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An Interesting Case of Localized Laryngeal Amyloidosis

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Abstract: Amyloidosis forms a group of disorders characterised by extracellular tissue accumulation of amorphous hyaline material. Amyloidosis in head and neck is a rare benign disease. Among the sites in head and neck, larynx is the most commonly involved. Laryngeal involvement in amyloidosis is rare and accounts for less than 1% of all benign tumors. It usually occurs in the 40 to 60 years age. It is essential to differentiate a primary laryngeal amyloidosis from malignancy for the management of the disease. A 65years old man presented to the Department of ENT with chief complaints of hoarseness of voice for the past 3 years, it is insidious in onset, gradually progressive, associated with mild dyspnoea. On Laryngoscopic examination, a pinkish white mass of size 2x1cm in supraglottic region over false vocal cords (ventricular folds) almost covering 2/3rd of the true vocal cords and anterior commissure was noted. Bilateral vocal cords mobile and bilateral aryepiglottic folds and arytenoid cartilages are normal. Patient is known smoker, all routine investigations are within normal range, and systemic diseases are ruled out. Micro laryngeal surgery done, mass excised and sent for histopathological examination, a provisional diagnosis of laryngeal amyloidosis was given. Follow up of the patient also did not show manifestation of any systemic pathology, consequently diagnosis of localized laryngeal amyloidosis was given.
Keywords: Amyloidosis, Localised laryngeal amyloidosis, Congo red.

INTRODUCTION

Amyloidosis is a rare benign disease of head and neck region. It can be of two types, systemic and localized. It is the commonest site of involvement in head and neck region. The localized form of amyloidosis is extremely rare in larynx accounting for between 0.2 and 1.2% of benign tumors of the larynx [1]. It is usually seen in the 4th to 6th decade of life, with a male to female preponderance of 2:1 [2, 3]. Amyloid in the larynx can be identified as subepithelial extracellular deposits of acellular, homogeneous and amorphous, eosinophilic material displaying apple-green birefringence with polarized light when stained with Congo red or that is metachromatic with crystal violet or methyl violet [4]. It may be either systemic or localized to an organ. Although the etiopathogenesis of the condition is unclear, in the localized type it is hypothesized that the extracellular protein (the monoclonal light chain deposits) is synthesized locally and deposited unlike the systemic amyloidosis [5]. Macroglossia is the most common head and neck manifestation of amyloidosis, and appears in between 15 and 20% of patients with light chain disease. It usually occurs as nodules or polypoid lesions and can be located anywhere in the larynx or trachea. Systemic involvement is very rare with laryngeal amyloidosis.

CASE REPORT

A 65years old man (Fig.1) who came to ENT OPD with C/O hoarseness of voice for the past 3 years, it is insidious in onset, gradually progressive, associated with mild dyspnoea. On Laryngoscopic examination (Fig.2), a pinkish white mass of size 2x1cm in supraglottic region over false vocal cords (ventricular folds) almost covering 2/3rd of the true vocal cords and anterior commissure was noted. Bilateral vocal cords mobile and bilateral aryepiglottic folds and arytenoid cartilages are normal. There were no signs and symptoms of any systemic disease. Patient is a known smoker for past 30 years and not a K/C/O tuberculosis, diabetes mellitus, hypertension, bronchial asthma. All routine investigations are within normal range, and systemic diseases are ruled out. Micro laryngeal surgery done (Fig.3), mass excised and sent for histopathological examination, Sections studied from the mass shows a polypoidal structure lined by stratified cuboidal epithelium. Subepithelium showing aggregate of acellular, extracellular, amorphous, eosinophilic material. Also seen are few benign glands and mild chronic inflammatory cells comprising of lymphocytes and plasma cells scattered throughout the lesion. (Fig.4). Congo red stain was also positive (Fig.4). Follow up of the patient (Fig.5) also did not show

manifestation of any systemic pathology, consequently diagnosis of localized laryngeal amyloidosis was given.

