ISSN: 0975-8585

Research Journal of Pharmaceutical, Biological and Chemical Sciences

Primary Laryngeal Amyloidosis: A Case Report.

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ABSTRACT

Primary laryngeal amyloidosis is a rare entity, but is the commonest site for deposits of amyloid in head and neck. A 59 years old healthy adult, a known case of allergic rhinosinusitis and chronic proximal nocturnal dyspnea came to the hospital with complaint of hoarseness of voice for the past 3 months. On examination the posterior pharyngeal wall appeared granular. Indirect laryngoscopy revealed edematous ventricular bands (Right< Left) .Mobility of vocal cords was not affected. A proliferative growth was noticed to be arising from the anterior 2/3rd of the left vocal cord. A biopsy of the lesion was taken that showed no evidence of malignancy of larynx. Two weeks later, the patient underwent repeat biopsy from left false cord growth under general anesthesia. Histopathological examination revealed amyloidosis in left false cord. Clinical presentation of the laryngeal amyloidosis often mimics neoplasm of larynx and so, a high degree of suspicion is required. Excision of the lesion with CO_2 laser is the treatment of choice.

Keywords: Larynx, Hoarseness, Amyloidosis

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ISSN: 0975-8585

INTRODUCTION

Amyloidosis is the term used for a group of diseases characterized by extracellular deposition of fibrillar proteinaceous substance called amyloid having common morphological appearance, staining properties and physical structure but with variable protein composition.[1] It may be systemic or localized which may occur in a variety of hereditary conditions. Amyloid deposits are characteristically Congo red stain positive and produces an apple-green birefringence under polarizing microscopy. [1] Amyloid deposits in the larynx are a rare entity, and account for less than1% of benign tumors of the larynx.[1] It was first reported by Borow in 1873.[2] In laryngeal amyloidosis , systemic involvement is rarely seen .[1,2] The diagnosis requires a high degree of clinical suspicion as patients usually present with complaints mimicking laryngeal neoplasms [2]. We present a case of primary amyloidosis of left false cord in a 59 year old male.

CASE REPORT

A 59 years old healthy adult, a known case of allergic rhinosinusitis and chronic proximal nocturnal dyspnea came to the hospital with complaint of hoarseness of voice for the past 3 months. There was no delay in onset of speech, voice fatigue, fluctuation in the quality of voice or foreign body sensation in the throat. There were no complaints of odynophagia, difficulty in breathing or noisy breathing. He was a chronic smoker but had stopped smoking 2 years prior. All laboratory investigations and urine examination were normal. On examination the posterior pharyngeal wall appeared granular. Indirect laryngoscopy revealed edematous ventricular bands (Right< Left) .Mobility of vocal cords was unaffected. A proliferative growth was noticed to be arising from the left ventricle and anterior 2/3rd of the left vocal cord. A biopsy was taken from the left vocal cord that showed no evidence of malignancy of larynx. Two weeks later, the patient underwent repeat biopsy from left false cord growth under general anaesthesia.Histopathological examination revealed amyloidosis in left false cord. This was confirmed by staining with Congo red. His post-operative stay in hospital was uneventful and on subsequent follow-up his voice had considerably improved. He continues to remain asymptomatic after 4 years.

DISCUSSION

Amyloidosis is related to the extracellular deposition in various tissues of an amorphous amyloid substance, formed from different protein precursors. It leads to tissue damage in the affected organs. It occurs more commonly in males(M:F=3:1) [3] It can either be localised amyloidosis or systemic amyloidosis. In a study conducted by Penner *et al*, in the head and neck region ,nearly 19% of amyloid cases were observed.[4] Amyloid deposits in the larynx are a rare entity, and account for only 0.2 and 1.2% of benign tumors of the larynx.[6] (Table 1)

Table 1
Sites involved in laryngeal amyloidosis
Ventricular folds(most common)
Laryngal venricle
Subglottic space
Vocal folds
Aryepiglottic folds
Anterior commissure

