

Localized subglottic laryngeal amyloidosis: a case report

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Amyloidosis is a heterogeneous group of diseases characterized by accumulation of amyloid protein in different organs of the body. Localized affection of the larynx by amyloid deposits is a rare event. This is a case of localized laryngeal amyloidosis in a 24-year-old female patient presented with hoarseness of voice. The amyloid mass was occupying the anterior part of the subglottic region and reaching up between both the vocal folds. Complete excision was achieved by cold instruments. Localized laryngeal amyloidosis despite being rare should be kept in mind as one of the causes of mass lesions in the larynx.

Keywords:

amyloidosis, hoarseness, larynx

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Introduction

Amyloidosis is a group of disorders that are characterized by extracellular deposition of abnormal proteins in various organs of the body which eventually can lead to their failure [1]. Virchow used the term amyloid to describe these abnormal proteins because of their starch-like reaction when treated with iodine and sulfuric acid [2]. Amyloidosis is a very rare systemic condition. In cases of localized amyloidosis of the head and neck region, the larynx is considered the commonest site of affection [3]. It usually affects age groups between 40 and 60 years and with a male to female predominance of 2 : 1.4 [4]. In this case report, we present a 24-year-old female patient with a complaint of hoarseness of voice that turned to be caused by localized amyloidosis.

Case report

A 24-year-old female patient presented to our Otorhinolaryngology Unit with a gradual onset of hoarseness of voice of 3 months duration not associated with dyspnea or cough. She is not a smoker with no history of alcohol consumption. She is not diabetic or hypertensive and her medical history is unremarkable. There is no weight loss, according to the patient's own words.

General and local external neck examination showed no abnormality. Flexible laryngoscopy showed a subglottic mass just inferior to the anterior commissure and occupying all the length of the subglottis anteriorly with superior extension that comes between the two vocal cords with phonation.

The mass was broad based, pinkish, with smooth surface, and no ulceration (Fig. 1).

Computed tomography with contrast was done (Fig. 2), which showed a small, soft-tissue non-enhanced lesion below the anterior commissure and extending to the subglottis with normal thyroid cartilage and thyroid gland and subcentimetric bilateral lymph nodes in the neck.

A decision of microlaryngosurgical biopsy of the mass was taken for histopathological consultation. An

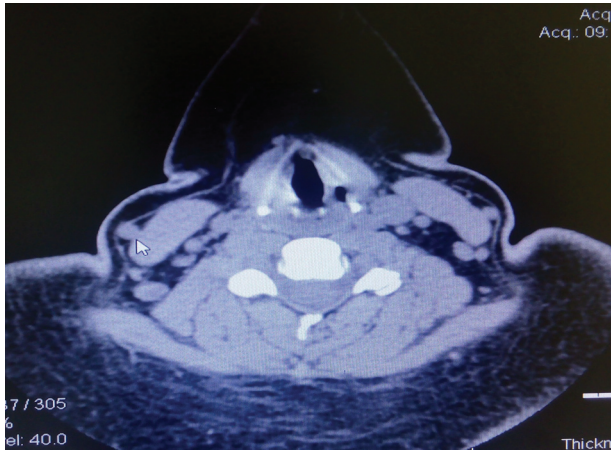
Figure 1



Mass is seen in the subglottic region.

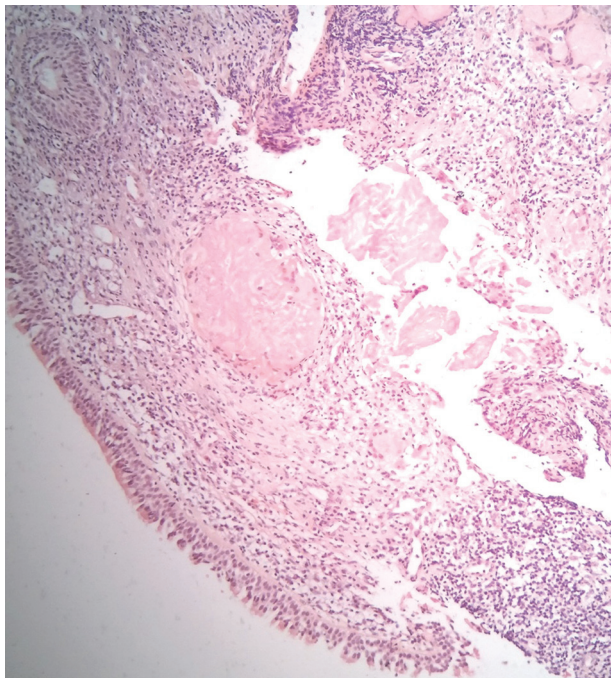
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Figure 2