Fig-1: Patient suffering from Localised Laryngeal amyloidosis

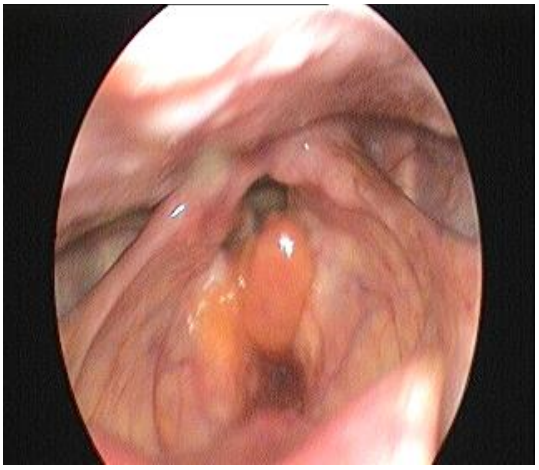

Fig-2: Laryngoscopic examination

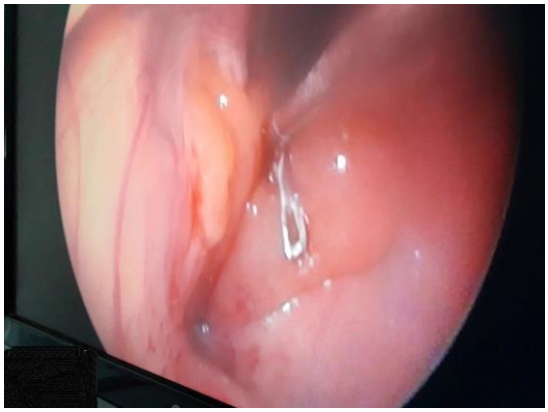

Fig-3: Micro laryngoscopic picture (videolaryngoscopy)

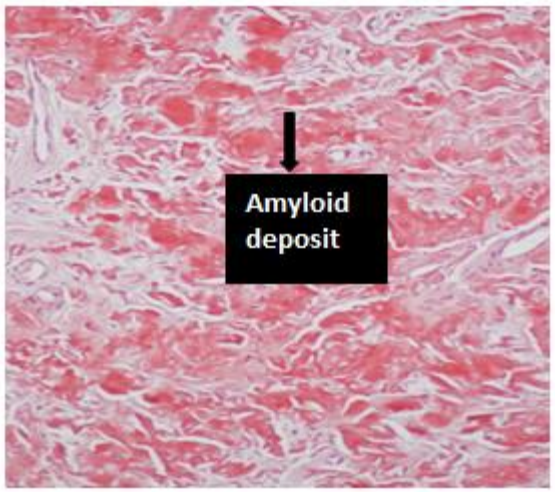

Fig-4: Histopathology findings

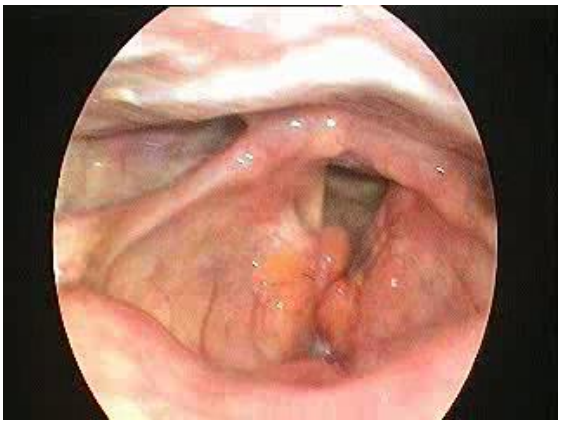

Fig-5: Postoperative findings

DISCUSSION

Amyloidosis is a metabolic disorder characterized by an abnormal deposition of the extracellular protein in various tissues of the body. Amyloidosis may be localized to a specific organ or may be generalized to various systemic organs [4]. The cause for localized laryngeal amyloidosis is unknown, it is hypothesized that it may be because of an inflammatory cell reaction of plasma cells to the antigens present [7] or the inability of the body to clear the monoclonal light chains produced by the mucosa associated lymphoid tissue [2]. Amyloid deposition leads to an alteration of the structure and affect the function of the organs [8]. The clinical appearance closely resembles that of laryngeal malignancy, thus differentiating such lesions from malignancy is essential for an effective management of the case. CT scan or MRI are useful investigation in knowing the extent of the disease. MRI is superior to CT as the amyloid deposits has intermediate T1 weighted and low T2 weighted intensity [9]. The definite diagnosis is achieved by tissue biopsy followed by the histopathological examination. The biopsied specimen reveals an eosinophilic acellular stromal proliferation which stains Congo red positive showing apple green

birefringence in polarized light suggestive of amyloidosis. Prior to labelling a primary localized laryngeal amyloidosis, it is essential to rule out the systemic involvement and other associated disorders like multiple myeloma and lymphoma. Surgery is the main stay of treatment in Laryngeal amyloidosis using CO2 laser excision. Though, the choice of laser or micro laryngeal forceps and scissors depends on the experience of the surgeon and his/her convenience. For a better postoperative outcome, CO2 Laser excision is superior due to its accuracy, healing and preservation of function [6]. Medical management in the form of corticosteroids and radiotherapy has been tried but didn't give any promising results [6]. The disease has a slow progression, but may recur which might require repeated excision requiring a regular 6 monthly follow-up. Despite the clinical resemblance of the disease to early laryngeal malignancy, a specific diagnosis of the case is mandatory for its appropriate management and cure of the disease. Thus, Laryngeal amyloidosis should be kept as a differential diagnosis

CONCLUSION

Laryngeal mass is a common presentation in patients presenting to an ENT clinic with hoarseness of voice. A systematic and thorough approach is mandatory for the effective management of such cases. Laryngeal amyloidosis should be kept as a differential diagnosis when we deal with such patients. Histopathology is the diagnostic investigation and ruling out the involvement of other systems for an appropriate management is essential.

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