Laryngeal amyloidosis can present with various different symptoms that include hoarseness, dyspnoea, dysphagia and stridor. [3,4] Hoarseness is by far the most common symptom. In our literature search, we found only 2 cases with dysphagia as the presenting complaint. [7] Our patient complained of hoarseness of voice for the past 3 months. On the basis of the presence of a proliferative growth arising from anterior 2/3rd of the left vocal cord a diagnosis of malignancy of the larynx was suspected. Any isolated lesion encountered in the larynx should be visualized by laryngoscopy and tissue biopsy should be examined under microscope. Radiological investigations should also be done. This helps to rule out malignancy. The first biopsy of left vocal cord yielded negative results. However, repeat biopsy two weeks later revealed amyloidosis of left false cord. H and E staining on light microscopy reveals structure of amyloid as an eosinophilic, amorphous, extracellular hyaline substance. (Fig 1)Congo red dye imparts a pinkish-reddish color to tissue amyloid deposits and under polarized microscopy green birefringence of stained amyloid is seen.(Fig 2) This should be distinguished from pseudoamyloid. Laryngeal amyloidosis is a localized amyloidosis that is made of AL (light chain) type amyloid. AL type of amyloid

ISSN: 0975-8585

is resistant to potassium permanganate while AA type amyloid dissolves in potassium permanganate. This is commonly used to differentiate the type of amyloid. [5] It has been shown by by electron microscopy that all types of amyloid consist of continuous, non-branching fibers(7.5-10 nm diameter). X ray crystallography and infrared spectroscopy demonstrate a characteristic cross β pleated sheet conformation. Amyloidosis should be correctly classified as it will influence the biological behavior of the lesion, and management and prognosis of the case. Isolated primary laryngeal amyloidosis will have different behavior compared to secondary amyloidosis, multiple myeloma and extramedullary plasmacytoma. Amyloid deposition can occur in neuroendocrine neoplasms also.

Different sources of amyloid have been described that include

1)Amyloid A in reactive amyloidosis 2) Immunoglobulin light chains

3) Hemodialysis-associated amyloid 4) Transthyretin type in familial or senile amyloid

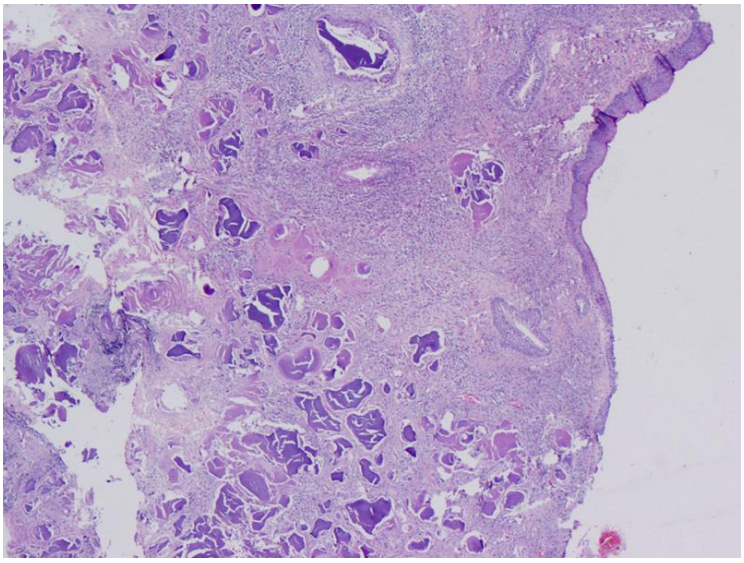

Figure 1: Amyloid appears as an amorphous eosinophilic hyaline extracellular substance (H and E stain)

