Thorax, 1979, **34**, 677–679

Short reports

Relapse of fibrosing alveolitis (desquamative interstitial pneumonia) after twelve years

A M HUNTER AND D LAMB

From the Chest Unit, City Hospital, and University Department of Pathology, Edinburgh, UK

A patient with desquamative interstitial pneumonia completely recovered after treatment with oral corticosteroids. Twelve years after stopping treatment the patient relapsed but has again recovered after treatment.

Case report

A 34-year-old woman presented with a 15 month history of dyspnoea progressing to breathlessness on minimal exertion. She had slight cough and scanty mucoid sputum. There was no history of arthralgia. She had taken no drugs and had been exposed to no recognised allergens.

Gross clubbing of fingers and toes was present. Chest expansion was reduced and numerous inspiratory crackles were heard over the lower two-thirds of both lungs. The chest radiograph showed nodular opacities in the lower half of both lung fields, being more pronounced on the right side. Lung function tests showed a restrictive pattern (table). Arterial oxygen saturation was reduced to 89%. Blood tests including a full connective tissue disorder screen gave normal results.

Serial pulmonary function tests

First presentation	*Predicted (litres)*	*23/2/62*	*17/4/62*	*12/5/62*	*1969*
FEV (0·75 s) l	2·25	1·15	1·51	2·12	2·13
FVC l	3·0	1·75	2·55	3·05	3·63
TLC l	4·5	2·6	3·37	3·96	4·15
VC l	3·0	1·5	1·85	2·45	2·8
Second presentation	*Predicted (litres)*	*8/2/77*	*14/4/77*	*August 1978*	
FEV (1 s) l	2·55	1·8	2·0	2·02	
FVC l	2·78	2·8	3·0	3·0	
TLC l	4·32	4·25	4·55	4·32	
VC l	2·6	2·62	2·76	2·7	
Transfer factor (single breath) mmol min^{-1} kPa^{-1}	7·8	3·1	5·39	—	

Open lung biopsy was carried out. The most obvious feature was the presence of large numbers of large eosinophilic cells filling alveoli and also extending into respiratory and terminal bronchioles. These cells (fig 1) were large, with abundant eosinophilic, somewhat granular, cytoplasm, and vesicular nuclei. There

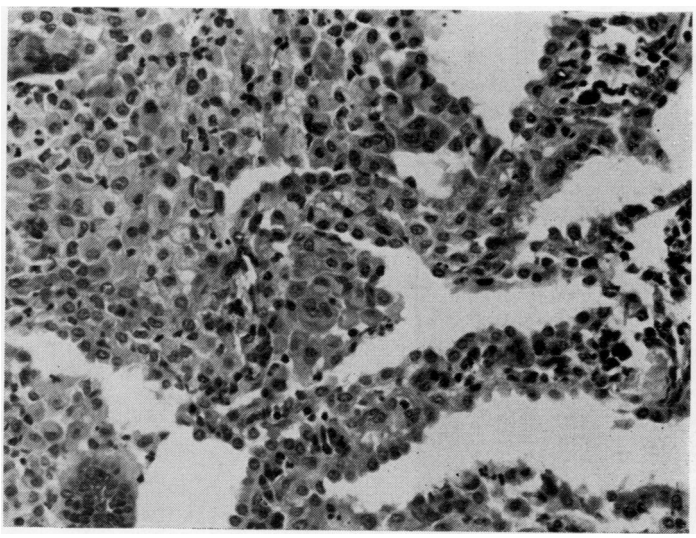

Fig 1 *Alveolar lumina are filled with large mononuclear cells with granular cytoplasm among which occasional multinucleate forms are seen (top left). Alveolar lining cells are cuboidal; alveolar wall is thickened with a moderate lymphocytic and plasma cell infiltrate (bottom right). Haematoxylin and eosin ×360 (original magnification).*

was some variation in size and shape of cells, binucleate and occasionally multinucleate forms being seen. Occasional mitotic figures were seen among the masses of cells filling the alveolar lumina. Alveolar lining cells were prominent and in places cuboidal in appearance. The alveolar walls themselves were rather thickened and showed a moderate infiltration with plasma cells and lymphocytes; occasional polymorphonuclear leucocytes were seen. The degree of alveolar wall thickening was well seen with a reticulin stain (fig 2). No obliteration of alveolar lumina was seen, and there was no gross scarring. No granulomas were seen; special stains for organisms and fungi were all negative and no doubly refractile material was present in the tissue examined. The histological features are those of desquamative interstitial pneumonia. Small eosinophilic intranuclear inclusions were present in occasional alveolar lining cells. These were noted by Liebow *et al* (1965) in a small proportion of their cases but no specific viral aetiology has been proposed.

