

## Case Report

# Laryngotracheal Amyloidosis Presenting as a Laryngeal Mass

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**Abstract**

Amyloidosis is a disease caused by improper three-dimensional folding of precursor proteins, which results in formation of protein aggregates, amyloid fibrils, that cause cell and organ damage. Amyloidosis localized to the respiratory tract is considered rare, with a spectrum of clinical symptoms. We report a case of a 72 year old man who presented with a laryngeal mass, found to be an amyloidoma. Cross-sectional imaging revealed disease involving the tracheobronchial tree. Despite its rarity, amyloidosis may be included in the differential diagnosis for diffuse nodularity and thickening involving the laryngeal folds and the tracheobronchial tree.

**ABBREVIATIONS**

CT: Computed Tomography

**INTRODUCTION**

Amyloidosis results from improper folding of precursor proteins which results in cell and organ damage [1]. Various classifications of amyloidosis exist, either describing the precursor protein that is misfolded [2] or based upon the distribution of abnormality [1]. Amyloidosis confined to the respiratory tract is rare, and is categorized as localized. It is subdivided into laryngeal, tracheobronchial, or parenchymal categories [3]. Amyloidosis characterized by submucosal amyloid deposition represents approximately 0.2-1.5% of benign laryngeal lesions [4-6]. Involvement of the tracheobronchial tree is characterized by diffuse nodularity and thickening, with areas of bronchial narrowing [7] which may cause symptomatic sequelae such as atelectasis and post-obstructive pneumonia [5,7]. Definitive diagnosis relies on histological analysis. Despite its rarity, the radiologist may include amyloidosis in the differential diagnosis of diffuse nodularity and thickening involving the laryngeal folds and tracheobronchial tree.

**CASE PRESENTATION**

A 72 year old man was referred to the Otorhinolaryngology clinic for evaluation of an asymptomatic laryngeal lesion, incidentally found in evaluation of dyspepsia. He also noted an

intermittent choking sensation with self limited cough. A former smoker, his past medical history included coronary artery disease, HTN, atrial fibrillation, and gout. Fiberoptic laryngoscopy revealed a laryngeal polypoid lesion involving the left false vocal cord.

The patient was electively brought to the operating room for microscopic direct laryngoscopy with excisional biopsy. At that time, the lesion was noted to be yellow, with a broad attachment to the false vocal cord along its entire base and mucosal covering (Figure 1). The lesion was excised and sent for pathology. A rigid



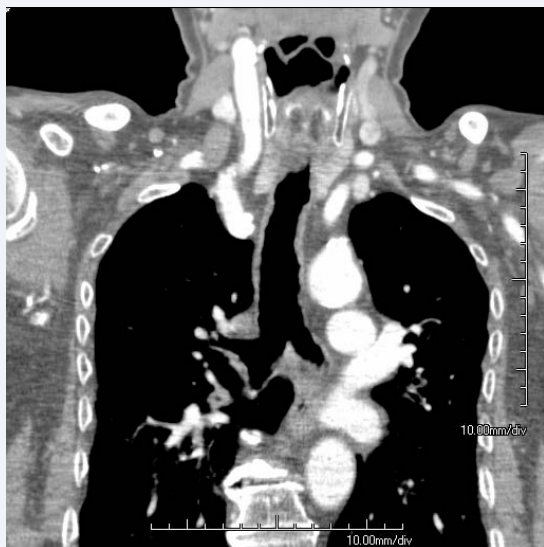
**Figure 1** Gross image of the larynx at the time of excisional biopsy demonstrates a yellow soft tissue mass with its attachment to the left false vocal cord.



**Figure 2** Endoscopic view of the trachea at the time of excisional biopsy revealing nodular and fatty changes within the trachea.



**Figure 3** Post biopsy axial CT image at the level of the larynx demonstrates nodularity and thickening involving the true vocal cords and the anterior commissure.



**Figure 4** Post biopsy reformatted coronal CT image of the tracheobronchial tree demonstrates nodularity and thickening involving the tracheobronchial tree. Additional images not included, revealed areas of bronchial narrowing.

bronchoscopy was performed, which revealed nodular and fatty changes throughout the entire trachea (Figure 2).

After excisional biopsy, the patient was sent for a CT scan of the neck and thorax. CT of the neck (Figure 3) demonstrated nodularity and thickening of the true vocal cords and the anterior commissure. The remaining soft tissues of the supra- and infrahyoid neck were unremarkable. Reformatted coronal image of the thorax (Figure 4) demonstrated nodularity and thickening throughout the tracheobronchial tree. Additional images not included revealed areas of bronchial narrowing. On clinical grounds, there was no suspicion of systemic amyloidosis.

## DISCUSSION

Amyloidosis comprises a group of diseases occurring secondary to improper three-dimensional folding of precursor proteins. As a consequence, insoluble aggregates, amyloid fibrils, lead to cell and organ damage [1]. While roughly 28 different proteins are known to cause amyloidosis [8], all forms characteristically develop cross- $\beta$  sheet structures and apple green birefringence with Congo red histological staining [1].

Several classification schemes exist to categorize amyloidosis. One includes classification based upon systemic versus localized clinical grounds. In the systemic form, the site of amyloid deposition differs from the site of the precursor protein synthesis. Alternatively, in the localized form, the abnormally accumulated amyloid fibrils occur in the same tissue or organ that synthesized the precursor protein [1]. The localized form is confined to the gastrointestinal system, genitourinary system, lungs, skin, or respiratory tract [8].

Localized amyloidosis affecting the respiratory tract is subdivided into laryngeal, tracheobronchial, and parenchymal categories [3]. The larynx is the most commonly affected organ within the head and neck. Patients may present with hoarseness, difficulty breathing, or abnormal phonation [4, 9]. The amyloid deposits are submucosal and homogeneous [6]. While the entire larynx may be involved, the supraglottic region, particularly the false vocal cords, is most commonly affected [4,6,10]. Concomitant involvement of the trachea may be seen [10].

Imaging features of a laryngeal amyloidoma on CT are nonspecific, consisting of a well defined submucosal mass, with possible calcification [4]. On MRI, an amyloidoma is isointense to skeletal muscle on T1 weighted images, and isointense to slightly hyperintense on T2 weighted images [4].

Tracheobronchial amyloidosis is the most common variant affecting the respiratory tract. Typically, there is sparing of the pulmonary parenchyma. Imaging features include tracheobronchial wall thickening with resultant luminal narrowing and stenosis [7]. Tracheal amyloid deposits on endoscopy may appear as submucosal nodules, superficial yellow lesions, or erythematous elevations [3].

Diffuse nodular thickening of the tracheobronchial tree without pulmonary parenchymal abnormalities is a nonspecific finding, eliciting a differential diagnosis of: laryngotracheobronchial papillomatosis, relapsing polychondritis, tracheopathia osteochondroplastica, Wegener's, sarcoid, and idiopathic laryngotracheal stenosis [3,7].

Clinical manifestations of systemic amyloidosis were not present in the patient reported. A cardiac MRI was negative for amyloid involvement. Of note, the patient presented with a prior medical history that included gout. An association between gout and amyloidosis is rare, with a paucity of case reports[11].

There is no definitive cure for laryngotracheal amyloidosis. Despite a benign course, complications include secondary pneumonia, atelectasis, and recurrent disease after bronchoscopic interventions [7,10,12]. Laryngotracheal amyloidosis should be considered in the differential diagnosis for cases presenting with nonspecific submucosal nodularity involving the larynx, and diffuse wall thickening and nodularity involving the tracheobronchial tree.

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