Acute respiratory distress syndrome due to pulmonary involvement by neoplastic plasma cells in multiple myeloma

D B Marmor, J L Farber, J E Gottlieb

Pulmonary involvement with multiple myeloma occurs infrequently and may be difficult to distinguish from more common primary lung tumours, metastatic disease, or other pleural and parenchymal abnormalities. A patient who developed acute respiratory distress syndrome (ARDS) was subsequently found to have multiple myeloma with involvement of lung parenchyma by neoplastic plasma cells. Only one other report of ARDS in association with multiple myeloma was found, and there are no previous reports where the appearance of ARDS antedated a diagnosis of multiple myeloma. In patients with ARDS, parenchymal involvement from multiple myeloma should be included in the differential diagnosis.

A 65 year old woman was admitted to hospital complaining of dull pleuritic retrosternal chest pain of 5 days’ duration. It had been somewhat relieved with non-steroidal anti-inflammatory drugs. She also complained of a cough and worsening dyspnoea upon exertion. A 2 month history of nausea was accompanied by a weight loss of 11 lb. Past medical history included non-insulin dependent diabetes, hypertension, asthma, and obesity.

On physical examination the patient initially appeared comfortable at rest but was dyspnoeic with limited activity. Her temperature was 101.7°F, heart rate 96 beats/min, respiratory rate 20 breaths/min, blood pressure 117/72 mm Hg. The only significant physical finding was bilateral basilar crackles. Laboratory data revealed a white blood cell count of 10 400/mm³ and haemoglobin of 9.7 g/dl. The serum sodium was 131 mEq/l, total calcium 8.5 mg/dl, and creatinine 1.0 mg/dl. Urinalysis revealed trace protein. The chest radiograph showed bilateral multifocal areas of nodular consolidation (fig 1), and the ECG showed normal sinus rhythm with diffuse non-specific T wave abnormalities. Serum troponin levels were normal. Oxygen saturation was 88% on room air and 94% on 5 l oxygen via nasal cannulae.

The patient’s respiratory status steadily declined over the ensuing days, despite treatment with antibiotics for presumed pneumonia. Four days after admission she was in acute respiratory distress with tachypnoea and the use of accessory muscles. Arterial blood gas showed pH 7.46, PaCO₂ 5.2 kPa, and PaO₂ 4.5 kPa on 50% FiO₂ non-rebreather mask. She required intubation and mechanical ventilation with a low tidal volume high positive end expiratory pressure protocol. Bronchoalveolar lavage yielded fluid that contained 40% macrophages, 25% neutrophils, and 35% lymphocytes. No malignant cells and no pathogens were identified. All blood, sputum, and urine cultures were negative. A chest CT scan showed diffuse, patchy, multifocal air space consolidation bilaterally with a nodular character, small bilateral pleural effusions, mediastinal lymphadenopathy, and a questionable lytic lesion of the T9 vertebra.

Ultrasound guided thoracentesis yielded 200 ml of bloody exudative effusion. Cytology was negative for malignant cells. Thoracoscopic biopsy revealed diffuse and nodular infiltration of the lung parenchyma by neoplastic plasma cells (fig 2). The 24 hour urine contained 918 mg protein, and immunoelectrophoresis revealed a monoclonal band in the gamma region identified as IgG kappa with excess kappa light chains. Serum immunoelectrophoresis similarly showed hypergammaglobulinaemia with a monoclonal IgG kappa band.

Treatment continued with piperacillin/tazobactam, azithromycin, and vancomycin for pneumonia, and methylprednisolone for myeloma. She was judged to be a poor candidate for more aggressive chemotherapy owing to progressive multisystem organ failure. In keeping with the patient’s advance directive and after consultation with her family, supportive care was withdrawn. Permission for bone marrow biopsy or necropsy was withheld.

DISCUSSION

This patient was diagnosed with multiple myeloma on the basis of a monoclonal serum protein spike, the presence of urinary Bence Jones proteins, and infiltration of the lungs by neoplastic plasma cells. She developed acute respiratory distress syndrome (ARDS) secondary to this nodular involvement of the lungs by the myeloma. Multiple myeloma presenting as ARDS has not been reported previously. Absence of leucocytosis on admission, failure to respond radiographically or clinically to antibiotics, the absence of positive cultures for microorganisms, and pathological findings without evidence of infectious pneumonia all argue against infection as a major factor contributing to the development of ARDS.

Multiple myeloma may involve the thorax in a variety of ways, but pulmonary parenchymal involvement is uncommon. Thoracic manifestations include skeletal abnormalities,
Multiple myeloma may produce varied patterns on chest radiographs including multiple masses mimicking solid tumour metastasis, diffuse interstitial disease from alveolar septal amyloidosis, or consolidation.11 12 Our patient presented with a chest radiographic pattern that was consistent with the diagnosis of ARDS, as defined by the American-European Consensus Conference on ARDS.13 However, both the ARDS Clinical Trials Network (in terms of inclusion criteria) and a recent review of “imitators” of ARDS highlight the pathophysiological heterogeneity that may underlie such a broad clinical definition.14 15 Since our patient had neither prior history nor clinical evidence of multiple myeloma, this pathological diagnosis was not made until an open lung biopsy was obtained. A greater clinical suspicion could conceivably have resulted in bone marrow biopsy, thereby obviating the need for lung biopsy. We suggest that multiple myeloma with direct plasma cell invasion of pulmonary parenchyma be added to the list of pathological mechanisms that can produce the clinical syndrome of ARDS.

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