

Case Report

Laryngeal Amyloidosis

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Amyloidosis is a rare condition that disturbs the normal function of cells and organs. We report a case of a 73 year-old gentleman who presented with hoarseness of voice and globus sensation in throat. There was an irregular non-ulcerated mass over the right laryngeal surface of the epiglottis.

Endolaryngeal microscopic laser surgery was performed to remove the tumor and histopathological examination turned out to be amyloidosis. (Rawal Med J 201;42:595-597)

Keywords: Laryngeal amyloidosis, Congo red stain, endolaryngeal microscopic laser surgery

INTRODUCTION

Amyloid is a heterogeneous family of extracellular proteinaceous deposits with characteristic microscopic, histochemical, and ultrastructural features.¹ Deposition of amyloid in the larynx are rare, accounting for less than 1 percent of all benign tumors of the larynx.² Amyloidosis can be localized or systemic disease. The distinction is crucial because localized amyloidosis has excellent prognosis and can be managed conservatively. Whereas systemic amyloidosis is associated with significant morbidity and mortality.^{1,2}

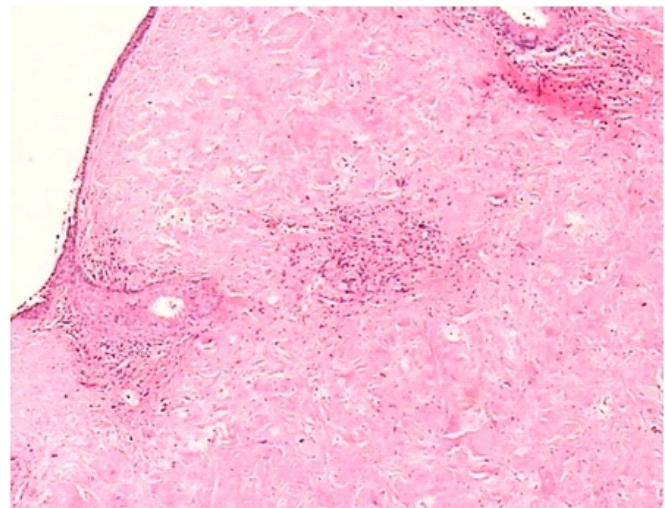
CASE PRESENTATION

A 73 year-old gentleman with multiple comorbidities of hypertension, diabetes mellitus, dyslipidemia and ischemic heart disease, presented to our clinic with 2 month history of hoarseness of voice in association with globus sensation in throat. There was no breathing difficulty or no odynophagia. He did not smoke and there was no family history of head and neck malignancy. Fiberoptic laryngopharyngoscopy examination revealed an irregular non-ulcerated mass over the right laryngeal surface of the epiglottis with normal vocal cords mobility with no airway obstruction. Head, neck and systemic examination were otherwise unremarkable. His routine blood investigations were also within normal limits.

Based on the clinical findings, a provisional diagnosis of supraglottic tumor was made. We proceeded with partial epiglottectomy and removal of the mass with laser excision. Intraoperative

finding showed the presence of a mass at laryngeal surface of epiglottis which measured 2cmx2cm. The false cord, vocal cord and subglottic space were otherwise normal. He showed marked improvement in voice quality. The vocal cords were mobile with no edema or scar. There was no recurrence seen during one-year follow up of the patient. Histopathological examination revealed diagnosis of laryngeal amyloidosis (Fig. 1).

Fig. 1. Hematoxylin and eosin stain showing deposition of amorphous material within the submucosa with overlying stratified squamous epithelium [single arrow]. Mixed inflammatory cells were seen in between the material [double arrow] (40x)



We proceeded with further evaluation to look for systemic involvement by performing full blood picture, kidney and liver function test, erythrocyte sedimentation rate and urine FEME but all results

were normal. Bone marrow aspiration and trephine biopsy showed normocellular marrow with no significant increase of plasma cells. Serum protein electrophoresis was reported as negative and echocardiogram revealed no cardiomyopathy. It was then concluded that this patient only had localized laryngeal amyloidosis with no systemic involvement.

DISCUSSION

Amyloidosis is an uncommon, progressive disease characterized by extracellular deposition of insoluble fibrillar proteins in tissues leading to organ failure. It can be classified into either systemic or localized based on number of organs involved. Only 9 to 15 percent of the total amyloidosis cases are classified as localized disease and head and neck region is the rarest place to have amyloidosis. Among the cases, larynx, most specifically epiglottis, is as the most common subsite to develop amyloidosis but laryngeal amyloidosis accounts for less than 1 percent of all benign laryngeal tumors.^{1,3}

Laryngeal amyloidosis commonly occurs in patient aged 40 to 60 years old with male to female ratio of about 2:1.² Patients commonly present with hoarseness of voice and globus sensation without airway compromise.^{1,3} This epidemiology profile is similar to our patient who is a 73 year-old gentleman. It is important to distinguish between localized and systemic disease because localized amyloidosis can be managed conservatively with an excellent prognosis, whereas systemic amyloidosis is associated with significant morbidity and mortality.² Therefore further evaluation for systemic disease is essential in these patients.³⁻⁵ In our patient, further evaluation for systemic amyloidosis was performed. Multiple myeloma renal and liver involvement and cardiomyopathy were ruled out with proper investigations.

Although laryngoscopy revealed an irregular submucosal mass over right laryngeal surface of the epiglottis, the diagnosis of amyloidosis was purely based on histopathology findings of deposits within the mass that appeared salmon pink with Congo red stain and under polarized light apple green birefringence.³⁻⁵ Initially, we had not thought of

amyloidosis because absence of other clinical evidence to support the diagnosis. Laryngeal amyloidosis can have similar presentation as other laryngeal pathology, e.g. laryngeal tumor, and it is important to consider laryngeal amyloidosis as a differential to mass in larynx.⁵

Surgery is the gold standard treatment for primary localized laryngeal amyloidosis. It is best done by endoscopic CO2 laser excision of the mass.^{6,7} Laser excision has advantages of precision in excision, minimal blood loss and better preservation of vocal cord function compared to conventional surgery.⁶⁻⁸

However, method of surgery largely depends on the experience and expertise of the surgeon.^{6,8-10} For our patient, he underwent endolaryngeal microscopic laser surgery to remove the lesion and no complication developed after the procedure.

The slow progression of the amyloidosis necessitates the need for continuous and regular follow-up to ensure early detection of any recurrence.^{1,2,4,6} We have followed this case for a period of one year till date there were no signs of recurrence and the patient is in good state of health.

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