

Localized laryngotracheobronchial amyloidosis: Management issues

Sir,

Amyloidosis represents a spectrum of disease characterized by abnormal deposition of insoluble extracellular proteinaceous material. Amyloidosis may affect multiple organs.^[1-3] Localized amyloidosis of laryngotracheobronchial region is rarely found and the patient usually presents with localized symptoms such as hoarseness of voice, breathlessness, and cough. We report a case of 60-year-old woman who presented with laryngotracheobronchial nodular amyloidosis that brings forth the issues in the management of such a case.

A 60-year-old-female presented to a hospital with weakness and hoarseness of voice, breathlessness and mild chronic cough for last 3–4 months. Laryngoscopic examination revealed a small nonulcerated subepithelial red mass arising from the right false vocal cord. Biopsy from the false vocal cord mass showed subepithelium composed of acellular eosinophilic material, which was congophilic on Congo red stain and showed apple-green birefringence under polarized light, suggestive of amyloid. Diagnosis of vocal cord amyloidosis was given. Serum protein electrophoresis ruled out multiple myeloma. She was given symptomatic treatment and discharged under follow-up.

She again presented with similar complaints 3 months later. Contrast-enhanced computed tomography (CT) neck, chest, and abdomen revealed focal bulge in the right false vocal cord, mild circumferential thickening of the tracheal wall with specks of calcification along with a nodule in posterior basal segment of the left lower lobe. On bronchoscopy, multiple, variably-sized subepithelial, nonulcerated nodules were noted on false vocal cords bilaterally, trachea, and right main bronchus [Figure 1a-c] and CT showed nodularity and thickening of tracheal wall [Figure 1d]. Bronchial biopsy was done, and section showed tissue lined by respiratory epithelium with extensive deposition of amyloid [Figure 2a and b]. These deposits also showed evidence of amyloid on Congo red and methyl violet [Figure 2c and d]. Histological diagnosis of bronchial amyloidosis was made.

Her pulmonary function test showed truncation of flow volume loop; however, all values were both above lower limit of normal and >80% of predicted values [Figure 1e-f]. A final diagnosis of localized laryngotracheobronchial amyloidosis was given. The patient is under regular follow-up for 1 year, with similar Pulmonary function test (PFT) values.

Amyloidosis can be systemic or localized according to clinical involvement. The localized form may remain confined to the gastrointestinal system, genitourinary system, lungs, skin, or respiratory tract.^[1-4]

Pulmonary amyloidosis can have four different manifestations: diffuse parenchymal amyloidosis, nodular parenchymal amyloidosis, infiltrative tracheobronchial amyloidosis, and nodular tracheobronchial amyloidosis.^[1] Our patient had localized nodular tracheobronchial amyloidosis associated with bilateral laryngeal nodular amyloid deposits on false vocal cords. Diffuse nodularity and thickening in the tracheobronchial tree along with areas of bronchial narrowing may result in complications such as atelectasis and postobstructive pneumonia.

Amyloidosis confined to the respiratory tract is rare and is affected by primary type.^[1] Localized amyloidosis in airway may occur due to ineffective clearing of light chains produced by plasma cells in mucosa-associated lymphoid tissue or may be plasma cell reaction caused by inflammatory antigens deposited locally.^[2] The larynx is the most common site of localized amyloidosis in the head and neck region followed by ventricle, vestibular folds, vocal folds, epiglottis, aryepiglottic folds, and subglottis.

The patient with respiratory tract amyloidosis usually presents with hoarseness, progressive dyspnea, abnormal phonation, hemoptysis, and dysphagia. Our patient presented with weakness and hoarseness of voice, breathlessness, and mild chronic cough. She was initially diagnosed with laryngeal amyloidosis only, and the workup for a systemic disease was found to be negative. She was evaluated for tracheobronchial involvement later when her complaints recurred. In a case series of 32 patients, synchronous or metachronous laryngeal and tracheobronchial tree was observed in 47% cases while lung involvement associated with airway disease was seen only in one patient.^[3] CT scan is more sensitive than chest radiograph in detecting the respiratory tract deposits in amyloidosis, although the findings are nonspecific.

Currently, the objective of treatment is to maintain luminal patency through surgical or Nd-YAG laser or cold knife.^[3,4] External beam radiation along with airway stenting, tracheostomy, partial or total laryngectomy has also been attempted with some success.^[5] Up to 50% of those who undergo these procedures may have relapse after surgical excision of the lesions.^[5] Therefore, a major dilemma in these patients is the timing of intervention.