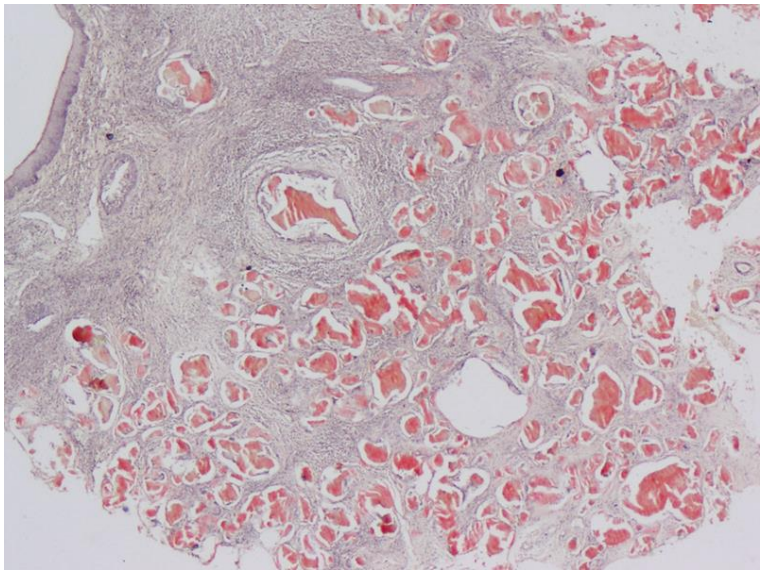

Figure 2 :Congo Red dye imparts salmon pink color to tissue amyloid deposits.

Clinical presentation of the laryngeal amyloidosis often mimics neoplasm of larynx and so, a high degree of suspicion is required.

It is not necessary to perform all the procedures and laboratory tests(Table 2)These investigative procedures can be tailored for each individual patient to help exclude systemic disease. Thus, a high disease

ISSN: 0975-8585

suspicion index followed by serum and urine electrophoresis, rectal biopsy, punch biopsy during direct laryngoscopy and MRI constitute an effective diagnostic procedure. Microlaryngoscopy with CO_2 laser is the treatment of choice.

Table 2		
Clinical and Laboratory assessment to differentiate between systemic amyloidosis and isolated laryngeal amyloidosis[6,7,8,9]		
Check for lymphadenopathy		
Radiological imaging	X-ray(especially chest) and MRI	
	Bone scan(Skeletal scintigraphy)	
	Evaluation of urinary and digestive tract	
ECG		
Endoscopic examination of aerodigestive tract		
Clinical laboratory studies		
Peripheral blood smear	Complete blood count	
Liver function tests	Renal function tests	
Quantitative immunological analysis	Electrophoresis of serum and urine	
Urine examination	Bence Jones protein analysis	
Serological test for rheumatoid arthritis	Tissue biopsy*(as clinically indicated)	
Erythrocyte Sedimentation Rate	Bone marrow biopsy	
*Rectal biopsy was earlier considered the best and most specific site to assess systemic amyloidosis but has been largely replaced by abdominal fat pad aspirate. Gingival biopsy is also commonly indicated		

Congo red dye imparts a pinkish-reddish color to tissue amyloid deposits and under polarizing microscopy apple green birefringence of stained amyloid is seen. Any other differential diagnosis -except systemic amyloidosis- may be safely excluded(Table 3) if Congo red staining is positive.

Table 3
Differential Diagnosis *of isolated laryngeal amyloidosis[8,10]
Laryngeal neoplasms
Ligneous conjunctivitis
Lipoid Proteinosis
Hyalinised type vocal cord nodule/polyp
Small cell carcinoma of larynx
Medullary thyroid carcinoma invading larynx
*Other than other causes of amyloid

CONCLUSION

Amyloidosis of larynx is usually seen to act as a benign disorder but can be progressive or , recurrence after treatment may be seen. To help detect any recurrence, regular follow-up and visualization with laryngoscopy is required. To control symptoms, multiple surgeries may be indicated.

ISSN: 0975-8585

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