

Amyloidosis of the respiratory tract treated by laser therapy

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We describe a case of amyloidosis localised to the bronchial tree that was treated by laser therapy. The initial diagnosis was thought to be bronchogenic squamous carcinoma. This case shows that amyloid is an unusual cause of bronchial obstruction that may simulate bronchogenic carcinoma and that in certain cases bronchoscopic laser cautery may be appropriate.

Case report

A 39 year old woman was admitted to hospital complaining of chest pain, breathlessness, cough, and haemoptysis. She had been discharged three days previously after an ovarian cystectomy. The past medical history included a hysterectomy for an ovarian cyst complicated by a pulmonary embolus 10 years before this admission. In the intervening time she had been perfectly well. She smoked five cigarettes daily, occasionally with marijuana.

On examination she was distressed with stridor and tachypnoea (30/min) but afebrile and not cyanosed. Expansion and breath sounds were reduced over the right hemithorax. The pulse rate was 120 beats/min and blood pressure 120/80 mm Hg, and there were no signs of heart failure. Forced vital capacity (FVC) was 1.6 litres (predicted 3.85 l) FEV₁ was 1 litre (predicted 2.72 l) and peak expiratory flow rate (PEFR) was 200 l/min⁻¹. An electrocardiogram showed a sinus tachycardia with normal axis and QRS complex. A radiograph was normal. A ventilation-perfusion lung scan showed virtual absence of ventilation of the right lung, with normal perfusion and no abnormality of the left lung. In view of these findings bronchoscopy was performed. The vocal cords and trachea were normal but the right main bronchus was almost completely occluded by apparent submucosal infiltration and swelling. The bronchial mucosa was reddened and oozed blood between nodules of pale, malignant looking tissue. Further nodules were seen at the carina extending into the left main bronchus. Multiple biopsy specimens and bronchial washings were taken.

Microscopy of the bronchial biopsy specimens showed chronically inflamed mucosa and foci of squamous metaplasia with some infiltrating cells strongly suggestive of invasive squamous cell bronchogenic carcinoma (figure, A). Bronchial washings were reported as showing large

abnormal cells suggestive, but not diagnostic, of carcinoma.

In view of these findings the patient was started on oral dexamethasone 4 mg four times daily and referred for urgent radiotherapy, which continued for two weeks. She responded quickly to treatment and within 48 hours her breathlessness and haemoptysis had ceased. Before discharge from hospital the FVC was 3.1 litres; FEV₁ 2 l and peak expiratory flow rate (PEFR) 200 l min⁻¹. Two months after discharge she presented with further haemoptysis and increasing breathlessness. A repeat V-Q scan showed absent ventilation of the right upper lobe. Bronchoscopy again showed inflamed bleeding mucosa over the carina and the origin of the right upper lobe with islets of pale, malignant looking tissue.

Microscopy of biopsy specimens again showed considerable squamous metaplasia but also a nodule of congophilic and thioflavin T positive material (figure, B). These changes are diagnostic of amyloidosis and review of the first biopsy specimens showed amyloid to be present in small amounts. Immunochemical staining showed this to contain the AL subunit protein. The urine was negative for Bence-Jones protein and a rectal biopsy specimen was negative.

