International Journal of Health Sciences and Research
www.ijhsr.org ISSN: 2249-9571

Case Report

Primary Diffuse Laryngeal Amyloidosis, a Rare Cause of Hoarseness of Voice

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ABSTRACT

Introduction: Amyloidosis is a heterogenous group of disorders characterised by deposition of amyloid proteins in various body organs. Of the numerous types, Laryngeal Amyloidosis is an extremely rare disease of unknown etiology.

Case report: We report a case of 30 year old woman who presented with hoarseness of voice and stridor necessitating a tracheostomy to secure the airway. A diagnosis of isolated laryngeal amyloidosis was made based on histopathology and thorough clinical and laboratory workup, and the patient responded to a combination therapy of oral corticosteroids and Bortezomib.

Conclusion: Laryngeal amyloidosis is a rare disease, and its diagnosis calls for a high index of suspicion as well as thorough workup to exclude systemic involvement.

Key Words: Larynx, amyloidosis, diffuse, hoarseness, stridor, bortezomib

INTRODUCTION

Amyloidosis is a benign, slowly progressive, heterogenous group of disorder characterised by extracellular proteinaceous deposits in various target organs. [1] This leads to organ malfunction and failure. Amyloidosis is derived from Greek word 'Amylon' which means starch and 'Eidos', which means resemblance. The term amyloid was first used by Virchow, because the protein showed starch like reaction when treated with iodine and sulphuric acid. [2]

Amyloidosis mainly affects 50-70 year old individuals with male: female predominance of 3:1. [1] Symptoms of amyloidosis are mainly dependant on site and size of deposits. Laryngeal involvement has been noted at all levels of larynx. Eyes, orbits, salivary glands, nose, paranasal sinuses, nasopharynx, oral cavity, bronchotracheal tree, lungs are other sites of involvement. [3]

Patients with amyloidosis of larynx present with symptoms like hoarseness of voice, cough, globus hemoptysis, stridor, dyspnoea, rarely dysphagia. Deposition of amyloid in the larynx may occur diffusely or in a single tumor nodule form. Microscopic examination typically shows acellular, homogenous and amorphous eosinophilic material displaying apple green birefringence under polarized light when stained with Congo red dye. [4]

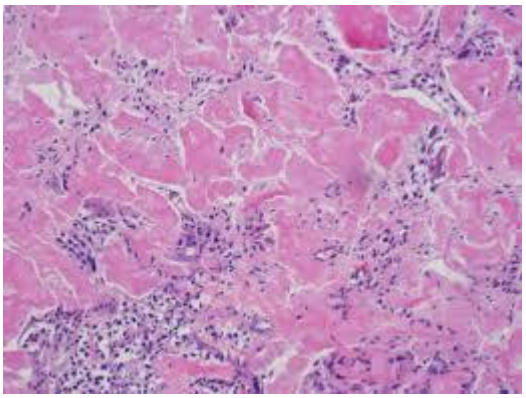

Figure1. Histological appearance of vocal cord amyloidosis showing acellular, eosinophlic amorphous deposit.

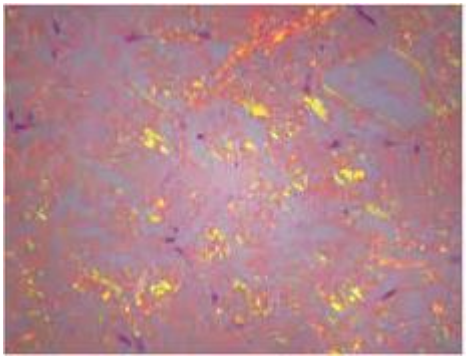

Figure2.Congo red staining of a biopsy specimen showing apple green birefringence under polarized light.

CASE REPORT

A 30 year old lady walked into our outpatient department with complaints of hoarseness of voice since two years along with shortness of breath and foreign body sensation in throat since 1 year, aggravated in past 1 month and stridor since 2 days. There were 3-4 episodes of hemoptysis also in the past one year. A chest X-Ray showed no obvious abnormalities, which to some extent ruled out any lung pathology. Indirect laryngoscopy revealed diffuse edema over bilateral vocal cords. However, both cords were mobile.