Treatment was started with oral prednisolone, 60 mg daily, and dramatic clinical improvement occurred. The dose was gradually reduced over a period of two years and then stopped. She became asymptomatic, the clubbing of the fingers and the crackles disappeared, and the radiographic appearances and the respiratory function tests returned to normal (table).

Fourteen years after initial presentation and 12 years after stopping treatment with prednisolone she developed increasing breathlessness after a non-specific febrile illness. Once again she had taken no drugs and there was no recognisable allergen. On this occasion, however, she had an arthralgia affecting elbows, shoulders, and, in particular, the small joints of the hands. The joints were normal in appearance. Clubbing of the fingers and crackles at both lung bases were again present. The chest film again showed nodular opacities at both bases. Respiratory function tests showed a reduced transfer factor and a slight reduction of vital capacity and total lung capacity (table). Rose Waaler test was positive 1 in 128. Lung biopsy was not repeated.

High dose oral prednisolone was again given with a highly satisfactory clinical, radiographic, and physiological response. She is again asymptomatic 18 months later.

Discussion

This patient developed the characteristic clinical, physiological, and radiographic features of fibrosing alveolitis. The pathological features corresponded to those characteristic of desquamative interstitial pneumonia as described by Liebow *et al* (1965). In the biopsy specimen examined there was no progression to a frank honeycomb pattern of pulmonary damage.

Whether one agrees with Liebow *et al* (1965) that desquamative interstitial pneumonia is a separate entity or with Scadding and Hinson (1967) that it represents one end of a spectrum of cryptogenic fibrosing alveolitis, such a prominent desquamative pattern appears to be associated with a better prognosis. The natural history, including the prognosis after treatment, of desquamative interstitial pneumonia has been reviewed recently by Carrington *et al* (1978). In this form of fibrosing alveolitis improvement was recorded in 61·5% of patients, some of whom needed indefinite maintenance treatment.

We know of no patient previously reported to have made a total recovery from fibrosing alveolitis and to have relapsed 12 years later. A very favourable therapeutic result has again been obtained.

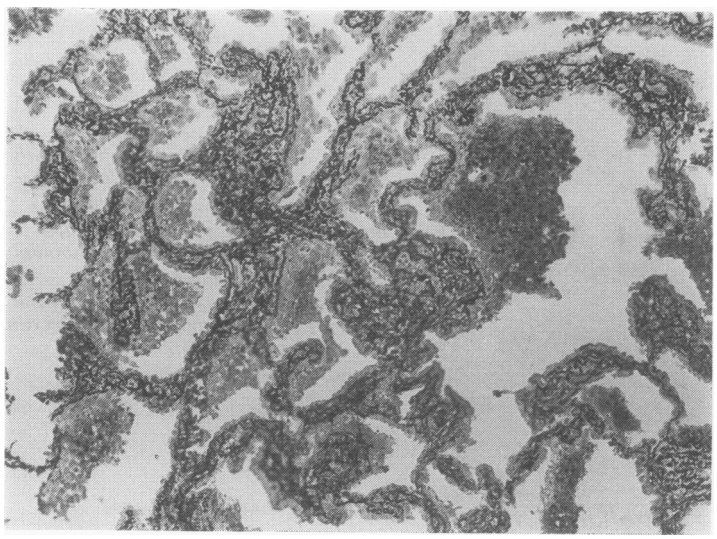

Fig 2 *Large airspace at top is part of a respiratory bronchiole. It and alveoli are filled with cells. Reticulin pattern shows widening of most alveolar walls but no gross scarring. Reticular stain ×150 (original magnification).*

References

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Requests for reprints to: Dr A M Hunter, Llandough Hospital, Penarth, S Glamorgan.