Mass (subglottis below the region of anterior commissure) as seen with contrast computed tomography.

Figure 3



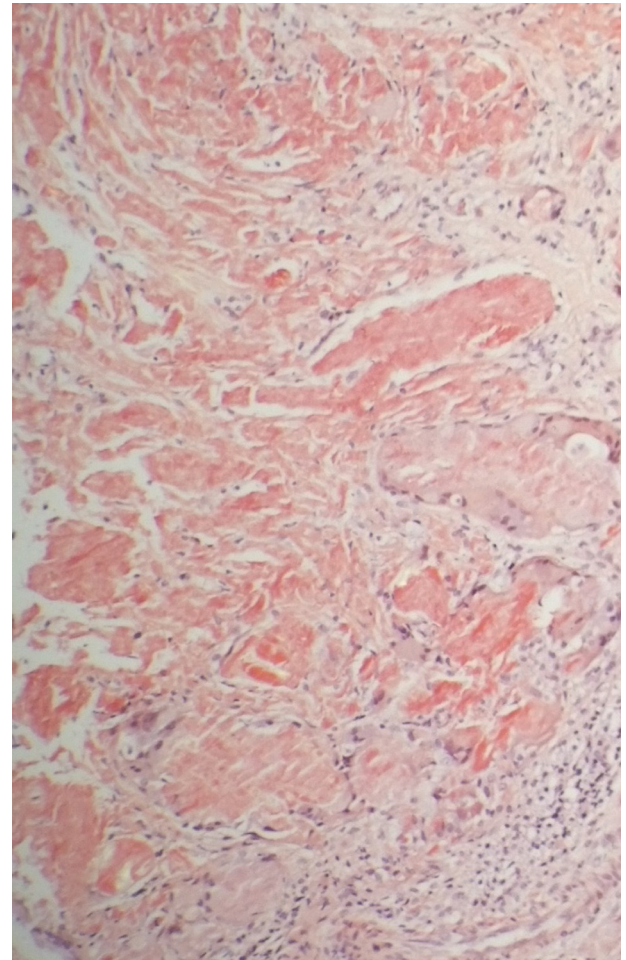
Eosinophilic deposits in the subepithelial stroma.

informed consent was taken from the patient after explanation of the procedure.

All the preoperative routine labs like complete blood count, kidney and liver function tests, serum electrolytes, and erythrocyte sedimentation rate were normal.

Under general anesthesia, microlaryngoscopy was done which showed the same criteria of the mass as seen by flexible laryngoscopy. Mass was biopsied first with no considerable bleeding and for the same complete

