is a high likelihood of recurrence after resection, and therefore long-term follow-up is required.⁶

There are some studies that suggest that debulking of the lesion with postoperative radiotherapy gives a better result.⁶

Our case of laryngeal amyloidosis was treated successfully by laser treatment with no signs of recurrence for 6 months.

At present, only a handful of cases have been reported from India, which makes this case report unique in its own way as the patient has been symptom-free till the last hospital visit.
CONCLUSION

Laryngeal amyloidosis is a rare entity and care should be taken to ensure complete clearance of disease. Systemic amyloidosis should also be ruled out. Follow-up should be maintained for a minimum of 6 months as chances of recurrence are high.

Surgical resection with or without radiation have shown good results in most of the reports published.

REFERENCES