A CASE OF PRIMARY DIFFUSE TRACHEOBRONCHIAL AMYLOIDOSIS TREATED BY LASER THERAPY

J C H Yap, Y T Wang, S C Poh

ABSTRACT
We report a case of primary diffuse tracheobronchial amyloidosis in a 72-year-old lady who presented with a long history of recurrent cough, dyspnoea, wheezing, haemoptysis and chest infection. She was treated successfully with three sessions of laser therapy. There were improvements in both clinical symptoms and measurements of airway obstruction. Bronchodilators and oral prednisolone were not required after treatment.

Keywords: Tracheobronchial Amyloidosis, laser therapy

INTRODUCTION
Amyloidosis is characterized by the extracellular deposition of the substance amyloid, which emits a unique green birefringence when viewed under a polarizing microscope after staining with Congo Red. Amyloidosis localized to the lower respiratory tract is rare; the first case was reported by Lesser in 1877\(^\text{1}\). We describe the first case of primary diffuse tracheobronchial amyloidosis in Singapore and its treatment by photocoagulation therapy using a neodymium:yttrium-aluminium-garnet (Nd:YAG) laser to relieve airway obstruction.

CASE REPORT
This Chinese woman first presented to our department in April 1977 at the age of 58 years. She complained of haemoptysis associated with pleuritic pain. She had a history of chronic productive cough for 20 years and had been treated by general practitioners whenever there were exacerbations. There was no history of any severe childhood respiratory illness or sinusitis. She was a non-smoker. Physical examination was normal. Chest radiograph showed some areas of atelectasis in both bases. She was diagnosed to have bronchiectasis and given a course of antibiotics.

She defaulted follow up but was again admitted in 1984 for dyspnoea at rest as well as haemoptysis. She developed type II respiratory failure with hypotension. Bilateral rhonchi were heard over both lungs. Chest radiograph was normal. She made rapid recovery with a short duration of respiratory support and eventually was well enough to be discharged with bronchodilators and oral steroids. The blood gas on discharge: arterial oxygen tension (PaO\(_2\)) of 54.4 mmHg and arterial carbon dioxide tension (PaCO\(_2\)) of 41.5 mmHg on atmospheric air. Her forced expiratory volume in the first second (FEV\(_1\)) was 0.55 litre, forced vital capacity (FVC) 1.43 litres and FEV\(_1\)/FVC 39%. There was no bronchodilator response. She continued to have wheezing episodes despite medications. She was last seen in May 1985 after which she did not turn up for further follow up.

She had recurrent attacks of dyspnoea which were often aggravated by upper respiratory tract infection. She was admitted in March 1987 for a severe episode of breathlessness associated with wheezing, productive cough and fever. She was in hypercapnic respiratory failure with acidosis. Leukocytosis was present. Her chest radiograph showed pneumonic patches in both bases and right lower lobe collapse. The airway obstruction was not relieved by bronchodilator. She was intubated and treated for status asthmaticus. She then developed a right middle lobe collapse. In view of this, a bronchoscopy was performed. The mucosa of the tracheobronchial tree was oedematous and erythematous. The orifice of the right upper lobe was obstructed by a "swollen mucosa". Aspirate grew Klebiella and Pseudomonas. Acid fast bacilli and malignant cells were not detected. Chest radiograph showed re-expansion of the collapsed lobes after the lavage during bronchoscopy. She was weaned off the respirator but after eight days, she had a similar episode of dyspnoea associated with collapse of the same lobe. A second intubation was required for respiratory support and bronchial lavage was performed again. Thick inspised mucus was reported. The Fig 1 - Chest radiograph (1988) showing lingular opacity

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mucosa was again noted to be oedematous. She was eventu-
ally weaned off the respirator after three weeks. Her blood gas 
on discharge: PaO₂ = 71.8 mmHg and PaCO₂ = 45.4 mmHg on 
air. The chest radiograph showed lingular opacities and right 
basal atelectasis. She defaulted follow up again.

In June 1988, she was referred for investigation of a lingular 
opacity (Fig 1). Sputum smear for acid fast bacilli was negative. 
An empirical course of anti-tuberculous treatment was insti-
tuted without response. The sputum culture for acid fast bacilli was

Fig 2 - Picture taken during bronchoscopy showing 
distortion of the mucosa with granules and plaques

also negative. In view of this, a bronchoscopy was carried out. 
The mucosal lining was noted to be very granulomatous (Fig 2). The 
orifice of the right upper lobe was partially obstructed by a 
huge granule. Several biopsies were taken from this granule. 
The orifices of the rest of the respiratory tree were obstructed by 
submucosal plaques as well as smaller granules. Histology 
showed hyperplastic bronchial mucosa with globular masses of 
amorphous material in the submucosal layer. The stain for 
amyloid was positive (Fig 3).

Retrospectively, our patient had no symptom of chronic 
extrapulmonary disease or systemic involvement by amyloid. 
Her urine analysis did not show the presence of Bence Jones 
protein and electrophoresis was normal. Both serum protein 
and immunoglobulin electrophoresis did not show any abnor-
mality. The ESR was 18 mm per hour. Liver function tests 
were normal. Two dimensional echocardiography of the heart 
did not show any amyloid deposit. The CT Scan of the thorax 
showed modular appearance and thickening of the tracheal 
wall as well as the lingual bronchus which was also narrowed 
with associated collapse and consolidation. Bone marrow and 
rectal biopsies were not done.

