

An Isolated case of Laryngeal Amyloidosis

Jagannatha Prasad¹, Kalyana Chakravarthi², Subba Rao M.V³, Rama Krishna Tirumala Bukkapatnam⁴

¹Assistant prof. Dept. of ENT, Medicit Institute of Medical Sciences, Ghanpur RR Dist. Telangana state, PIN: 501401

²Assistant Professor, Medicit Institute of Medical Sciences, Ghanpur RR Dist. Telangana state, PIN: 501401

³Professor, Dept. of ENT Medicit Institute of Medical Sciences, Ghanpur RR Dist. Telangana state, PIN: 501401

⁴Professor, Dept. of ENT Shadan Institute of Medical Sciences, Peeramcheruvu Hyderabad, PIN: 500008

*Corresponding author

Dr. Ramakrishna Tirumala Bukkapatnam

Email: tbramkrishna@gmail.com

Abstract: A variety of benign and malignant lesions involve larynx. Though rare, amyloidosis can involve paraglottic region of larynx, false cords and true cords. An isolated case of laryngeal amyloidosis has been treated with Microlaryngoscopy and excision. The main stay of confirmation was by hisytopathological examinartion with special staining with congo red. A three year follow up of the patient showed no residual or recurrent disease. Systemic examination did not reveal any involvement of other organs.

Keywords: malignant lesions , larynx , amyloidosis, Microlaryngoscopy.

INTRODUCTION

More than a century ago, when Borow [1] described three amyloid ‘tumors’ in the larynx, it was not known that amyloidosis is essentially a metabolic disorder involving light-chain proteins. With a less than 1% incidence amongst laryngeal masses, Laryngeal amyloidosis constitutes a singularly curable entity by surgery whether by Microlaryngoscopy (MLS) or by Laser [2, 3]. Though poses a formidable diagnostic dilemma with malignant tumors, the ‘birefringence’ nature of amyloid with apple green colour under polarized light with congo red staining of tissue exemplifies the fact that Pathological examination is the gold standard. We are presenting a case of laryngeal amyloid which presented to the department of ENT at an early age

CASE REPORT

A 26 year old male patient, agricultural labourer, non- alcoholic, non-smoker presented with change of voice of 6months duration in February 2011.He has no history of vocal abuse or dysphagia. Clinical examination of neck did not reveal any enlargement of cervical nodes.

Video-laryngoscopy examination showed a smooth, bosselated, elevated, mucosa-covered, diffuse mass involving laryngeal surface of epiglottis, left false cord, with hindrance to visualize the true cord. The right true and false cords and aryepiglottic folds were normal (Fig- 1).Both vocal cords were found to be mobile. CT scan of larynx was done which showed a non-enhancing soft tissue homogeneous shadow in the region of left translottic region. The thyroid, cricoid

and arytenoid cartilages were unremarkable. Though MRI scan was recommended, it couldn’t be done due to personal constraints. After thorough blood chemistry investigations, Patient was taken up for Microlaryngoscopy and excision of the mass under general anaesthesia. Adequate preparations were done to conduct tracheostomy per operatively, however, the need didn’t arise.

The tissue was subjected to histopathological examination (Fig-2) and the true nature of amyloid was confirmed by extra cellular nature of homogeneous, eosinophilic filaments which were not soluble on potassium permanganate wash. Confirmation was done on congo red staining with apple green ‘birefringence ‘under polarized light.

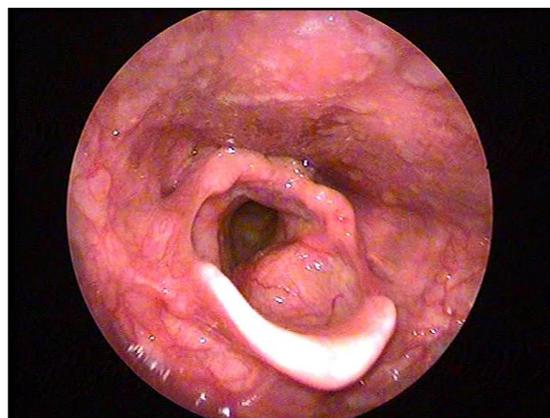


Fig-1: Video laryngoscopy Picture showing smooth, diffuse, bulge in the region of petiole of epiglottis and false cord

