

Obliterative bronchiolitis and upper-zone pulmonary consolidation in rheumatoid arthritis

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We report a patient with two of the rare intrapulmonary complications of rheumatoid disease. Necropsy showed that obliterative bronchiolitis was the cause of the bilateral upper-zone consolidation noted on the chest radiograph.

Case report

A woman aged 55 years, who had had rheumatoid arthritis for 13 years, was admitted to hospital with a five-week history of increasing shortness of breath. She had had no previous respiratory problems. She had never received systemic corticosteroids or penicillamine and had stopped gold treatment 10 years earlier because of a rash.

Examination showed advanced rheumatoid arthritis affecting the hands, wrists, elbows, and knees, with nodules below both elbows. She was breathless at rest with prolonged expiration and an occasional inconstant, mid-inspiratory squeak was heard over the upper anterior chest.

The chest radiograph two years earlier had been normal. Her current chest radiograph showed lung fields of normal size with bilateral upper-zone homogeneous shadowing confirmed on tomography (fig 1). The FEV₁ was 0.42 l (16% of predicted) and FVC 1.46 l (41% of predicted). The total lung capacity (helium dilution) was 85% of the predicted value. The volume-flow loop confirmed severe airways obstruction. Her arterial pH was 7.45, Pao₂ 5.1 kPa, and Paco₂ 7.3 kPa. Rheumatoid serological tests showed F 11 LP 1/640; SCAT 1/28. Antinuclear and anti-DNA antibodies and aspergillus precipitins were not detected. Sputum cultures for aspergillus and tubercle bacilli gave negative results.

A diagnosis of obliterative bronchiolitis was made. Despite treatment with prednisolone at an initial daily dose of 60 mg the patient's breathlessness and airways obstruction showed no improvement and the upper-zone consolidation remained unchanged. Her condition continued to deteriorate and she died four months after her breathlessness began.

The lungs were examined in detail after fixation by bronchial infusion with 10% formal saline. Both apical segments were pale and firm on section; the lungs were otherwise macroscopically unremarkable apart from congestion and the presence of peripheral thromboemboli. The changes in the apical segments were due to organising obstructive pneumonitis. The lung architecture was intact but there was patchy interstitial fibrosis with filling of alveolar

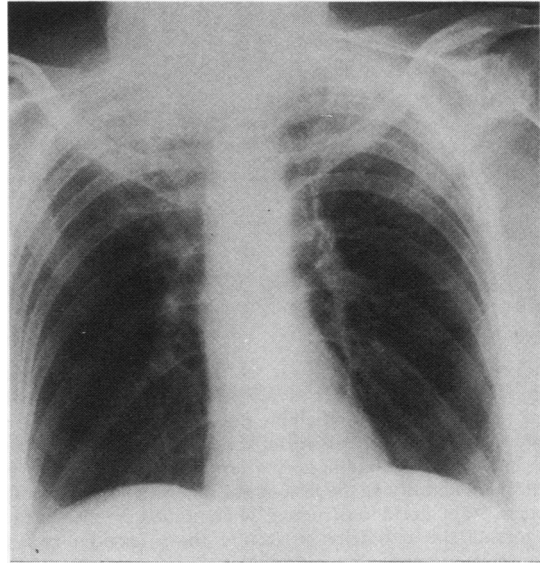


Fig 1 Chest radiograph on admission showing bilateral upper-zone homogeneous shadowing.

spaces by foamy macrophages, plasma cells, polymorphs, cholesterol crystals, and fibrous plugs.

Throughout both lungs the lobar and segmental bronchi showed severe active chronic mucosal inflammation with focal ulceration and squamous metaplasia. Macrophages, many of them foamy, and elongated cells having the features of fibroblasts predominated. There was focal fibrinous exudation but no distinct palisading of the cells. Plasma cells were present but lymphocytes were rather sparse and lymphoid follicles were not a feature. Mucus glands were not hypertrophied.

The most striking histological change was bronchiolitis obliterans. This was found to be extensive in the apical segments. In view of the physiological evidence of severe and widespread airways obstruction the macroscopically unremarkable parts of the lungs were also examined carefully and bronchiolitis obliterans was almost as widespread here as it was in the apical segments. Two-thirds of all the tissue blocks (58 in all) contained one to three severely narrowed or completely obliterated small bronchi or bronchioles. The affected airways measured about 0.5-2 mm in diameter. The changes, which were focal within the affected airways, consisted of luminal occlusion or severe

