CASE REPORT

Tracheobronchial Amyloidosis Masquerading as Bronchial Asthma

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ABSTRACT

A case of localized tracheobronchial primary amyloidosis masquerading as "bronchial asthma" is presented. Computed tomography of the chest and fiberoptic bronchoscopy image are included. Tracheobronchial primary amyloidosis is a rare, but potentially curable cause of airway obstruction mimicking asthma.

Key word : *Tracheobronchial amyloidosis.*

[Indian J Chest Dis Allied Sci 2004; 46 : 117-119]

INTRODUCTION

Primary amyloidosis involving the tracheobronchial tree is extremely uncommon. It produces tumour-like lesions in the tracheobronchial tree. The clinical presentation includes symptoms such as dysponea, cough, haemoptysis, stridor and wheezing. We present a case report of localized tracheobronchial primary amyloidosis masquerading as "bronchial asthma".

CASE REPORT

A 50-year-old nonsmoker male was symptomatic with episodic cough and wheezing for the last five years. He was being treated as a case of "bronchial asthma" in another hospital with partial response to oral and inhaled bronchodilators. He was also complaining of intermittent streaky haemoptysis for the same duration. The patient reported to our hospital for the treatment of acute exacerbation of his symptoms. On examination, he was in respiratory distress. Stridor was audible over the trachea and rhonchi were auscultated bilaterally. Examination of other systems was not remarkable. Investigations revealed hemoglobin 13 g/dl with a total leukocyte count of 15×10^9/L (82% neutrophils, 18% lymphocytes). The ESR was raised at 51 mm/hour (Westergren's method). All biochemical investigations were within normal limits. The chest radiograph was normal[1]. Computed tomography of the chest showed circumferential thickening of the tracheobronchial tree [Figures 1(a), 1(b)]. Pulmonary function test revealed the following (as % predicted) FVC 52%, FEV_1 37%, FEV_1/FVC 73%, PEFR 33%, FEF_{25-75} 17%, FEF_{50} 14% and FEF_{75} 11%. However, there was no

[Received : August 20, 2002; accepted after revision : December 4, 2002]

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reversibility of the airway dysfunction. Arterial blood gases, while breathing room air, were PaO_2 7.49 kPa (normal range 11.3-12.6 kPa), $PaCO_2$ 4.81 kPa (normal range 4.7-6.0 kPa).

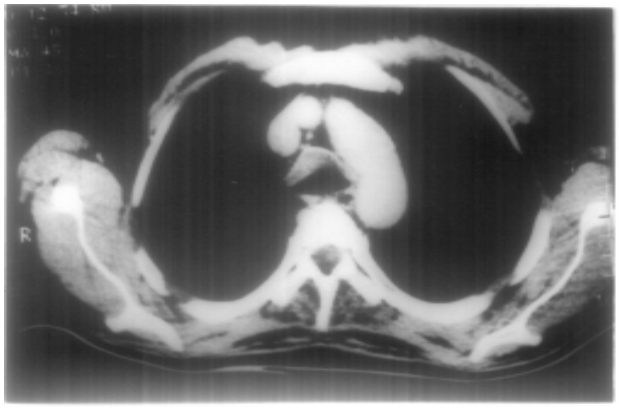

Figure 1(a). Contrast enhanced CT scan at the level of aortic arch showing diffuse circumferential thickening of the tracheobronchial airway just above the carina.

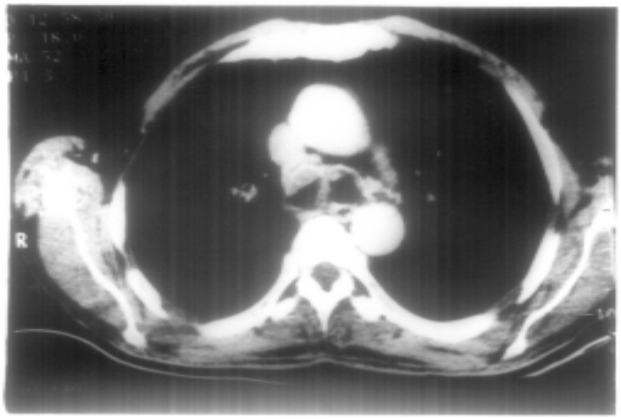

Figure 1(b). Contrast enhanced CT scan just below the carina showing diffuse circumferential wall thickening in both right and left main bronchi.