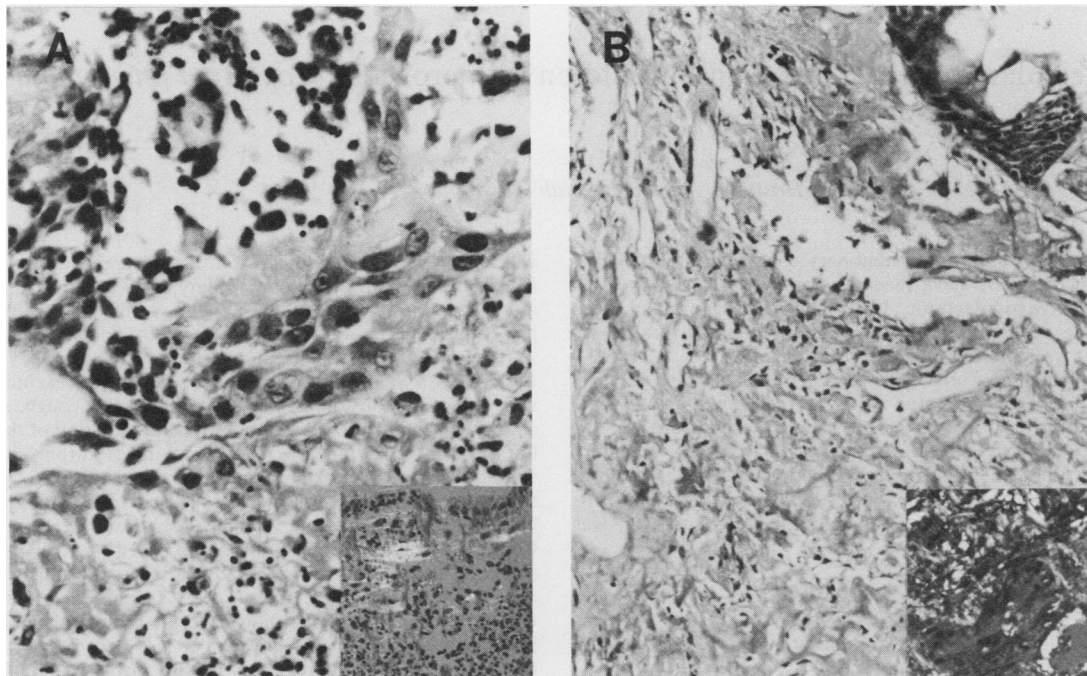
An attempt at bronchoscopic removal of the obstructing tissue during rigid bronchoscopy under general anaesthetic was impeded by bleeding. The patient was referred for laser therapy to relieve the haemoptysis. Laser therapy was given twice, a neodymium-yttrium-aluminium-garnet crystal laser (Barr and Stroud Fiberlase 100) being used, to photocoagulate the sessile endobronchial lesions. After treatment the haemoptysis and breathlessness ceased. At follow up 18 months after presentation the patient's FVC (3.8 l), FEV₁ (2.2 l), and PEFR (234 l min⁻¹) remained satisfactory. There has been one further episode of blood streaking the sputum, which resolved with antibiotic treatment.

Discussion

This case of primary tracheobronchial amyloid is unusual in that the onset was acute and the patient was young (the mean age at presentation being 53 years) and female, the disease being commoner in men. The diagnosis is histological, the clinical appearances being often highly suggestive of malignancy. Squamous metaplasia is a well recognised feature and a diagnostic trap histologically.² Traditionally, amyloid has been classified as primary or secondary, but now differences in the amyloid fibrils have led to a chemical classification of clinicopathological entities.³ Two major

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(A) Fragment of one of the initial biopsy specimens (A) showing metaplastic and dysplastic squamous epithelium, overlying degenerative and inflamed bronchial mucosa. (Haematoxylin and eosin, $\times 370$.) Inset: A small amount of birefringent amyloid is present. (B) Specimen from the second biopsy showing squamous metaplasia with submucosal eosinophilic material and chronic inflammatory cells. (Haematoxylin and eosin, $\times 160$.) Inset: A large amount of amyloid is present. (Both Congo red, $\times 160$, and cross polarised filters.)

subunit proteins, AA and AL, have been identified. Protein A is thought to be derived by proteolysis from certain immunoglobulin light chains and AA proteins from serum acute phase protein.³ AI is found in primary amyloid, as in this case.

The mainstay of treatment is symptomatic, although corticosteroids, colchicine,⁴ melphalan,⁵ radiotherapy,⁶ and pneumonectomy⁷ have been used. Intermittent bronchoscopic resection may be successful⁸ but in our patient it was limited by local bleeding. Laser therapy may be a useful method in endobronchial obstructing malignancy.⁹ In our patient it gave considerable symptomatic relief and improvement in lung function.

This case illustrates that primary pulmonary amyloidosis, though rare, may simulate carcinoma clinically and is usually associated with squamous metaplasia, leading to diagnostic difficulty. Laser therapy may be the treatment of choice in some patients with diffuse tracheo-bronchial amyloid, though our current experience is very limited.

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