Mesenchymal cystic hamartoma of the lung: a rare cause of relapsing pneumothorax

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Abstract
A 14 year old boy is described with recurrent spontaneous pneumothoraces due to a mesenchymal cystic hamartoma, a very rare disease with a multicentric nature and a benign course in most patients.

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We present a very unusual cause of relapsing pneumothorax due to a mesenchymal cystic hamartoma of the lung, a distinct clinicopathological entity first described by Mark in 1986.

Case report
A 14 year old Turkish boy, born in the Netherlands, was admitted for a left spontaneous pneumothorax once in August and twice in September 1990. They were unsuccessfully treated with suction, and later by pleurodesis with talc, tetracycline, and blood. The patient had one small haemoptysis, but denied dyspnoea, fever, night sweats, or chills. His past was unremarkable with no history of tuberculosis, recurrent respiratory infections, or smoking. Physical examination revealed dullness and increased breath sounds at the left base. Routine biochemical studies, haematology and urine analysis were normal. The chest radiograph and computed tomographic scan showed a partial left pneumothorax and an atelectatic left lower lobe. Bronchoscopy revealed normal airways. In October 1990 a thoracotomy was performed for persistent pneumothorax. The left lower lobe was collapsed and multiple adhesions and a thickened pleura were found, but no bullae or blebs. In the lateral part of the lower lobe a bloody crater-like defect was found from which a biopsy sample was taken. The initial diagnosis of this biopsy material was pulmonary blastoma. For complete surgical removal of the tumour the patient was referred to the University Hospital in Nijmegen. A lobectomy was performed and a yellow-brown tumour of about 5 cm in diameter with a superficial defect was found. Pathological examination showed glassy cystic tumours in several places and some fibrosis. Microscopy showed epithelial structures of varying size in which the epithelium was flat cuboid to low cylindrical. The nuclei were sometimes slightly atypical, the surrounding stroma being cellular and consisting of sinuous bundles of long elongated cells. The finding of several nodules containing cystic spaces lined by cuboidal epithelium and a band-like layer of cells (cambium layer) composed of mesenchymal-looking cells with dark nuclei and scanty cytoplasm is in keeping with a diagnosis of mesenchymal cystic hamartoma (figure).

Two years after thoracotomy the patient is well without evidence of recurrence or metastasis.

Discussion
This case report illustrates that mesenchymal cystic hamartoma of the lung enters into the differential diagnosis for a patient with pneumothorax or haemoptysis and nodules or cysts on a chest radiograph. By strict definition the

Cyst lined by cuboidal epithelium and stroma composed of mesenchymal-looking cells with dark nuclei and scanty cytoplasm. Stain: haematoxylin and eosin, original magnification × 150 reduced to 65% in origination.
Aortic valvulitis complicating Wegener’s granulomatosis

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Abstract

In a case of Wegener’s granulomatosis aortic valve replacement was performed for worsening congestive cardiac failure secondary to aortic incompetence. Two paravalvular lesions and an isolated intra-leaflet defect of the non-coronary cusp were identified at operation. Historical changes were consistent with a connective tissue disease.

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Case report

A 20 year old man presented with arthralgia, haemoptysis, inflamed fauces, conjunctivitis and weight loss. On clinical examination he was distressed, pale, febrile (39.3°C) with a tachycardia (blood pressure 110/80 mm Hg). A vasculitic rash was present on his shins and elbows. Auscultation revealed bibasilar fine crackles and a blowing left parasternal diastolic murmur. He was anaemic (haemoglobin 8.7 g/dl) with a leucocytosis (13.2 × 10⁹/l), urea 12.5 mmol/l and creatinine 199 µmol/l. Radiographic examination demonstrated cardiomegaly and fluffy basilar pulmonary opacifications. Echocardiography showed mild aortic and mitral regurgitation, mild left ventricular dilatation, and normal valve cusps. No evidence of pulmonary hypertension or oedema was demonstrated. Renal ultrasound revealed normal sized kidneys of increased echogenicity consistent with glomerulonephritis. Antineutrophil cytoplasmic antibody (ANCA) titres (>1/320) confirmed the diagnosis. Treatment with prednisolone, cyclophosphamide and plasma exchange commenced immediately.

Five months later worsening congestive cardiac failure, recurrent epistaxes, pleurisy and anaemia (haemoglobin 5.9 g/dl) necessitated readmission. Echocardiography revealed left ventricular dilatation and partially prolapsing, thin aortic valve cusps. Despite treatment his condition deteriorated.

Examination demonstrated a collapsing pulse, blood pressure of 240/120 mm Hg, a displaced apex beat, a precordial thrill, and a pandiatrophic murmur. Bibasilar crepitations persisted. A diagnosis of severe aortic incompetence with significant left ventricular failure was made.

Left axis deviation and left ventricular hypertrophy (without strain) was noted on the electrocardiogram. Repeat echocardiography confirmed a moderately dilated left ventricle with good contractility, very severe aortic regurgitation with prolapsing aortic cusps, a normal mitral valve, and an ejection fraction of...