In view of the computed tomography findings, he was subjected to a fibreoptic bronchoscopy examination. It revealed yellowish, flat submucosal lesions starting at the level of the vocal cords. In addition, there were friable and glistening yellowish polypoid projections from the walls of the trachea (Figure 2). The carina was also covered with these projections. The rest of the bronchial tree could not be evaluated due to marked narrowing of both main bronchi by these projections. Histo-

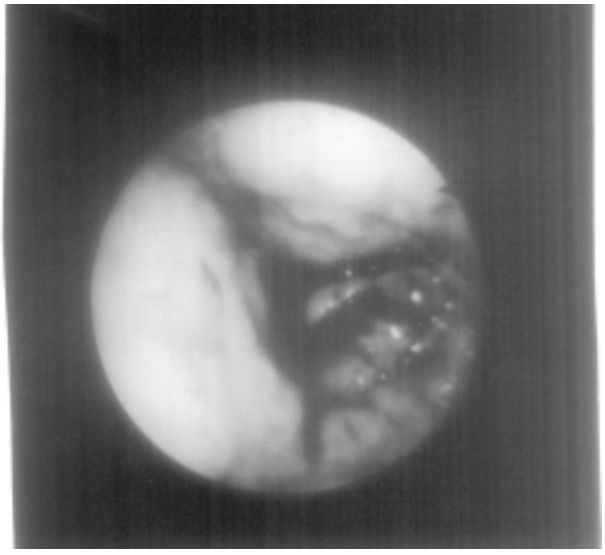

Figure 2. Fiberoptic bronchoscopy view shows glistening yellow polypoid projections from the tracheal wall.

pathology of the biopsy specimen from these projectios confirmed the presence of amyloidosis. There were no Bence Jones proteins in urine. Serum protein electrophoresis result was normal. Electrocardiogram and echocardiogram were normal. A rectal biopsy specimen did not show any evidence of rectal amyloidosis. The patient was treated with amoxycillin-clavulanate combination, intravenous corticosteroids, oral and inhalational bronchodilators. Thereafter, bronchoscopic piecemeal excision of nodular amyloidosis was performed. A few weeks later, the patient was also subjected to neodymium:yttrium-aluminium-garnet (Nd YAG) laser treatment. A repeat bronchoscopy examination did not show much improvement. Subsequently, he was started on melphalan (0.25 mg/kg/day) and prednisolone (2 mg/kg/day) for four days every month. In addition, colchicine 0.5 mg twice a day orally was also administered. He was reviewed at three months. There was an excellent clinical response. There were no stridor or rhonchi and he did not require any bronchodilator therapy. A repeat pulmonary function test revealed the following (as % predicted): FVC 95%, FEV_1 97%, FEV_1/FVC 83%, PEFR 80%, FEF_{25-75} 98%, FEF_{50} 74% and FEF_{75} 118%. On bronchoscopy examination, there was a

significant degree of regression of lesions. Follow-up bronchoscopy after six months revealed almost normal tracheobronchial tree.

DISCUSSION

Amyloidosis is characterized by insoluble protein fibrils with characteristic physico-chemical properties. Amyloidosis may be primary or secondary to other diseases. Localized amyloidosis has not been chemically identified but is usually defined by the absence of systemic features.

Primary amyloidosis involving the tracheo-bronchial tree is rare. It produces tumour-like lesions in the tracheobronchial tree. The patients present with a variety of symptoms such as dyspnoea, cough haemoptysis, stridor, rhonchi and crepitations. These nodular lesions produce progressive airway obstruction resulting in symptoms suggestive of bronchial asthma, atelectasis and obstructive pneumonitis.

Management includes endoscopic excision of amyloid deposits, thoracotomy and even radiotherapy[2]. Spontaneous regression of diffuse tracheobronchial amyloidosis has also been described[3]. Fatal haemorrhage can occur following endoscopic piecemeal excision of amyloid nodules[4]. Bronchoscopic laser excision is considered to be a better option of treatment as amyloid is very sensitive to laser photo-irradiation. Repeated sessions are required and recurrence is rare. Carbon dioxide laser may have an advantage over the neodymium YAG laser in the management of localized tracheobronchial amyloidosis because of lesser bleeding[5]. Successful treatment of primary amyloidosis with intermittent chemotherapy consisting of melphalan and prednisolone has also been reported[6]. However, this mode of treatment has not been tried earlier in localized tracheobronchial amyloidosis. Recently, Aggarwal *et al*[7] have described two patients with tracheobronchial amyloidosis. Both these patients were reported to be doing well without any definitive treatment.

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