

Benign lymphocytic angiitis and granulomatosis

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ABSTRACT A 37 year old woman underwent a lobectomy for a lesion with a tumour like appearance on the chest radiograph. This was shown microscopically to be benign lymphocytic angiitis and granulomatosis, a rare condition that responds well to cytotoxic drugs and has a good prognosis.

In 1973 Liebow¹ introduced the concept of pulmonary angiitis and granulomatosis. The diseases grouped under this heading included classical Wegener's granulomatosis, limited Wegener's granulomatosis, lymphomatoid granulomatosis, necrotising sarcoid granulomatosis, and bronchocentric granulomatosis.² Four years later Saldana *et al.*³ suggested an alternative classification corresponding to Liebow's first three categories. Three histological types were recognised: Wegener's granulomatosis (lymphocyte depleted angiitis and granulomatosis), lymphomatoid granulomatosis (malignant lymphoproliferative angiitis and granulomatosis), and benign lymphocytic angiitis and granulomatosis. This last is a very rare disease, but because its prognosis is good and it responds well to cytotoxic chemotherapy its recognition is important.

Case report

A 37 year old non-smoking woman was admitted to hospital because a lesion unresponsive to antibiotics had been detected in the apical segment of the left lower lobe (fig 1). She had had a heavy, hacking cough for several months, but had no fever and was otherwise healthy. Routine investigations, including fiberoptic bronchoscopy, disclosed no abnormality. Subsequently the left lower lobe was resected. On gross examination the lobectomy specimen contained a tumour like mass a few centimetres in diameter, infiltrating locally into the visceral pleura.

Histologically there was a prominent vasculitis. Several vessels were occluded, and their walls were infiltrated by lymphocytes, plasma cells, and occasional histiocytes and giant cells (fig 2). The adjacent lung parenchyma was fibrotic and contained aggregates of lymphocytes. No necrosis was present. The lymphoid cells were highly differentiated, and there were no mitoses. The lesion was consistent in

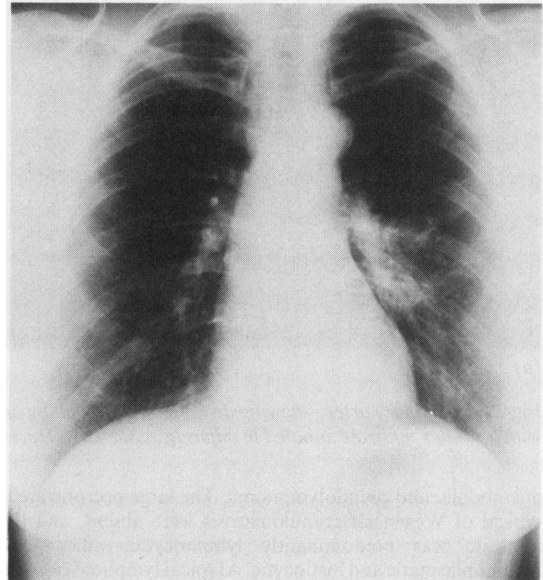


Fig 1 Chest radiograph showing a tumour like lesion in the left lower lobe.

appearance with benign lymphocytic angiitis and granulomatosis as defined by Saldana *et al.*³

The patient has been followed up for two years. There has been no evidence of recurrent lung disease or lesions of other organs. Cytotoxic chemotherapy has been withheld.

Discussion

Since the original 14 cases of benign lymphocytic angiitis and granulomatosis were described,^{3,4} only one further example has been published.⁵ This paucity of published cases can be attributed not only to the rarity of the disease and failure to recognise it as such but also to the controversy surrounding its existence as an entity.⁶ The controversy stems from Churg's⁶ observation that some of the original cases turned out to be lymphomatoid granulomatosis⁴ and in others the pathological description would fit pseudolymphoma or plasma cell granuloma.

