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Localised Laryngeal Amyloidosis Endoscopic Excision -A Case Report

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Abstract:

We report a case of primary laryngeal amyloidosis in a 35 year old adult patient who presented with hoarseness of voice for 6 months duration with no other symptoms. Patient was treated successfully with endoscopic excision using a microdebrider with a laryngeal blade and review of literature.

Keywords: laryngeal amyloid · Endoscopy · Debrider

Introduction:

Amyloidosis represents a heterogenous family of extracellular proteinaceous deposits of amyloid in various target organs of the body leading to its malfunction¹. Clinically amyloidosis is classified in to 2 forms- systemic and localized. Localized form is extremely rare, frequently involving the head and neck region. The larynx is affected most commonly (61%) followed by oropharynx, trachea, orbit and nasopharynx². Laryngeal amyloidosis remains a rare entity accounting about 0.2% to 1.2% of all benign laryngeal tumours. Systemic manifestation is rare with laryngeal amyloidosis, and carries an excellent prognosis. Multifocal disease is present in 15% of cases³. The disease typically manifests as hoarseness or change of voice in the fifth to sixth decade and affects men: women in ratio of 3:1¹.

Case report:

A 35 year adult reported with hoarseness of voice for 6 months with no other complaints. He was a non smoker and not an alcoholic with no significant family history. The videolaryngoscopy examination revealed a smooth swelling in the left supraglottic region extending to the left vocal cord . Both vocal cords were mobile and normal. The CT scan fig(1) showed a homogenous mass in left supraglottic region. Endoscopic excision and biopsy of the mass with a debrider fig(2) using a laryngeal blade revealed deposition of acellular, homogenous amorphous, and eosinophilic deposits showing a positive staining with congo red features of amyloidosis. Postoperative period was uneventful and hoarseness of voice improved significantly.

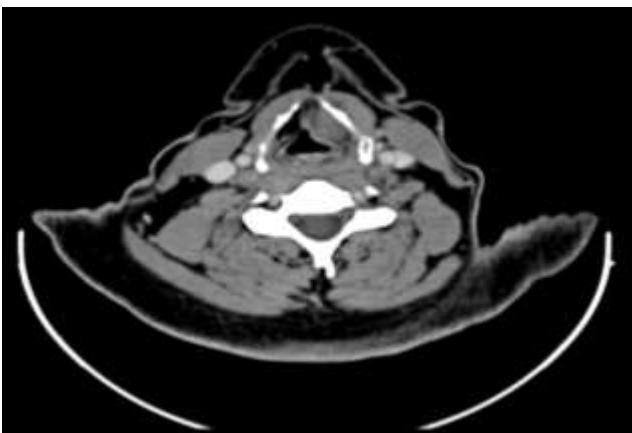


Fig 1



Fig 2

Discussion:

Amyloidosis is a benign, slowly progressive condition that is characterized by the presence of extracellular fibrillar proteins in a variety of organs and tissues and are not metabolized thus impair the function of the organ where they accumulate⁴. Virchow coined the term amyloid in 1853. Borrow described the first case in 1873 most cases of amyloid of the larynx are composed of immunologically identical to light chain fragment of immunoglobulin and is classifiable as fibril type. The source of immunoglobulin is unclear, the lesion may develop from a localized monoclonal immunoproliferative disorder in which plasma cells which are intimately associated with amyloid deposits are thought to produce light chain immunoglobulin that is deposited as amyloid rather than an inflammatory infiltrate reacting to the deposited amyloid. The second theory for the amyloid deposition suggests that a circulating precursor protein is deposited in the stroma after a change in vascular permeability as a result of local inflammation the plasma cells may be either inciting the inflammation or reacting to the amyloid⁵. The classification suggested by Symmer is

1. Generalised Primary amyloidosis
2. Generalised Secondary amyloidosis
3. Localized.

The localized form is rare the larynx affected more frequently than any other site. In larynx the sites most commonly involved are vestibular fold (55%), followed by subglottis(36%), ventricle(36%), vocal folds(27%), aryepiglottic folds(23%) and anterior commissure(14%)¹. The “gold standard” for diagnosis of amyloid is tissue biopsy demonstrating characteristic hematoxylin and eosin changes and congo red birefringence or metachromatic pink- violet staining with methyl violet or crystal violet⁵.

The treatment for localized amyloidosis is symptomatic removal. The various methods for excision are microlaryngeal instruments with conservation of surrounding tissue, CO₂ laser excision³. In our case we utilized the 15⁰ endoscope fixed with chest suspension and excision was done with microdebrider using the laryngeal blade successfully.

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