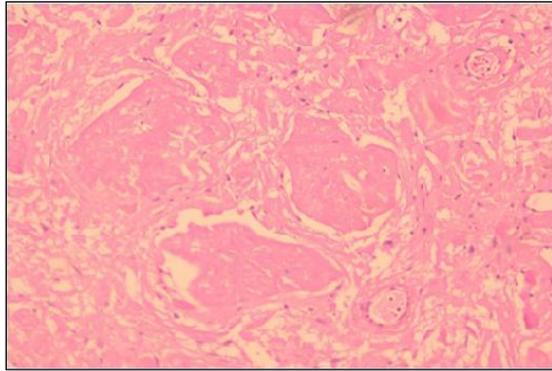


Fig-2: Eosinophilic stained amorphous substance with scanty plasma cell infiltration.

Subsequent to pathology report, thorough investigations were carried out like ESR, RFT, LFT, thyroid function tests, Echo cardiography, urine bence-jones proteins, abdominal fat FNAC, and rectal biopsy. None of the reports came positive for systemic amyloidosis.

DISCUSSION

Amyloid (starch like) was first described by Rokitsky [4] in 1842 and looks like pale yellow to red-gray, amorphous substance that gets deposited under the sub epithelial region in the lamina propria extracellularly. The origin of such deposition caused a lot of debate. It was believed that sparse infiltration of plasma cells around the blood vessels and at the periphery of the amyloid was suggested to be the source of such deposition. Immuno-phenotyping which demonstrated that the substance was in fact was a fibrillar protein which is similar to variable region of light chain fragment of immunoglobulins. It is theorized that the secretions of the monoclonal plasma cystic infiltrates of the larynx are metabolised into immunoglobulin light chains fragments by adjacent histocytes and deposited extracellularly as amyloid.

The incidence of amyloidosis male to female ratio is 3:1 with most cases occurring in the 5th decade of life .The youngest reported case of localised laryngeal amyloidosis is that of an 8yr old girl .Incidence of the disease increases with increased age .The pathogenesis of laryngeal amyloidosis remains unknown. Although rare, laryngeal amyloidosis may occur in patients with an underline lymphoid neoplasm or in association with systemic amyloidosis. Amyloidosis can be hereditary or acquired, systemic or localised.

The common site of laryngeal involvement was found to be false cords and trans glottis region [5-7] ,in our case it was laryngeal surface of epiglottis extending on to false cord .The average age of presentation was around 37years , but in our case ,he was younger by 10 years. Laryngeal amyloid may be a part of systemic amyloidosis or may be secondary to

Rheumatoid arthritis. Amyloidosis can be secondary to lymphomas. Hence, a complete plethora of investigations have to be carried out. Recently FNAC of abdominal fat was found to be positive in 95% primary systemic amyloidosis and 66% of secondary systemic amyloidosis [8, 9].

A review of laryngeal amyloidosis in the literature showed more often than not these are isolated. So it may be appropriate to consider laryngeal amyloidosis to be a 'localized monoclonal immune proliferative disorder'. In a systemic involvement of amyloidosis, the plasma cell infiltrates are spatially separated from the deposited amyloid [10].

Magnetic resonance Imaging (MRI) scan is gaining popularity [11], more than CT scanning in identifying Amyloid disease. The deposits present an intermediate intensity on T-1 weighted scan, and low signal intensity on T-2 weighted image. Thus MRI scan seems to be the best option; however, it could not be done in our case.

Treatment of localised laryngeal amyloidosis varies from simple observation of lesion to partial laryngectomy. Management of localised laryngeal amyloidosis should be limited to local excision of laryngeal lesion when symptomatic .Observation is the best treatment in asymptomatic patients. Localised tumour like lesions may be completely removed endoscopically with cold knife, conventional micro laryngoscopy or with incorporation of carbon dioxide laser/ KTP532 laser.

Though laser ,both carbon dioxide and KTP [12] were regarded as highly effective in reducing the haemorrhage, with high accuracy of excision and low recurrence rate , in our study, with a follow up period of 3 years , there was no recurrence with simple MLS excision.

CONCLUSION

A smooth, non -ulcerative, diffuse, firm mass with intact mucosa in the supra glottic region should raise the suspicion of amyloidosis. A systemic search in the form of abdominal fat FNAC and serum immunoglobulin study should rule out a generalized disease. Microlaryngoscopy with or without Laser will be the treatment of choice as there is no role for radiotherapy in laryngeal amyloidosis.

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