Case report

An unusual case of hoarseness of voice

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ABSTRACT

Amyloidosis is a condition wherein there is an over-expression of specific proteins culminating in the extracellular deposition of insoluble beta pleated sheets of fibres. These deposits disrupt function of the target organ. Its aetiology remains unknown. Primary Amyloidosis localized to the trachea and bronchus is a rare entity. It produces tumour like lesion in the tracheo-bronchial tree. We hereby discuss a patient who presented with persistent hoarseness of voice who was referred to a tertiary care centre to rule out malignancy.

1. Introduction

Primary tracheal or endobronchial lesions may be benign or malignant. The definitive diagnosis of tracheobronchial lesions is frequently delayed, since the signs and symptoms caused by these lesions are quite nonspecific and chest radiographs are rarely diagnostic [1]. This case report is an attempt to make the clinicians aware that not all cases of tracheo-bronchial mass are malignant.

2. Case

An 80 year old male farmer, presented with a dry cough, hoarseness of voice and shortness of breath for 6 months. The breathlessness was gradually progressive MMRC grade II to III. There was no orthopnea or paroxysmal nocturnal dyspnoea. There was no haemoptysis, sputum production, chest pain, wheezing, or fever. He had loss of weight of 10 kg in 6 months and associated loss of appetite. He denied any dysphagia, abdominal pain, nausea, vomiting, swelling of the limbs, jaundice, and altered sensorium.

He had a past history of pacemaker insertion and hypertension for 10 years treated with amlodipine. He was diagnosed with Parkinson's disease for the last 5 years and was on treatment with levodopa/carbidopa combination.

He was an ex-smoker with a 40 pack year history of smoking. He left smoking for the last 5 years.

On examination, his BMI was 16; he was in respiratory distress requiring oxygen support with 4L/min through nasal prongs. General examination revealed pallor, with no palpable lymph nodes, clubbing, cyanosis or oedema. There was decreased air-entry in the right lung base. All other systemic examination was normal.

Routine investigation showed haemoglobin of 10 gm/dl, Chest radiograph showed a right lung basal haziness. The CT scan showed a large right pleural effusion with no abnormality in the mediastinum or bilateral lung parenchyma as described in Fig. 1.

Diagnostic thoracocentesis of 500 ml of haematogenous fluid from the right side of the chest failed to give any definitive diagnosis. Medical pleuroscopy of the right hemithorax revealed multiple nodules in the right lung parenchyma but the parietal pleura appeared normal. Multiple parietal pleural biopsies were taken.

Flexible fibre optic bronchoscope to visualize the bilateral vocal cords and endobronchial tree revealed a fleshy, hyperaemic mass lesion just above the true vocal cord in the larynx, as shown in Fig. 2 and an endo-bronchial lesion which was a hyperaemic lobular mass lesion in the right upper lobe apical segment. This endo-bronchial lesion was infiltrating the right bronchial tree apical segment of the right upper lobe as described in Fig. 3. Both the lesions were bleeding on touch. Multiple biopsies were taken from the lesion above the vocal cord and also from the endo-bronchial mass.

Histopathology of both the lesions revealed homogenous eosinophilic material deposition in the wall of blood vessels. This material was congophilic and gave apple green birefringence under polarized light as described in Fig. 4. Based on the pathology findings a diagnosis of Amyloidosis was confirmed. This material was also noticed on the surface of the pleural biopsy specimen.

Considering his age, co-morbid condition, overall quality of life and side effects of treatment, the patient, his family members decided on conservative management only. He was started on oral colchicine therapy and discharged in a stable condition.

3. Discussion

Amyloidosis is a group of clinical syndromes caused by deposition of...
insoluble amyloid fibrils in extracellular matrix. This in turn produces mass effects and cytotoxicity, thereby damaging normal tissue and organ structures leading to target organ dysfunction [2].

Primary tracheo-bronchial amyloid (TBA) accounts for 1.1% of all amyloidoses [3]. This was first reported around 50 years ago. In contrast to systemic amyloidoses, the other organs are spared from the disease. With the increased use of flexible fibre-optic bronchoscopy and computed tomography (CT) imaging in the past two decades, the number of reported cases has increased significantly [4]. Clinical presentation varies from chronic cough, dyspnoea, haemoptysis and even stridor. The final diagnosis is made during bronchoscopy, which includes bronchial biopsies revealing positive Congo red staining [5] as in our patient. Thus bronchoscopy remains the cornerstone in the diagnosis of TBA that allows better visualization of the lesions and has the advantage of allowing excision of amyloid deposits for histopathological analysis [6].

The treatment of primary tracheo-bronchial amyloid remains a challenge. There are various modalities that have been reported in literature which includes mechanical debulking, laser therapy, radiation therapy. All these options are directed towards maintaining the patency of the airway [7].

Although TBA is localized disease, two series reported low survival rate like 31% (13 of 41 patients) and 43% (3 of 7 patients). Other case series with mid airway diseases demonstrated that 7 of 15 patients (47%) progressed to respiratory failure. According to these reports, the average lifespan of these patients was only about 9 years after diagnosis. Morbidity and mortality were associated with quantity and rate of amyloid deposition in the airways [8].

Primary tracheo-bronchial amyloid may be an under diagnosed and hence under reported entity. In patients with difficult to treat asthma or chronic cough of unknown aetiology, or persistent dyspnoea without an identifying cause, recent hoarseness of voice; a more extensive workup, including flexible fibre optic bronchoscopy and computed tomography of the chest, would enable the identification of rare diseases including primary tracheobronchial amyloidoses [9]. Our current case report is just an attempt towards increasing the awareness amongst the physician.

4. Conclusion

Primary tracheo-bronchial amyloid needs a multidisciplinary approach, including interventional pulmonology, radiology and pathology for optimal management. Although this is a rare clinical condition, its importance is discussed regarding the differential diagnosis of localized tracheal and endo-bronchial lesions.

Disclosure of conflict of interest

None.
Fig. 4. Legend: Histopathology revealed homogenous, eosinophilic material deposit in the wall of the blood vessels. This material was congophilic with apple green birefringence under polarized light.

Disclosure of financial interest

None.

Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.rmcr.2018.01.013.

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