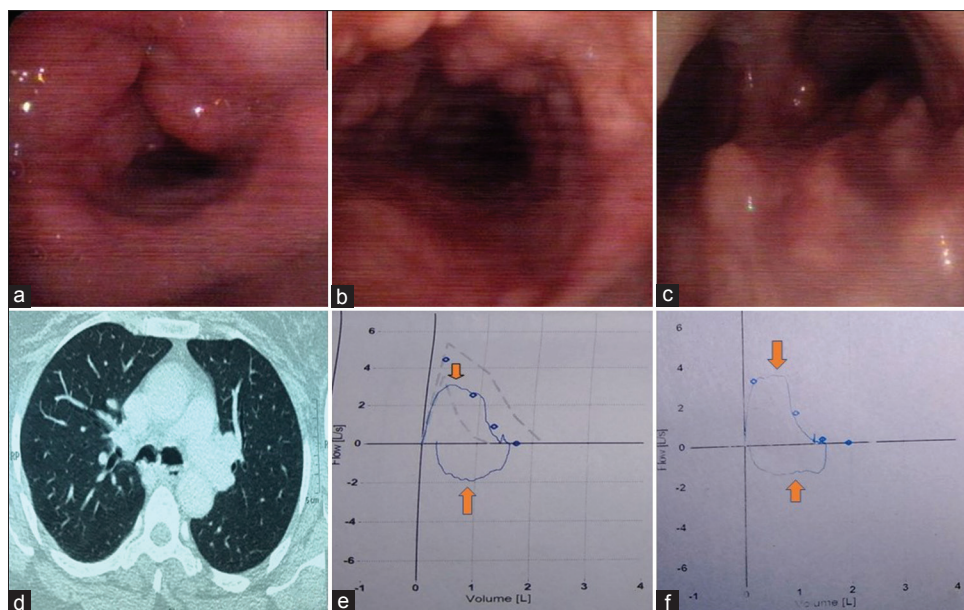


Figure 1: (a) Bilateral laryngeal nodules; (b) trachea studded with amyloid nodules; (c and d) nodules in trachea, at carina, and in right main bronchus with corresponding computed tomographic section (d) showing nodularity and thickening of posterior tracheal wall. (e and f) Truncation of flow volume loop in laryngotracheobronchial amyloidosis with progressive flattening of expiratory and inspiratory curves over 9 months

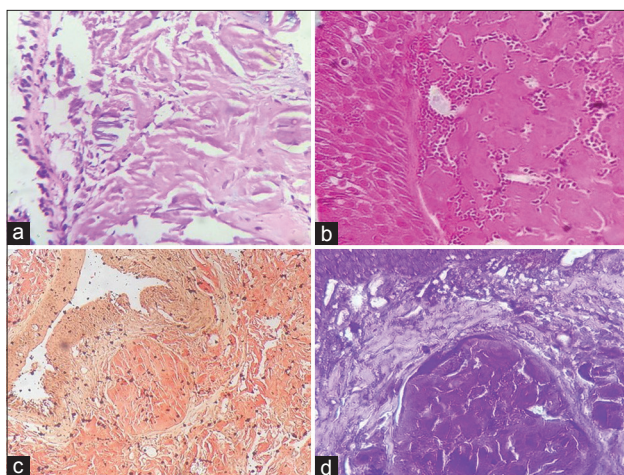


Figure 2: (a and b) Sections showing subepithelial dense deposits of amyloid (H and E, $\times 400$); (c and d) amyloid deposits highlighted by Congo red (c) and methyl violet (d) stains, respectively ($\times 400$)

Spirometry is not a good tool to monitor these patients as large airway obstruction presents late. Monitoring with CT or magnetic resonance imaging (MRI) is more objective than bronchoscopic examination. However, CT has a significant radiation exposure, MRI is expensive and time-consuming, and bronchoscopy is invasive, so there is no single modality for monitoring that is ideal.

Our case emphasizes that when a diagnosis of laryngeal amyloidosis is made patient should be simultaneously evaluated for tracheobronchial involvement as it may coexist because of likelihood of the same etiopathogenesis. Management needs multidisciplinary approach, highly specialized intervention, and patient's

informed will. It can be said that a benign disease that needs resource intensive management has a malignant prognosis in a resource-limited setting.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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Case Letters

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