

# Isolated Primary Subglottic Laryngeal Amyloidosis Presenting as Sudden Airway Obstruction

Hemanth Vamanshankar, Shreekantha B Kakkillaya, Shantanu Tandon, Ravi C Nayar

## ABSTRACT

Laryngeal amyloidosis of head and neck is a rare benign disease of unknown etiology. It accounts for less than 1% of all the benign laryngeal tumors. It is seen most commonly in the 5th to 7th decade of life with 3:1 male to female predominance. It may present with hoarseness, pain or difficulty in breathing.

We report a case of isolated laryngeal amyloidosis in a 46-year-old female who presented to us as a surgical emergency in stridor, requiring a tracheostomy. The patient was managed by staged microlaryngoscopy and CO<sub>2</sub> laser excision, and is on regular follow-up.

Laryngeal amyloidosis is an uncommon benign pathology, and can present with acute airway obstruction. Isolated subglottic laryngeal amyloidosis should be kept in mind in any case presenting with acute upper respiratory tract obstruction. Long-term follow-up is essential because of the slowly progressive nature of the disease.

**Keywords:** Amyloidosis, Laryngeal, Subglottic, Airway obstruction.

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## INTRODUCTION

Laryngeal amyloidosis of head and neck is a rare benign disease of unknown etiology. It accounts for less than 1% of all the benign laryngeal tumors. It has a 3:1 male to female predominance, and commonly presents between 5th and 7th decades of life with symptoms of hoarseness, pain or difficulty in breathing. Staged surgical excision by microlaryngoscopy using a CO<sub>2</sub> laser is the therapy of choice. However, long-term follow-up is essential because of the slowly progressive nature of the disease.

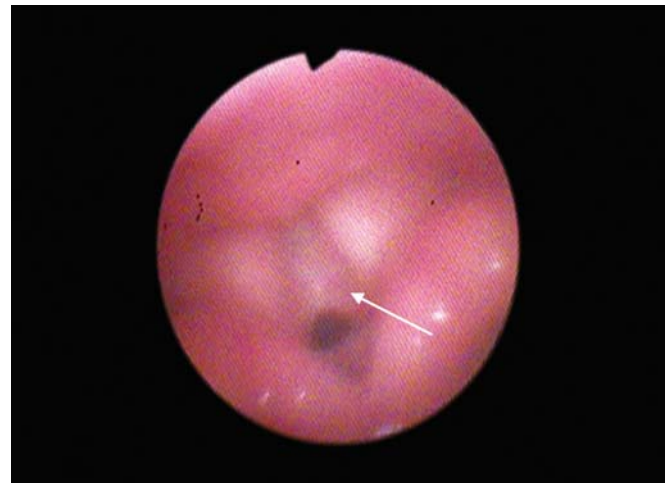
We report a case of isolated laryngeal amyloidosis in a 46-year-old female who presented to us as a surgical emergency in stridor, requiring a tracheostomy.

## CASE REPORT

A 46-year-old, obese lady, presented to the otolaryngology Outpatient Department at St John's Medical College Hospital, a tertiary care center, in Bengaluru, South India, with complaints of difficulty in breathing on exertion since 6 months and noisy breathing since 15 days. She was diagnosed and treated as a case of bronchial asthma with

history of multiple visits to the physician and worsening of her symptoms, before she presented to our outpatient department with features of acute upper airway obstruction. On examination the patient was in stridor, with use of accessory muscles of respiration.

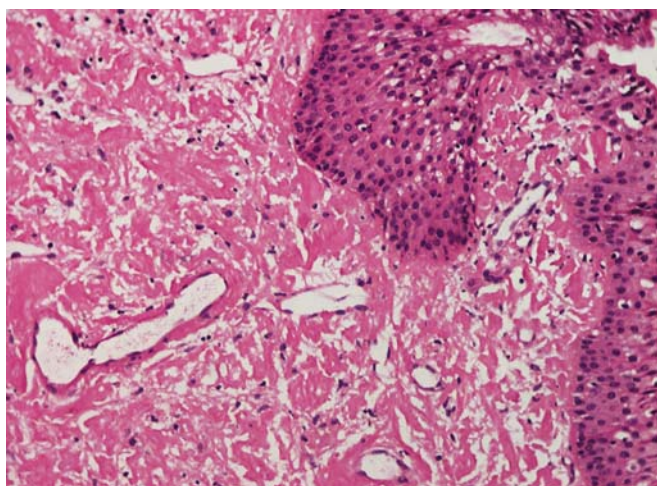
A 90° videolaryngoscopy with Hopkins rigid endoscope showed a subglottic growth compromising the airway (Fig. 1). A computed tomogram of the neck showed narrowing of the subglottis (Fig. 2). In view of acute stridor, emergency tracheostomy was performed to secure the airway. A subsequent direct laryngoscopy showed a proliferative mass, involving the subglottis and obstructing more than 80% of the airway. A biopsy was taken from this



