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Relapsing organising pneumonitis in a man with primary biliary cirrhosis, CREST syndrome, and chronic pancreatitis

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We report a relapsing, steroid-responsive, organising pneumonitis, in a man with the CREST syndrome (calcinosis, Raynaud's phenomenon, oesophageal dysfunction, sclerodactyly, and telangiectasia) variant of systemic sclerosis and primary biliary cirrhosis. This pattern of pulmonary disease has not previously been described in systemic sclerosis. The association between primary biliary cirrhosis and the CREST syndrome is well recognised in women[1] and we report the association in a man.

Case report

A 58-year-old man, a non-smoking clerical worker, presented in May 1978 with an eight-month history of lethargy, weight loss, and a cough productive of mucoid sputum, and increasing shortness of breath for two months. Raynaud's phenomenon had been present for 36 years. On examination he was ill but apyrexial, dyspnoeic on the slightest exertion, with telangiectasia on his face, crackles in the chest, and splenomegaly. His chest radiograph is shown in figure 1. Sputum and blood cultures were sterile. Serological tests for common respiratory viruses, *Mycoplasma pneumoniae, Coxiella burnetti,* the psittacosis agent, and *Legionella pneumophila* gave negative results. His haemoglobin concentration was 15·2 g/dl, white blood count 13·7 × 10^9/l (normal differential), ESR 49 mm in one hour, serum IgM 1192 IU/ml (normal 58–197), and IgG 200 IU/l (normal 57–172). The serum alkaline phosphatase activity was 1549 IU/l (normal 50–200), aspartate transferase 33 IU/l (normal 5–17), and γ-glutamyl transferase 269 IU/l (normal 5–28). The test for antimitochondrial antibody was strongly positive (>1/40) and that for anti-DNA antibody negative. A trephine lung biopsy showed organising pneumonia (fig 2). A liver biopsy specimen was compatible with a diagnosis of primary biliary cirrhosis. Eleven days after starting 50 mg/day of prednisolone he could walk up three flights of stairs and there had been considerable resolution of the radiological shadowing.

Over the next year the dose of prednisolone was gradually reduced to 15 mg on alternate days and treatment continued at this dose until August 1980. Sclerodactyly was noted at this time and he complained of acid regurgitation. A barium swallow showed a dilated oesophagus and diminished peristalsis (a previous one had shown nothing abnormal). The chest radiograph had not changed for many months (that is, during maintenance steroid treatment) and showed only residual shadowing in the lingula. Radiographs of the hands showed calcinosis. Liver tests continued to show evidence of cholestasis, and endoscopic retrograde cholangiopancreatography performed at this time showed a normal biliary system but an abnormal pancreas, with stricturing and irregularity of the main pancreatic duct in the tail of the pancreas.

The dose of prednisolone was gradually reduced over the next six months, and when he had been having 5 mg on alternate days for three months he developed increasing shortness of breath and lethargy. There was fresh confluent shadowing in the right upper zone. The findings at bronchoscopy were normal. This shadowing rapidly resolved with an increase in the dose of prednisolone.

Discussion

Pulmonary disease in systemic sclerosis is usually manifest radiologically as lower-zone shadowing, but confluent

