

Intravascular bronchioalveolar tumour

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The intravascular bronchioalveolar tumour, a rare pulmonary neoplasm, was first described under this name by Dail and Liebow in 1975.¹ Subsequent reports have emphasised the histological appearances and histogenesis. We present the clinical and pathological findings in a further case.

Case report

A 24-year-old nulliparous woman, with no important past illness, presented in April 1981 complaining that she had had painful ankle swelling for six months and exertional dyspnoea for six weeks. She had been nursing since 1975, was a non-smoker, and had been taking an oral contraceptive until the onset of her symptoms. On examination there was gross clubbing of fingers and toes, with tender, non-pitting swelling of the wrists and ankles. Chest expansion was restricted and there were a few inspiratory crackles at the left base posteriorly. The chest radiograph, which had been normal in 1975, showed bilateral nodular shadowing, the nodules being uniformly distributed and varying from 0.5 to 1.0 cm in diameter (fig 1). Radiographs of ankles

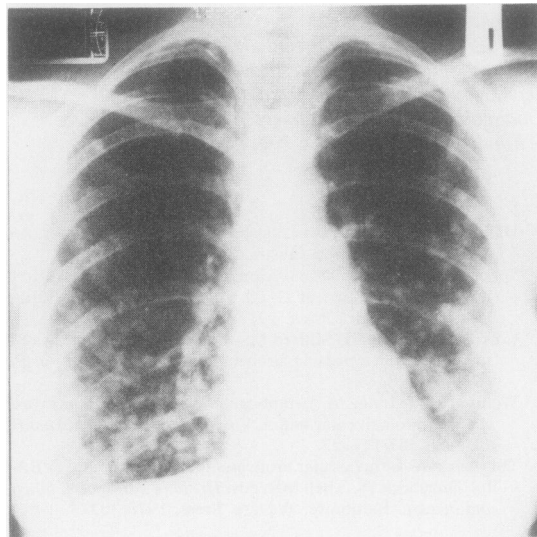


Fig 1 Chest radiograph on presentation showing bilateral nodular shadowing.

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and wrists showed the changes of hypertrophic pulmonary osteoarthropathy.

The erythrocyte sedimentation rate (ESR) (Westergren) was 46 mm in the first hour; the full blood count, blood urea and electrolyte concentrations, results of liver function tests, serum calcium concentration, and serum immunoglobulin levels were all within normal limits. The tests for rheumatoid factor (latex) and antinuclear factor and the Mantoux test gave negative results. There was a restrictive ventilatory defect with associated reductions in lung volumes to 70% of the predicted values. Gas transfer was normal. Fibreoptic bronchoscopy showed mucosal erythema but no other abnormality. The histological appearance of a transbronchial lung biopsy specimen was inconclusive and therefore an open-lung biopsy was performed through a left thoracotomy. Firm nodules up to 1.0 cm in diameter were found throughout the lung, with puckering of the overlying visceral pleura. Two wedge biopsy specimens were taken from the lingula.

The patient's joint symptoms were relieved by treatment with indomethacin and she returned to work. Azathioprine treatment was commenced. At follow-up in November 1981 there had been no change in her symptoms or in the radiographic appearances.

The biopsy samples were fixed immediately in 10% phosphate buffered formalin and 5- μ m thick paraffin sections were stained with haematoxylin and eosin, elastin Van Gieson, and Congo red and by the periodic acid Schiff reaction (PAS). All the nodules had a similar histological appearance and although not encapsulated they were well demarcated from the surrounding lung tissue. The nodules had a micronodular appearance with cellularity more pronounced peripherally. Collections of atypical vesicular cells were mixed with nodules of eosinophilic hyaline material, which protruded into alveoli and bronchioles in a micropolypoid manner (fig 2a). The centres of the nodules were fibrotic and only sparsely cellular, and "ghosts" of polypoid projections and occluded vessels could be recognised. Occasional atypical tumour cells were recognised within vessel walls (fig 2b). There was a mild interstitial inflammatory infiltrate, consisting of lymphocytes and plasma cells, within alveolar septa at the edge of the lesions, and adjacent alveoli were lined by cuboidal cells. The hyaline material at the periphery of nodules was weakly PAS positive but did not stain positively for amyloid. The appearances of the lesions were considered typical of the intravascular bronchioalveolar tumour.

