CASE REPORT

Pulmonary sarcoidosis and the acute respiratory distress syndrome (ARDS)

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A 50 year old man presented with 3 weeks of exertional dyspnoea. His chest radiograph on admission revealed diffuse bilateral interstitial infiltrates. He did not respond to antibiotics but subsequently improved on high dose corticosteroids. Bronchoscopic examination with transbronchial biopsy specimens revealed the presence of non-necrotising granulomas. This case demonstrates an unusual clinical presentation of life threatening pulmonary sarcoidosis characterised by the development of acute respiratory distress syndrome (ARDS) with acute respiratory failure.

Sarcoidosis is a multisystemic granulomatous disorder of unknown aetiology that most commonly affects the lungs and lymphatic system. The case history is described of a man with a distinctly unusual clinical presentation of life threatening pulmonary sarcoidosis characterised by the development of acute respiratory distress syndrome (ARDS) with acute respiratory failure.

CASE HISTORY

A 50 year old man presented with a 3 week history of progressive exertional dyspnoea, poor appetite, and a cough productive of whitish sputum. He denied fever, night sweats, or haemoptysis. He had a history of 20 pack years of cigarette smoking and type 2 diabetes mellitus. He was afebrile with a blood pressure of 120/60, pulse of 100 beats/min, and a respiratory rate of 16 breaths/min. There was no jugular venous distention, chest examination revealed bibasilar inspiratory crepitations, and cardiac auscultation was normal. The chest radiograph at presentation revealed extensive bilateral "interstitial" infiltrates (fig 1). Arterial blood gas analysis performed while the patient was breathing room air gave pH 7.42, $Paco_2$ 4.8 kPa, and Pao_2 7.1 kPa. Complete blood count, electrolytes, and renal and liver function tests were within normal limits except for a lactate dehydrogenase level of 376 U/l (normal 80–200 U/l). Electrocardiography showed sinus tachycardia.

Empirical treatment with intravenous erythromycin and cefotaxime was started and supplemental oxygen was given by nasal cannula at 2 l/min. His clinical state deteriorated and trimethoprim/sulfamethoxazole (TMP/SMX) and corticosteroids were added to cover for possible *Pneumocystis carinii* pneumonia (PCP). Forty eight hours after admission his respiratory rate was 28 breaths/min with a Pao_2 of 8 kPa on a 100% non-rebreather mask. A repeat chest radiograph showed diffuse bilateral air spaces (fig 2).

Fibreoptic bronchoscopy with bronchoalveolar lavage (BAL) was performed which revealed no evidence of PCP, acid fast bacilli, fungi, or malignancy. TMP/SMX was stopped but corticosteroids were continued because of a fairly marked clinical improvement following their initiation.

By day 6 after admission the patient's respiratory status was notably improved with a respiratory rate of 20 breaths/min and a repeat chest radiograph showed an improvement in the diffuse bilateral air space disease (fig 3). Arterial blood gas analysis the next day gave a Pao_2 of 11.6 kPa on supplemental oxygen given by nasal cannula at 4 l/min. Bronchoscopic examination with biopsy specimens was performed on day 8 and revealed the presence of non-necrotising granulomas. Special stains and subsequent cultures were negative for fungal, mycobacterial, viral, and other organisms.

The patient continued to improve and was discharged home 2 weeks after admission on oral prednisone 60 mg/day with a Pao_2 of 8.3 kPa while breathing ambient room air. His chest radiograph showed continuous improvement in the bilateral air space disease. The serum angiotensin converting enzyme level was 83 IU/l.

DISCUSSION

The clinical presentation and natural course of sarcoidosis can vary greatly. Approximately one half of patients are asymptomatic with characteristic abnormalities noted on routine chest

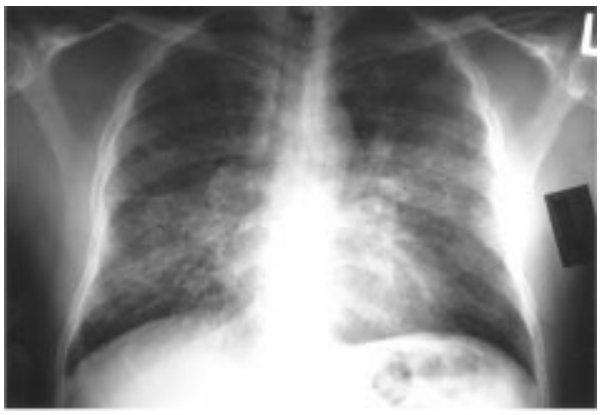

Figure 1 Chest radiograph on admission.