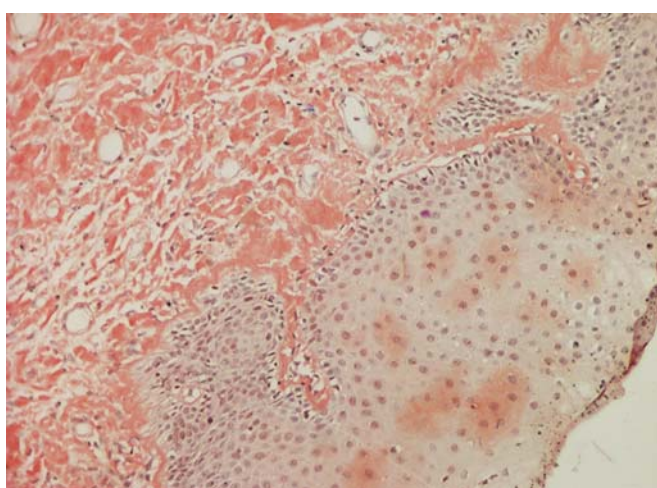
**Fig. 1:** Videolaryngoscopy showing lesion in the subglottis, compromising the airway



**Fig. 2:** Computed tomogram of the neck (axial view) showing narrowing at the subglottis



**Fig. 3:** Amyloid showing glossy eosinophilic staining on H and E



**Fig. 4:** Congo red stain showing the typical Salmon pink staining of amyloid

mass and sent for histopathological examination, which was suggestive of amyloidosis (Figs 3 and 4).

Further investigations were done to rule out systemic amyloidosis. The patient's complete blood count, erythrocyte sedimentation rate, basic metabolic and biochemical tests were within normal limits. A serum electrophoresis showed mild increase in gamma globulin of polyclonal origin, but the urine electrophoresis was negative for Bence Jones proteins. Abdominal fat biopsy was negative for amyloidosis. Based on these findings, systemic amyloidosis and multiple myeloma were excluded. The amyloid deposits were removed by staged microlaryngoscopy and CO<sub>2</sub> laser excision. Patient is now on regular follow-up with no evidence of air way disease since past 1 year.

## DISCUSSION

Virchow was the first to use the term amyloidosis because of the starch like reaction when treated with iodine and

sulfuric acid.<sup>1,2</sup> Laryngeal amyloidosis was first recognized at postmortem examination in 1842.<sup>2,3</sup> Although localized amyloidosis is a relatively uncommon; larynx is the most common involved site in the upper aerodigestive tract.<sup>1</sup>

Amyloidosis can be hereditary or acquired.<sup>2,4</sup> Of the 15 described biochemical forms of amyloidosis, the commonest three types are the following:

- AL (light chain)*: derived from plasma cells and contains kappa and lambda immunoglobulin light chains.
- AA (amyloidosis associated)*: amyloidosis is a unique nonimmunoglobulin protein synthesized by the liver. It is a reactive systemic amyloidosis.
- AB*: amyloidosis seen in Alzheimer's diseases and occasional familial cases.

Amyloidosis can also be classified according to the clinical involvement into systemic or localized forms.<sup>2</sup>

Laryngeal amyloidosis is reported in literature at ages ranging from 11 to 80 years the highest incidence lies in the 5th decade.<sup>2,5</sup> It is more common in men with a M:F ratio of 3:1.

Hoarsness is the most common symptom, followed by progressive dyspnea, hemoptysis and dysphagia. Laryngeal amyloidosis presenting with stridor is rare.<sup>1</sup> The few cases of (10) laryngeal amyloidosis presenting with airway obstruction have had a large supraglottic lesion. However, based on the case in point, it may be prudent to rule out amyloidosis in an undiagnosed case of subglottic pathology, causing airway obstruction.

Based on world literature, there is a predilection for supraglottic involvement in 54.05% of all the cases, with the ventricle and false cords being the most common, followed by glottis (18.91%), transglottic (16.21%) and tracheal involvement (10.8%).<sup>1,2</sup>

Lewis et al from the Mayo clinic, recommended urine and serum electrophoresis as a basic initial approach for diagnosis of patients suffering from amyloidosis located exclusively in the larynx.<sup>6</sup> Amyloid protein is commonly detected histologically by staining the biopsy samples excised with congo red stain, which would impart an apple-green color to the amyloid protein. In polarized light, amyloid appears birefringent. Extensive investigations are not necessary to rule out systemic amyloidosis and it can be tailored to suit individual patients, in the absence of systemic symptoms.<sup>6</sup>

Currently the most effective treatment is CO<sub>2</sub> laser-assisted surgical excision. Long-term follow-up is essential because of the slowly progressive nature of the disease.<sup>2,5</sup>

## CONCLUSION

Laryngeal amyloidosis is an uncommon benign pathology, and can present with acute airway obstruction. However, progression to systemic involvement is rare. Isolated subglottic laryngeal amyloidosis should be kept in mind in any case presenting with acute upper respiratory tract obstruction. More often than not, multiple managements for recurrent disease are required; hence, techniques that minimize trauma to the larynx must be used. Staged surgical excision by microlaryngoscopy using a CO<sub>2</sub> laser is the therapy of choice. Long-term follow-up is essential because of the slowly progressive nature of the disease.

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