Figure 4

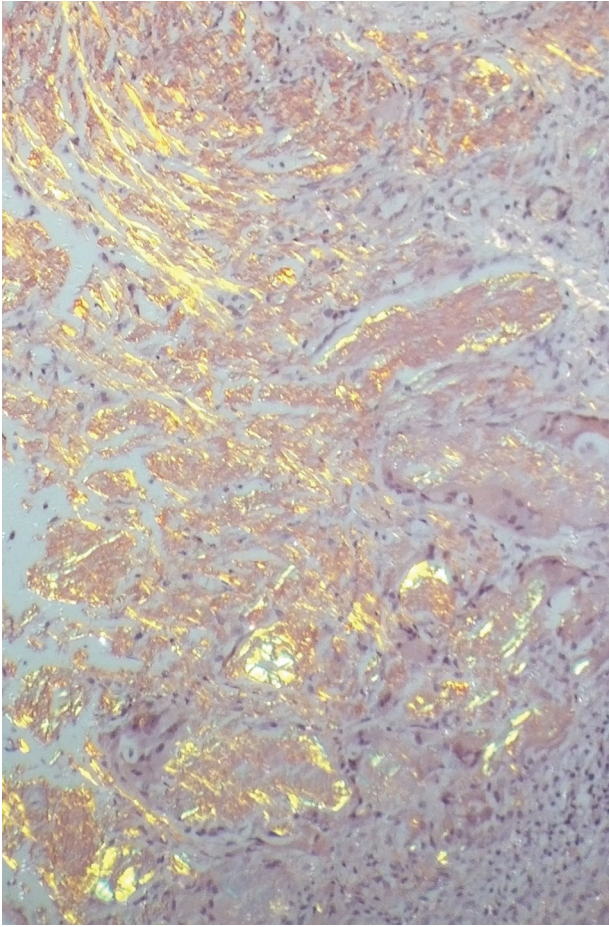


Specimen shows Congo red positive stain.

excision was done. The tissue taken was sent for histopathological examination which showed homogeneous amorphous eosinophilic deposits in the subepithelial stroma (Fig. 3). It was Congo red positive (Fig. 4) and showed apple-green birefringence under polarized light with Congo red stain which is consistent with amyloid deposits with no signs of malignancy (Fig. 5). Also, the lesion showed positive staining with periodic acid-Schiff and periodic acid-Schiff-D and negative mucicarmine.

The patient withstood the operation well and the postoperative period was uneventful. She was discharged from the hospital in the next day with no respiratory difficulty and no hoarseness of voice. She was investigated later on for exclusion of systemic amyloidosis. Chest radiography, echocardiogram, erythrocyte sedimentation rate, renal and liver functions, and complete blood count were normal. Urine analysis and 24h collection of urine for protein were also normal. Her Mantoux test and sputum acid-fast bacilli were negative. The patient was followed up for more than 18 months after

Figure 5



Apple-green birefringence.

surgery with no change of voice, no other clinical manifestations, or other examination findings suggesting recurrence of the disease.

Discussion

Amyloidosis represents a variety of conditions, characterized by extracellular deposition of abnormal, insoluble protein fibrils which can lead eventually to failure of organs and systems [5]. It may acquire systemic or localized forms, yet the latter is so rare.

Laryngeal amyloidosis remains a rare entity accounting for about 1% of all benign laryngeal tumors [1]. The first case of laryngeal amyloidosis has been reported by Burow and Neumann in 1875 [6].

Laryngeal sites which might be involved are ventricular folds (55%), laryngeal ventricle (36%), subglottic space (36%), vocal folds (27%), aryepiglottic folds (23%), and anterior commissure (14%) [1].

Despite that the immunoglobulin nature of localized amyloid is accepted generally, its pathogenesis is still unclear [7].

Laryngeal amyloidosis can simulate many other benign and even malignant lesions in the larynx like laryngocele, schwannoma, carcinoma, rhabdomyosarcoma, hemangioma, and pemphigus and because of that fact it should be put in the differential diagnosis of laryngeal masses despite its rarity.

In the case presented in this study, the lesion was smooth and no ulcerations were noted. Also, it was firm as palpated by the instruments intraoperatively. Removal of the lesion totally was done without endangering the vocal cords.

A case of familial primary localized laryngeal amyloidosis in two sisters had been documented by Oguz *et al.* [8]; however, familial primary localized amyloidosis of the larynx is a very rare finding. The case in this study showed no family history of such a condition.

The most common presentation of laryngeal amyloidosis is changes in voice [9] as has been met in our case. Amyloid protein is detected histologically by staining the samples with Congo red stain which gives an apple-green appearance to the amyloid material. Also, in polarized light amyloid appears birefringent. It has been documented that there is no need for extensive investigations to rule out systemic amyloidosis as long as no systemic manifestations are found [10].

Cases with laryngeal amyloidosis need long-term treatment due to the slowly growing nature of the lesion. The case presented in this report showed no recurrence over a period of 18 months.

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Conflicts of interest

There are no conflicts of interest.

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