She had two more admissions for acute exacerbation of 
breathlessness with chest infection before she agreed to laser 
therapy in September 1989. The Nd-YAG laser was used and 
the procedure was performed under general anaesthesia during 
the first and second sessions and under local anaesthesia dur-
ing the third session. We used a power of 30 watts for laser 
burn of one second duration. The total joules used for each 
session was 1185, 1075 and 2787 respectively. All three ses-
sions were not associated with any complication. Her symp-
toms of dyspnoea and wheezeing were markedly reduced. She 
was eventually taken off bronchodilators and steroids and has 
remained relatively well. Her lung function results are shown 
in Table I.

Table I - Showing Lung Function Tests Before and After 
Laser therapy

<table>
<thead>
<tr>
<th></th>
<th>Before</th>
<th>After</th>
<th>Percentage Change</th>
</tr>
</thead>
<tbody>
<tr>
<td>VC (L)</td>
<td>1.86 (76.5)</td>
<td>2.07 (85.2)</td>
<td>11.3</td>
</tr>
<tr>
<td>FEV1 (L)</td>
<td>1.07 (54.6)</td>
<td>1.53 (78.1)</td>
<td>43.0</td>
</tr>
<tr>
<td>FEV1 (%)</td>
<td>57</td>
<td>73</td>
<td>28.1</td>
</tr>
<tr>
<td>MMFR (L/sec)</td>
<td>0.77 (30.1)</td>
<td>1.23 (48.0)</td>
<td>59.7</td>
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</table>

Parentheses indicate percent of predicted normal

DISCUSSION

Amyloidosis is a rare condition which may be widespread 
throughout the body or confined to an organ. Primary pulmo-

nary amyloidosis presents in two main forms: tracheobronchial 
type with multifocal submucosal plaques and tumour-like 
masses or the parenchymal type as multiple or solitary nodules 
or diffuse alveolar septal infiltrations. Primary pulmo-

tary amyloidosis is seen more frequently than bronchial trophic 
and the incidence is higher in males. The duration of symptoms 
is quite variable in the cases reported in the literature so far.

The clinical presentation depends on the type of pulmo-

nary amyloidosis. The diagnosis of amyloidosis in the respira-
tory tract may be difficult as it can simulate bronchiectasis, 
tuberculosis, bronchial carcinoma, asthma and many other res-
piratory diseases. In our case, the patient was first diagnosed 
to have bronchiectasis and subsequently bronchial asthma. She 
was also treated empirically for tuberculosis. Finally a diagno-

sis of amyloidosis was made after a bronchoscopic assess-
ment. Patients with tracheobronchial submucosal plaques may complain of cough, dyspnoea, stridor or haemoptysis. Haemoptysis may become life-threatening as the disease progresses. They frequently suffer from repeated episodes of bronchial infection and may eventually develop areas of bronchiectasis. Bronchoscopy is useful and important in the diagnosis. Characteristically, the lesions are widely dispersed, irregular in size and shape and diverse in colour. They may vary from small, flat elevations to broad plaques and longitudinal folds. Biopsy is required for histological confirmation[19]. Bronchography may show tracheal lesions encroaching upon the main bronchi and occupying most of its length. The walls of the lumen are frequently thinned and the lumen can be completely occluded. The chest radiograph may be normal or show areas of atelectasis secondary to obstruction by amyloid lesions[20]. In contrast, the endobronchial tumour-like masses usually occur in the main bronchi as in our case. Hence the clinical features, radiographic changes and macroscopic appearance at bronchoscopy may be indistinguishable from that of a neoplasm. Of the two forms, patients with tracheobronchial submucosal plaques may be associated with tracheobronchopathia osteoplastica in the long run[14-15]. Patients with parenchymal nodules are usually asymptomatic and referred for investigation after an incidental finding on their chest radiograph. The nodules may be solitary or multiple, and may affect both lungs. They are usually peripheral and subpleural, ranging in size from 1 to 15 cm in diameter. There may be cavitation or calcification in up to one third of them. The group with the diffuse parenchymal lesions may present with an abnormal chest radiograph or more frequently, they complain of progressive dyspnoea. The radiographic abnormality may be confused with pulmonary oedema or with pulmonary fibrosis.

The treatment for pulmonary amyloid depends on the type. In our case, bronchoscopic laser cautery was the treatment of choice[16-18]. The amyloid is very sensitive to laser photoablation with a power setting of less than 30 watts. This contrasts with pulmonary neoplastic tissue which frequently requires 70 watts or greater to result in coagulation and necrosis[20]. Fissural necrosis does not occur following photoablation but a membrane of coagulated tissue is formed. It is not associated with severe bleeding as in bronchoscopic piecemeal resection because the Nd-YAG laser coagulates small vessels[21,22]. Recurrence is very rare[18,19]. However, repeated sessions are required[22]. The treatment for the other forms include total resection in the nodular form as well as corticosteroid, immunosuppressants and radiotherapy but with limited success[23].

Pulmonary amyloid may be more common than is generally recognised. It can mimic various diseases and should therefore be thought of when a clinical diagnosis proves difficult to confirm on initial investigation.

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REFERENCES