Discussion

In 1975 Dail and Liebow, reporting on 20 cases seen from 1962 to 1975, drew attention to an apparently undescribed neoplasm of the lung which they thought to be an atypical

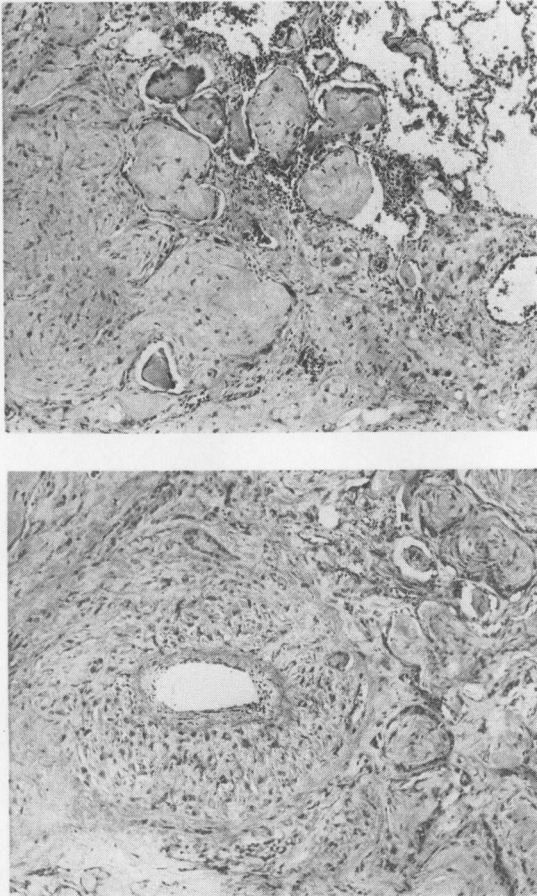


Fig 2 (a) Alveoli containing nodules of eosinophilic hyaline material and atypical cells (H and E, $\times 50$). (b) Nodules adjacent to a vessel with atypical cells within its walls (H and E, $\times 50$).

form of bronchioloalveolar tumour.¹ The multiple lesions, which had hypocellular fibrotic centres surrounded by a cellular zone, showed a great propensity for invading veins, arteries, alveoli, and bronchioles, which led the authors to coin the term intravascular bronchioloalveolar tumour. Six subsequent cases have been reported.²⁻⁵ In his 1977 review Spencer added "sclerosing" to the title⁶ and accepted the case of so-called pulmonary deciduosis recorded in 1972⁷ as the first report. On the basis of ultrastructural studies on material from three cases Corrin and colleagues rejected the hypothesis of an alveolar cell origin and suggested that

an angioblastic stem cell, such as the vasoformative reserve cell, is the most likely source of this tumour.² Weldon-Linne and colleagues support this conclusion and have advanced the term "sclerosing angiogenic tumour" as a more appropriate description.⁴

Only scanty descriptions of the clinical features of this tumour are available because the major report is in abstract form.¹ In that series over two-thirds of the patients were women, the age range was 14-71 years, and half were under 40. The seven subsequent cases (including our own) have all been women (ages 55, 49, 35, 28, 27, 26, 24). Patients have usually presented either with mild dyspnoea or with the appearances of multiple bilateral nodules on a routine chest radiograph. Chest pain,^{2,5} clubbing,⁸ haemoptysis, and arthralgia⁴ have also been recorded. In the case presented here the initial symptoms were due to hypertrophic pulmonary osteoarthropathy, which has not previously been described in this disorder.

The disease usually progresses slowly at first, with patients remaining well for up to six years after presentation.^{1,5} Later deterioration may be rapid leading to respiratory insufficiency. Three patients with the intravascular bronchioloalveolar tumour are known to have died¹ (at two, eight, and 12 years from presentation) but in many other cases details of the clinical progression are not available, and there is a need for more detailed reporting of the clinical features and results of treatment in patients with this unusual pulmonary tumour.

We are grateful to Drs CC Evans and JM Jefferson for permission to report this case and for help in the preparation of the report.

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