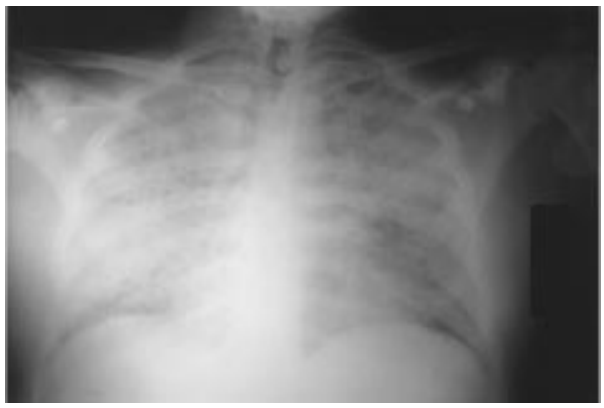

Figure 2 Chest radiograph on day 2.

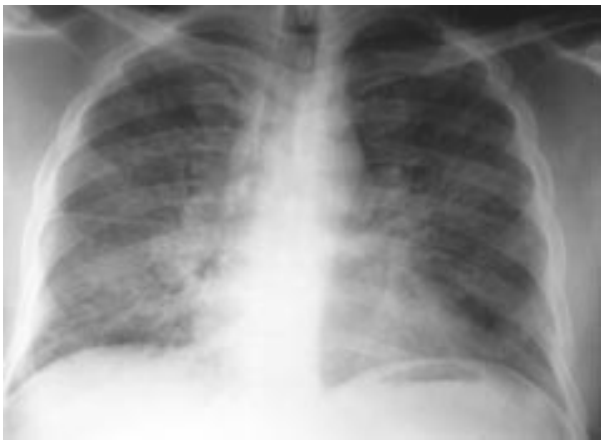

Figure 3 Chest radiograph on day 6.

radiographs. In symptomatic patients an insidious onset of symptoms from pulmonary, ocular, or cutaneous involvement is usual.

Although sarcoidosis with lung involvement is quite common, the development of ARDS with acute life threatening respiratory failure has not, to our knowledge, been previously reported in the English literature.[1 2] A single case report exists in the Japanese literature.[3]

Our patient clearly met the diagnostic criteria established by the American-European consensus conference on ARDS[4] and the diagnostic tests performed did not reveal any condition known to cause ARDS. Fever and leucocytosis were both absent and he had no evidence of sepsis or systemic inflammatory response syndrome. Bronchoscopic sampling with BAL on two occasions, sputum, and blood cultures as well as serological studies were all negative.

The early reaction in sarcoidosis is characterised by the accumulation of activated T cells and macrophages at the sites of ongoing inflammation. These cells spontaneously release interferon γ, interleukin 2 (IL-2), and other cytokines. ARDS may have developed in our patient as a result of the release of these cytokines. Previous publications have reported the presence of chest radiographic abnormalities in recipients of recombinant IL-2, including a case of ARDS.[5 6]

It is well recognised that sarcoidosis is a diagnosis of exclusion. While our patient had an occupational history of welding, hence raising concern about the possibility of hypersensitivity pneumonitis, he had been unemployed in that field since he immigrated from Cuba to the United Sates 8 years before presentation and previous chest radiographs prior to the acute presentation had been unremarkable. Tuberculosis was considered in the initial differential diagnosis, and his purified protein derivative (PPD) skin test was positive. However, extensive investigation did not reveal any evidence of tuberculosis and numerous cultures were negative. Most importantly, despite approximately 12 months of continued treatment with corticosteroids for sarcoidosis, there has been no evidence of tuberculosis or other infectious processes.

The importance of this report is primarily related to the possibility that sarcoidosis may present as ARDS which, in the absence of an appropriate diagnosis and treatment, could potentially be fatal. As far as we are aware, this is the first case reported in the English literature of acute life threatening pulmonary sarcoidosis presenting with ARDS.

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