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

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A patient with simultaneous bilateral spontaneous pneumothorax (SBSP) due to pulmonary and pleural manifestations of recurrent multiple myeloma is presented. The patient died in shock of unknown cause. The diagnosis was suspected from pleural fluid examination showing an exudate with numerous plasmocytes. Macroscopically and histologically, the visceral organs and the bone marrow were infiltrated with multiple monoclonal proliferations of 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.

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 levofloxacin 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 phenprocoumone, 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 resulted negative. A chest radiograph showed cardiomegaly, bilateral pleural effusions, and simultaneous bilateral pneumothorax [arrows indicate the visceral pleural line [left] and a small apical area of gas in the pleural space [right]].

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 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.
Simultaneous bilateral spontaneous pneumothorax

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, sarcoidosis, lymphangioleiomyomatosis, Marfan’s syndrome); and neoplasms (for example, lymphoma, sarcoma). Our patient presented with typical signs and symptoms of pneumothorax. The clinical course and laboratory results suggested an infection as the underlying disease; this could not be ruled out completely since she had received antibiotic therapy before admission. Given her history of recent steroid therapy, we were concerned about tuberculosis or P carinii pneumonia, which both may present with SBSP. The diagnosis of extramedullary manifestation of her multiple myeloma, established cytologically in the pleural effusion as well as histologically at necropsy, was clinically unexpected because the patient had shown a favourable response to standard therapy. However, rapid and fatal disease progression with extraosseous manifestation even after complete remission has been observed by others and may result from clonal evolution of malignant, dedifferentiated plasma cells. In a recent series of 52 consecutively necropsied patients with multiple myeloma, over 60% showed extraosseous involvement, including the lung in 15.4%. Pneumothorax was not found in that series, and to our knowledge, the patient reported here is the first with SBSP due to pleuropulmonary infiltration of multiple myeloma.

Summary points

- Simultaneous bilateral spontaneous pneumothorax is rare, accounting for approximately 1%–4% of all cases of spontaneous pneumothorax.
- Aetiologies include trauma, infections, parenchymal lung disease, malignancy, and idiopathic.
- In the largest published series (12 patients), the majority of patients had underlying lung disease.
- Typically, bronchogenic carcinoma does not cause simultaneous bilateral spontaneous pneumothorax.
- Rather, the malignancies leading to this complication are of mesenchymal and haematological origin.
- Treatment includes pleural drainage on one or both sides. Early definitive treatment by surgical pleurectomy on at least one side has been recommended, particularly in patients with underlying lung disease.

REFERENCES

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