A thorough clinical examination and extensive laboratory workup ruled out systemic amyloidosis, and involvement of other organs like liver or spleen. A computerised tomography (plain + contrast) scan of neck revealed enhancing mucosal thickening involving bilateral true and false vocal cords, anterior commisure, bilateral ary-epiglottic folds extending inferiorly to involve the sub- glottic region of the larynx, with mild to moderate narrowing of the airway. It also showed few sub-centimeter sized non necrotic lymph nodes involving bilateral level II cervical regions.

A microlaryngoscopy with biopsy was planned. In view of stridor and difficult intubation an elective preoperative tracheostomy was done followed by microlaryngoscopy, which revealed severe narrowing of glottic inlet with diffuse laryngeal swelling involving bilateral cords, aryepiglottic fold extending inferiorly to the sub- glottis. De-bulking procedure was done and the specimen was sent for biopsy.

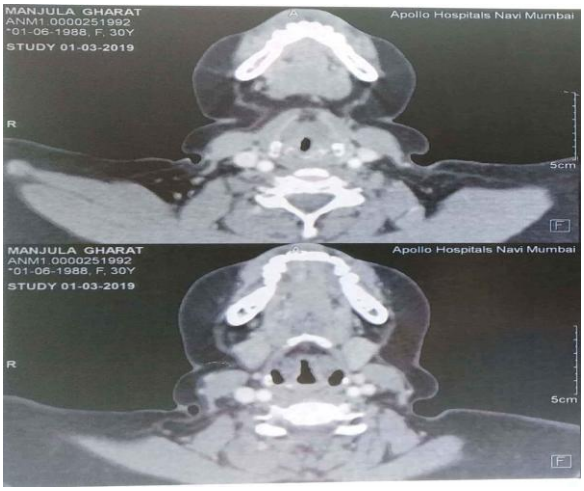

Figure3. CT Neck (P+C) showing edema of bilateral vocal cords.

On histopathological examination and staining with Congo red dye, the specimen showed apple green birefringence on polarised microscopy, which confirmed the diagnosis of laryngeal amyloidosis. Serum protein electrophoresis showed high alpha 2 levels. Serum immunofixation electrophoresis was positive for IgG band and kappa band. Serum immunoglobulin, serum light chains, serum B-2 microglobulin were present within normal limits. After a referral to haematologist who prescribed a combined therapy of oral Bortezomib and corticosteroids, the patient was discharged on a metallic tracheotomy tube. A nasopharyngolaryngoscopy performed two months later showed significant improvement and patient was then weaned off the tracheostomy.

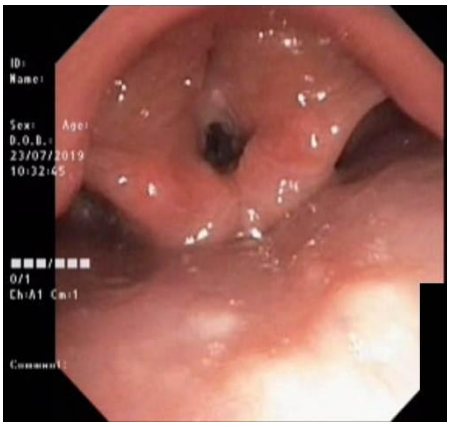

Figure4. Nasopharyngolaryngoscope showing diffuse edematous bilateral vocal cords significantly compromising the lumen.

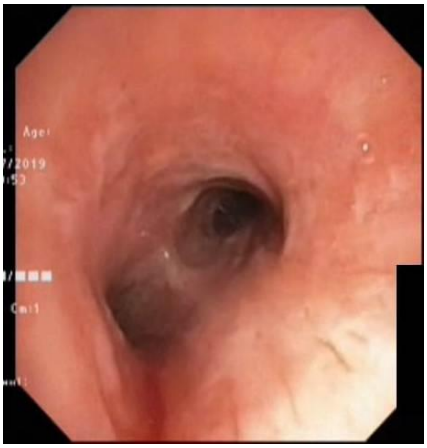

Figure5. Post tracheostomy image of the trachea and carina showing no abnormality.

