

Sarcoidosis in an adult with cystic fibrosis

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ABSTRACT Sarcoidosis in an adult patient with cystic fibrosis lung disease was diagnosed on the basis of pulmonary function and radiographic data. It should be considered in the differential diagnosis of new diffuse interstitial infiltrates or hilar adenopathy in a patient with cystic fibrosis; biopsy of lung, lymph node, or skin lesions and interleukin-2 receptor levels may help to obtain a diagnosis.

The diagnosis of coexistent pulmonary disease in the presence of chronic lung disease due to cystic fibrosis can be difficult. The clinician must remain vigilant for other causes of deteriorating lung function, however, as delay in diagnosis may result in inadequate treatment.

Case report

A 29 year old white man with cystic fibrosis was admitted to hospital because of deteriorating pulmonary function values and the appearance of a new interstitial infiltrate on the chest radiograph. Cystic fibrosis had been diagnosed when he was 13, with a sweat chloride concentration on pilocarpine iontophoresis¹ of 860 mmol/l. At the ages of 13 and 22 years the patient was admitted to hospital because of dehydration but was otherwise generally healthy, having pancreatic enzyme replacement therapy.

At the age of 28, seven months before admission to hospital, he had noted the onset of occasional episodes of chest tightness at rest, which were relieved with salbutamol from a metered dose inhaler. Physical examination showed nothing remarkable. A chest radiograph showed bilateral hilar enlargement, diffuse interstitial infiltrates, bronchiectasis localised to the right upper lobe, and hyperinflation. Pulmonary function tests (table: 21 January 1988) indicated a decline in the forced expiratory volume in one second (FEV₁), vital capacity, and flow at 25–75% of vital capacity from previous measurements. A full blood count and differential count were within normal limits. Serological studies for *Histoplasma capsulatum* and *Mycoplasma pneumoniae* gave negative results. Serum IgE levels and human immunodeficiency virus titres were normal. The tuberculin skin test

response was negative. Sputum cultures grew *Pseudomonas aeruginosa*. Intravenous tobramycin and ticarcillin, chest physiotherapy, and bronchodilators were given for 14 days with little clinical improvement.

One month after discharge the chest radiograph and pulmonary function tests (table: 17 February 1988) showed no improvement. Fibreoptic bronchoscopy disclosed no endobronchial lesions, and transbronchial lung biopsy was performed. This showed multiple non-necrotising granulomas. Special stains for acid fast and fungal organisms gave negative results, and no birefringent foreign particles were present. Cultures of bronchial washings were negative for mycobacteria and fungi. The serum calcium concentration was 2.4 (normal 2.25–2.75) mmol/l, and serum angiotensin converting enzyme activity was 54 (normal 20–60) nmol/min/ml. A gallium-67 scan showed moderately increased uptake in both lungs with focal areas of increased activity in the left lung base and right perihilar region. The serum interleukin-2 receptor level was raised at 640 U/ml (normal <477 U/ml²). On the basis of the radiographic pattern and histological findings the diagnosis of sarcoidosis was made (radiological stage II).

Discussion

We describe a patient with lung disease associated with cystic fibrosis and coincident sarcoidosis. A review of published reports brought to light only one report (of two cases) of sarcoidosis in the setting of cystic fibrosis.³ Though pulmonary function and radiographic data suggested the diagnosis of sarcoidosis in our patient, the diagnosis may be

Results of pulmonary function studies (% predicted normal in parentheses) in a patient with sarcoidosis and cystic fibrosis

	30 June 1987	21 Jan 1988	17 Feb 1988	19 April 1988
FEV ₁ (l)	4.92 (112)	3.72 (85)	4.22 (96)	3.92 (90)
FVC (l)	6.33 (120)	5.69 (108)	5.83 (111)	5.64 (107)
FEV ₁ /FVC (%)	77 (93)	65 (78)	72 (87)	69 (83)
FEV ₂₅₋₇₅ (l)	4.26 (91)	2.07 (45)	2.86 (62)	2.39 (51)
TLC (l)	7.28 (114)	7.26 (113)	—	—
TLC (mmol min ⁻¹ kPa ⁻¹ l ⁻¹)	—	9.74 (73)	—	—

FEV₁—forced expiratory volume in one second; FVC—forced vital capacity; FEV₂₅₋₇₅—volume expired during time interval between 0.25 × FVC and 0.75 × FVC; TLC—total lung capacity; TLCO—single breath transfer factor for carbon monoxide, corrected for haemoglobin concentration.

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elusive in patients with more advanced lung disease. The diagnosis may be made on the basis of pulmonary function values, chest radiographs, serum interleukin-2 receptor levels, and biopsy specimens of lung, lymph node, or skin lesions. Sarcoidosis should be considered in the differential diagnosis of new diffuse pulmonary interstitial infiltrates or hilar adenopathy in patients with cystic fibrosis.

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References

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