Despite these objections, in our opinion benign lymphocytic angiitis and granulomatosis is a definite entity. In the present case the alternative diagnoses taken into consideration included Wegener's granulomatosis, lymphomatoid granulomatosis, lymphocytic interstitial

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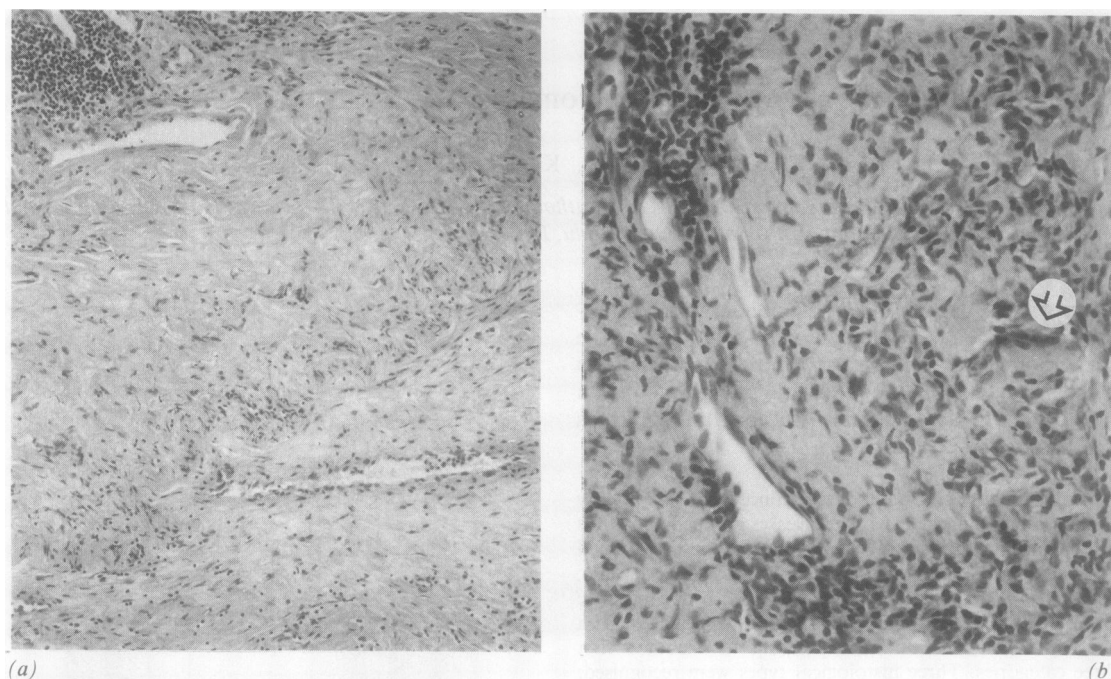


Fig 2 Pulmonary artery showing disruption of its wall by an infiltrate of lymphocytes, macrophages, and plasma cells (a), with a similar infiltrate situated in adjacent tissue (b) (Haematoxylin and eosin.)

pneumonia, and pseudolymphoma. The large necrotic areas typical of Wegener's granulomatosis were absent, and the infiltrate was predominantly lymphocytic rather than lymphoplasmatic and histiocytic. Atypical lymphoid cells are found in lymphomatoid granulomatosis, and the overall cytological appearance is more pleomorphic. A mild vascular infiltrate may be associated with pseudolymphoma, plasma cell granuloma, and lymphocytic interstitial pneumonia,⁶ but in our patient the vasculitic component was very prominent.

Benign lymphocytic angiitis and granulomatosis responds favourably and promptly to cytotoxic drugs, and where chlorambucil has been used resolution has been complete with no recurrences.^{3,4} On the other hand, after surgical resection alone the disease has recurred in almost half of the reported cases. In our patient no recurrence has been detected during the two year postoperative follow up.

A definitive diagnosis of benign lymphocytic angiitis and granulomatosis can be made only by histological examination, although the diagnosis is supported by the absence of extrapulmonary manifestations in most cases and a favourable response to cytotoxic drugs. The importance of a good tissue specimen, which can be obtained only by open biopsy, must be emphasised.^{4,5}

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