Case report of Laryngeal Amyloidosis: Unusual cause of hoarseness

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ABSTRACT

Background: Laryngeal amyloidosis is a rare pathology of the airway in which the diagnosis can only be made via tissue biopsy. We reported a case of laryngeal amyloidosis presenting with symptoms of hoarseness and voice fatigue.

Case report: This is a case report of laryngeal amyloidosis in a middle-aged female patient who presented with hoarseness of voice. Upon obtaining a further history and performing a clinical examination, it was noted that the patient had a laryngeal mass. She underwent surgery, and the mass was histopathologically confirmed as laryngeal amyloidosis. She was successfully treated with a combination of surgical and medical treatment. This case presentation describes these rare pathological findings, their clinical manifestations, the histopathological and surgical diagnostic problems, treatment and patient’s progress that we followed up till presence.

Conclusion: Isolated laryngeal amyloidosis is a rare benign laryngeal tumor that is often presented with various symptoms depending on the tumor location and can even cause upper airway obstruction. It is a histological diagnosis confirmed by histopathological specimens that are stained with Congo red. Treatment of laryngeal amyloidosis includes symptomatic relief, surgical or even conservative treatment.

Keywords: vocal cord mass; airway; laryngeal mass treatment.


INTRODUCTION

Amyloidosis is a rare disease distinguished by deposition of extracellular fibrillar protein, which subsequently progresses to organ failure.1 Isolated laryngeal amyloidosis is uncommon, constituting only 0.2-1.2% of all non-malignant tumors of the larynx.1,2 It was first reported by Borrow & Neuman in 1873.3,4 The most common site for amyloid deposition is the ventricle, followed by the false and true vocal cord. Other laryngeal subsites include aryepiglottic folds and subglottis.5 Symptoms of laryngeal amyloidosis are related to the involvement of the anatomical location and size of the tumor. We are reporting a case of laryngeal amyloidosis with presenting symptoms of hoarseness and voice fatigue.

CASE REPORT

Twenty-eight years old female complained of hoarseness of voice for four and a half years. She described her voice as rough, and the symptom progressively worsened. There was also voice fatigue throughout the day. She was very disappointed as her voice did not recover. She had neither shortness of breath nor noisy breathing. She could tolerate a normal diet without odynophagia or dysphagia. There were neither reflux symptoms nor aspiration symptoms. She denied any excessive use of voice or voice abuse. Upon further questioning, she denied any history of prolonged cough, night sweat or tuberculosis contact. She did not smoke cigarettes, but she drank alcohol during special occasions. There were also no constitutive symptoms such as loss of appetite or significant loss of weight. She has no previous medical illness and no background of malignancy in her family. She had undergone her first surgery, direct laryngoscopy and biopsy for laryngeal mass four years ago. Biopsy of the mass was reported as laryngeal amyloidosis.

During the examination, she was sitting comfortably without respiratory distress. Her vital signs were normal. No additional breathing sound such as stridor can be heard. Upon neck examination, her laryngeal framework was palpable and no neck mass or cervical lymph node present. Seventy-degree laryngoscopy examination...

Figure 1. a smooth surfaced mass seen over the right false cord
Amyloidosis is a disease characterized by generalized amyloid deposition to all tissues or isolated to a single location or organ. There are two hypotheses for the amyloid depositions. First, it is caused by the production of amyloid proteins specific to an organ or site. The other cause is the pathological process like inflammation of an organ causing amyloid deposition from the systemic light chain.

Isolated laryngeal amyloidosis is usually presented in a patient aged 50-60, but previously there are reports involving a younger age group (8 – 12 years old). Patient may be presented with symptoms of hoarseness, voice fatigue, dysphonia or stridor. Some may present with difficulty in swallowing. Symptoms vary as it depends on the location and size of the tumor.

Although rare, laryngeal amyloidosis may lead to upper airway obstruction due to significant airway compromise. Laryngeal amyloidosis usually appears as a smooth-looking mass. There has been a reported case of laryngeal amyloidosis with papillomatous changes of the false vocal cord mucosa. The surgeon’s initial diagnosis was carcinoma until proven as laryngeal amyloidosis via histology examination.

Diagnosis of laryngeal amyloidosis is made via histology examination. Positive Congo red immunohistochemical staining revealed a pink or red color under normal light. However, under polarized light, the classical apple-green birefringence appearance can be seen.

Radiological investigations can help to support the diagnosis. The appearance of the mass as the intermediate signal on T1 and low signal intensity on T2 weighted magnetic resonance imaging (MRI) are in resemblance of skeletal muscle. The similarity was caused by depositions of amyloids in the form of parallel sheers, same as the organization of skeletal muscle fibers. Computed tomography (CT) scan helps identify the site and extension of the mass and a pre-operative preparation before general anesthesia. That can help the surgeon or even the anesthetist plan the surgery or anticipate upper airway difficulties.

Laryngeal amyloidosis is most often treated surgically. The surgery aims to be normal. Epiglottis, bilateral arytenoids and bilateral pyriform fossa were also normal (Figure 1). Tumor removal was performed using both cold instruments and also carbon dioxide laser. Performing the surgery was challenging as it was microsurgery with a very small surgical field. The surgeon meticulously excised the tumor to avoid injury to the true cord.

On the first day after the surgery, she was able to tolerate a soft diet without aspiration. She was comfortable without noisy breathing or difficulty breathing. She was discharged with proton pump inhibitor, T. Esomeprazole 40mg BD and adequate analgesia. After the surgery, the slough was occupying the right false cord from a 70-degree laryngoscopy upon examination in the ENT clinic. Both vocal cords were mobile and symmetrical bilaterally. After five weeks post-operation, the sloughs had resolved with no residual mass at the right false cord.

Histopathology report of the mass showed fragments of fibro collagenous tissue with extracellular, eosinophilic deposits seen within the stroma. There was a focal respiratory type epithelium lining seen. There was no dysplasia or evidence of malignancy. The eosinophilic deposits were highlighted by Congo red stain with an apple green birefringence appearance under a polarized microscope after special staining. Therefore, the findings were confirmed as laryngeal amyloidosis.
confirm the histopathological diagnosis and improve the patient's quality of life in breathing, voice projection, and swallowing. Carbon dioxide laser is one of the surgical instruments that has been proven to reduce trauma compared to its cold instruments counterparts. In our case, a carbon dioxide laser was used. Laryngeal surgery is fine microsurgery that requires a skilled surgeon. It is important to carefully eradicate the tumor as recurrence is common if not fully excised. Therefore, the patient requires long term follow-up after the surgery.

We must not take lightly when patients complain of long-standing voice hoarseness. Although it is most likely benign pathology of the larynx, proper history taking, detailed physical examination, and investigations must be carried out. Proper counseling and information must be given to patients in deciding the treatment method, complications of the treatment and the possible outcome.

CONCLUSION
Isolated laryngeal amyloidosis is a rare benign laryngeal tumor often presented with various symptoms depending on the location and can even cause upper airway obstruction. It is a histological diagnosis confirmed by histopathological specimens, stained with Congo red. Treatment of laryngeal amyloidosis includes symptomatic relief and surgical intervention.

REFERENCES