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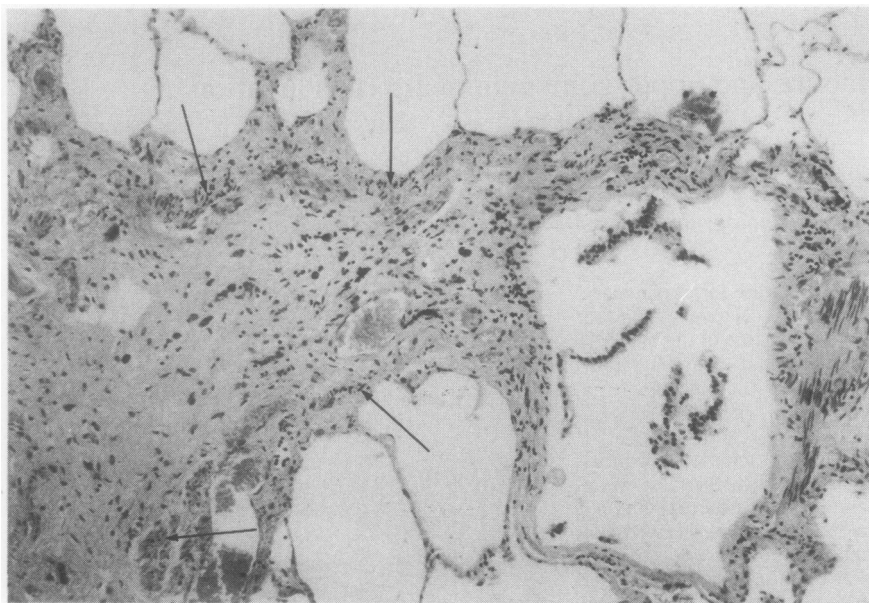


Fig 2 Cross-section of a small bronchus (right) and a completely obliterated branch in longitudinal section (left). Arrows indicate bronchial muscle. The surrounding lung parenchyma was essentially normal. (Haematoxylin and eosin, $\times 68$.)

reduction by vascular fibrous tissue (fig 2). Although an occasional obliterated airway contained tubular structures lined by columnar epithelium the nature of these scars could easily have been overlooked but for their constant location alongside pulmonary arteries. Elastic stains were helpful in identifying the lesions as obliterated bronchioles. Apart from florid obstructive pneumonitis in the apical segments the parenchyma around the affected airways showed no appreciable abnormality.

Discussion

Obliterative bronchiolitis is one of the pulmonary complications of rheumatoid disease,¹⁻⁴ usually being diagnosed only months before the patient's death. As this case shows, lungs which appear normal macroscopically may be affected by very extensive obliterative bronchiolitis, which only detailed and careful histological study will show. If a clinical and physiological diagnosis of bronchiolitis obliterans had not been made, the extent to which the macroscopically normal lung was affected might well have been overlooked.

Consolidation and fibrosis of the upper lobe in rheumatoid arthritis has been described in six patients.^{3,5,6} None had histological examination. This condition has been likened to the upper-zone fibrosis found in ankylosing spondylitis.⁷ In the present case this bilateral upper zone consolidation was due to an obstructive pneumonitis secondary to obliterative bronchiolitis.

Spencer states that in ankylosing spondylitis "Bronchiolitis obliterans also commonly occurs and causes obstructive airways disease and accumulation of foamy macrophage cells in the alveoli."⁸ In two of the cases reported by Davies⁷ the histological appearances might now be interpreted as obstructive pneumonitis. In one this appearance occurred at the transition from fibrotic to normal

lung. Neither author related the upper-zone fibrosis to the obstructive pneumonitis.

We believe that this is the first reported case in a patient with rheumatoid arthritis of bronchiolitis obliterans as the cause of bilateral upper zone consolidation. We might speculate about whether the previously reported upper zone consolidation and fibrosis in rheumatoid arthritis, and possibly in ankylosing spondylitis, were also secondary to obliterative bronchiolitis.

References

- Gosink BB, Friedmann PJ, Liebow AA. Bronchiolitis obliterans. Roentgenologic-pathologic correlation. *Am J Roentgenol* 1973;117:816-32.
- Geddes DM, Corrin B, Brewerton DA, Davies RJ, Turner-Warwick M. Progressive airway obliteration in adults and its association with rheumatoid disease. *Q J Med* 1977;46:427-44.
- Macfarlane JD, Dieppe PA, Rigden BG, Clark TJH. Pulmonary and pleural lesions in rheumatoid disease. *Br J Dis Chest* 1978;72:288-300.
- Murphy KC, Atkins CJ, Offer RC, Hogg JC, Stein HB. Obliterative bronchiolitis in two rheumatoid arthritis patients treated with pencillamine. *Arthritis Rheum* 1981;24:557-60.
- Petty TL, Wilkins M. The five manifestations of rheumatoid lung. *Dis Chest* 1966;49:75-82.
- Petrie GR, Bloomfield P, Grant IWB, Crompton GK. Upper lobe fibrosis and cavitation in rheumatoid disease. *Br J Dis Chest* 1980;74:263-7.
- Davies D. Ankylosing spondylitis and lung fibrosis. *Q J Med* 1972;41:395-410.
- Spencer H. *Pathology of the lung*. Vol 2. 3rd ed. Oxford: Pergamon Press, 1977:752.