Simultaneous bilateral spontaneous pneumothorax in a patient with recurrent, extraosseous multiple myeloma

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A 72 year old white woman was referred to our emergency department because of cough and left sided pleuritic chest pain. The patient had not suffered a trauma. On the day of admission she was started on levofloxacine therapy by the referring physician.

The patient was known to have arterial hypertension. Eleven months before admission she had presented with intermittent atrial fibrillation and left ventricular hypertrophy with normal left ventricular ejection fraction on echocardiography. Ten months before admission she had been diagnosed with pulmonary embolism and a multiple myeloma of the IgG/lambda type, stage IIIA according to the Durie-Salmon staging system, presenting with anaemia, hypercalcaemia, and a serum IgG level of 57.1 g/l. She was treated with phenprocoumon, monthly infusions of pamidronate, and a total of five cycles of prednisone and melphalan until four weeks before admission. Under this treatment she was normocalcaemic, and IgG levels had dropped to 14.1 g/l. On physical examination the patient was in poor overall condition. She was orthopnoeic with a respiratory rate of 30 breaths/min; blood pressure was 213/109 mm Hg, and the pulse rate 110 beats/min. Examination of the heart was normal. The left lower lung was dull to percussion, and cracks were heard over both lungs. There was no abdominal tenderness or organomegaly. A chest radiograph showed bilateral pneumothorax that was more prominent on the left side, cardiomegaly, pulmonary venous congestion, and bilateral pleural effusions (fig 1). Laboratory data were remarkable for microcytic anaemia (haemoglobin 104 g/l), a low normal thrombocyte count (96 × 10⁹ µl⁻¹), raised creatinine (142 µmol/l) and C reactive protein (121.8 mg/l) levels, and prolonged prothrombin time (international normalised ratio was not measurable). Urinalysis revealed trace protein and haemoglobin. Arterial blood gas analysis showed hypoxemia (arterial oxygen tension 8.21 kPa), discrete hypercapnia (arterial carbon dioxide tension 5.75 kPa), and metabolic acidosis (pH 7.32, bicarbonate 21.8 mmol/l).

The patient was taken to the intensive care unit and treated with oxygen, furosemide (frusemide), amoxycillin/clavulanic acid, fresh frozen plasma, and vitamin K. A chest tube was inserted into the left pleural space. A few hours after admission, she was resuscitated because of pulseless electrical activity, and mechanical ventilation was initiated. Because of progression of the right sided pneumothorax, an additional chest tube was placed on the second day. Analysis of the right pleural fluid revealed an exudate containing leukocytes at 14 × 10⁹/l with >50% granulocytes and 25%–50% plasma cells (fig 2). A Gram stain showed no micro-organisms, and bacterial cultures were sterile. Repeated blood cultures were sterile, and a test for urinary antigen of Legionella pneumophila resulted negative.

Bronchoscopy was performed on the second day. The right main and left lower lobe bronchi were occluded with blood clots. Bronchoalveolar lavage revealed unspecific lymphocytic and granulocytic inflammation; no acid-fast bacilli were detected microscopically and in cultures, and silver staining for Pneumocystis carinii was negative.

A presumptive diagnosis of extramedullary multiple myeloma involving the pleura was made. The patient became hypotensive and oliguric and was treated with saline and vasopressors. A morning cortisol level was 1092 nmol/l; no steroids were supplemented. The antibiotic regimen was changed to piperacillin/tazobactam and clarithromycin. Repeated radiographic controls confirmed expansion of both lungs. Echocardiography showed a hypertrophic left ventricle with a normal ejection fraction. On the morning of the third day, acute severe hypotension occurred. The patient died 36 hours after admission.

The necropsy revealed extensive medullary and extraosseous manifestation of multiple myeloma, involving multiple lymph nodes, all lobes of both lungs, the visceral pleurae, chest wall, liver, pancreas, the epicardial fat and right atrium with compression of the right coronary artery, both kidneys and adrenal glands, and the femurs as well as the spine. Immunohistochemistry confirmed monoclonal IgG/lambda positive plasma cells staining positively for IgG and lambda chains.

SBSP is a rare condition and may be caused by trauma, parenchymal lung disease, infections, or neoplasms. This is the first report of SBSP caused by pleuropulmonary infiltration of multiple myeloma.
The myocardium was notable for severe amyloidosis and peracute necrosis of the posterior left wall.

DISCUSSION

SBSP comprises a small fraction of all cases of spontaneous pneumothorax. An earlier report on a series of 12 patients with SBSP over 19 years estimated this fraction to be around 1%–4%.

Non-traumatic causes of SBSP include idiopathic, infections (for example, *Mycobacterium tuberculosis* or *Mycoplasma pneumoniae*), parenchymal lung disease (for example, histiocytosis X,

The diagnosis of extramedullary manifestation of her multiple myeloma, established cytologically in the pleural effusion as lambda light chains in tissue obtained from several visceral organs (not shown).

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