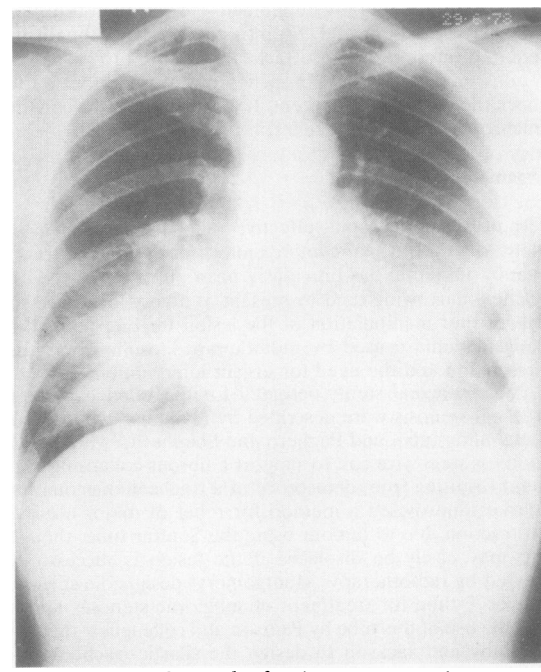

Fig 1 *Chest radiograph of patient at presentation.*

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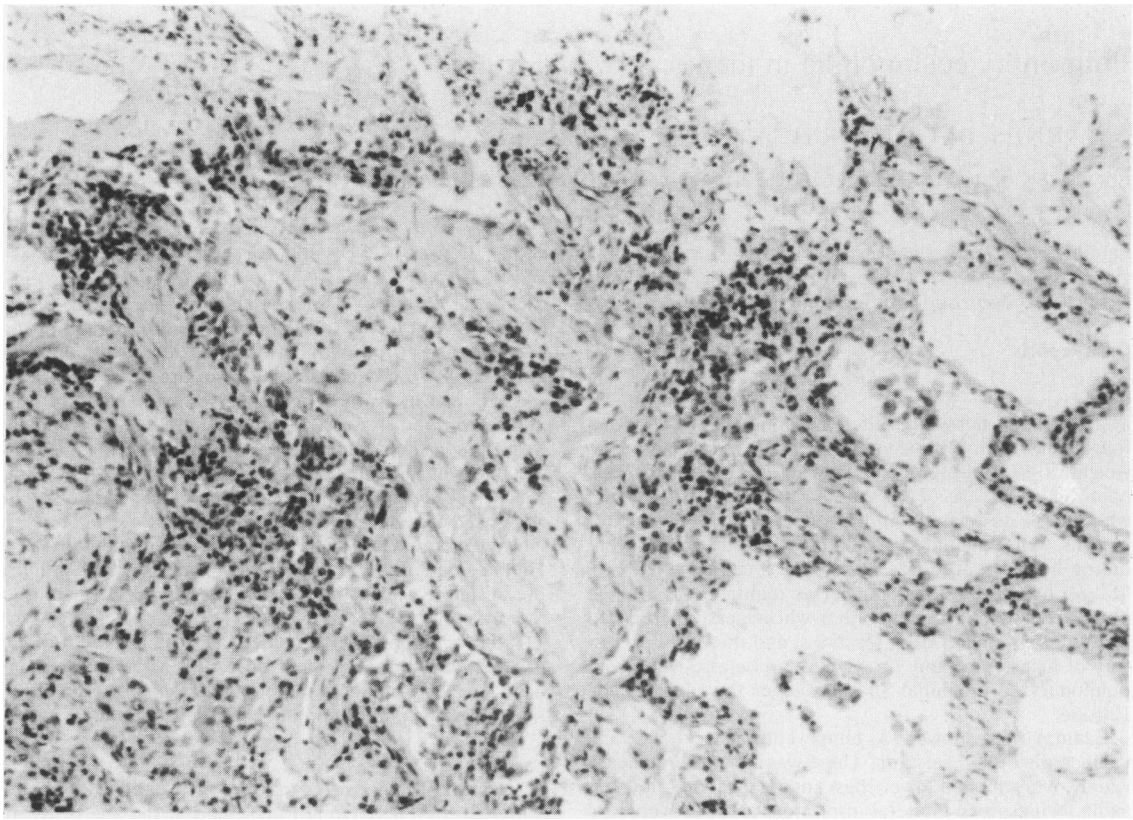

Fig 2 *Lung biopsy specimen showing a focus of organising pneumonia with small buds of connective tissue ("bourgeons conjunctifs") in the air spaces and proliferation of fibrous tissue through the pores of Kohn. (Haematoxylin and eosin, × 200.)*

shadowing has occasionally been described.[2] The pathology of the latter has not been defined but in our case an organising pneumonia was found. The aetiology of this is not clear; an organising pneumonia may result from incomplete resolution of infection but no infective agent was identified (and the mode of onset and relapse after three years would not suggest this). Aspiration of gastric acid or contents has been suggested as the cause of the confluent shadowing in systemic sclerosis[2] but our patient had a barium meal examination showing nothing abnormal at presentation; and the other histological features of aspiration, foreign material, foreign-body multinucleate cells, and extrinsic lipid were absent.[3] The response to prednisolone, and also the subsequent relapse, was both rapid and dramatic, and similar to that described by Grimblat *et al* in two cases of relapsing organising pneumonias; neither of these, however, had evidence of connective tissue disorders.[4]

In the course of investigating the lung disease, the results of cholestatic liver function tests were noted; a concurrent diagnosis of primary biliary cirrhosis was made and radiographic evidence of chronic pancreatitis was found. The association between systemic sclerosis and primary biliary cirrhosis, although well recognised in women, has been described in only three men,[5] none of whom had the CREST variant. This case demonstrates the association in a man.

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