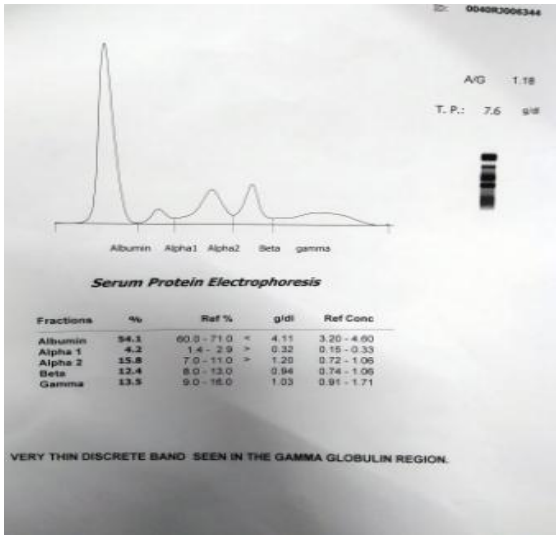

Figure 6. Serum electrophoresis was normal and was done to rule out multiple myeloma.

DISCUSSION

Amyloid fibrils are protein polymers comprising identical monomer units. Functional amyloids play a crucial role in a variety of physiological processes like long term memory and gradual release of stored peptide hormones. Amyloidosis results from the accumulation of pathogenic amyloids in a variety of tissues of the body.

Amyloidosis is diagnosed when Congo red–binding material is established in a biopsy specimen. Since different modalities of treatment exist for different types of amyloidosis, hence determining the type of amyloidosis in addition to its diagnosis is also helpful. A condition may suggest the type of amyloidosis, but the diagnosis must be established by immunostaining a biopsy specimen in order to offer definite treatment to the patient.

Amyloidosis of head and neck can be of two types, a primary isolated disease or secondary to systemic involvement. Larynx is the most common site of involvement in the head and neck region and is frequently affected by localized rather than systemic amyloidosis. Monoclonal deposits of light chain type (AL) are characteristic of laryngeal amyloidosis. [5] The ventricles and the false and true vocal cords of the larynx are the most common sites of involvement in laryngeal amyloidosis. It accounts for 0.2-1.2% of benign tumors of larynx. [6]

When a diagnosis of laryngeal amyloidosis is made, workup should include studies to rule out systemic disease, as well as a correct assessment of the laryngeal involvement. Multiple myeloma, rheumatic diseases, and tuberculosis are some of the systemic causes that must be considered as differential diagnosis. Amyloidosis related with familial syndromes and endocrinopathies, such as medullary thyroid cancer, also needs to be investigated. In our patient, after laryngeal amyloidosis was confirmed by biopsy, systemic involvement as well as any other systemic illness was ruled out.

Treatment of laryngeal amyloidosis may vary from simple observation to partial laryngectomy depending upon the level of involvement of larynx. Endoscopic CO_2 laser excision of the mass is considered be the first choice of therapy in case of localised laryngeal lesion. [7]

In AL amyloidosis, the proteasome inhibitor bortezomib has been proved effective. A systematic review and meta-analysis established that the addition of bortezomib to treatment of AL amyloidosis resulted in appreciably improved overall response rate (ORR), complete response, cardiac response rate, and 2-year overall survival; risk of neuropathy and overall mortality were both reduced. [8]

Since our patient had a diffuse vocal cord edema and not a localised mass, CO_2

laser or surgical excision was not the preferred mode of treatment. After consultation with a haematologist, the patient was started on a combination therapy of oral Bortezomib and corticosteroids to which she responded well and could be successfully weaned off the tracheostomy, after 3 months of therapy.

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How to cite this article: Wadhwa A, Dabholkar YG, Bhatt BV et.al. Primary diffuse laryngeal amyloidosis, a rare cause of hoarseness of voice. Int J Health Sci Res. 2019; 9(11):111-114.

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