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Endobronchial involvement in Wegener's granulomatosis

RAJNI AMIN
M.B.B.S., M.R.C.P., F.R.C.R., D.M.R.T.

Radiotherapy Department, Royal Devon and Exeter Hospital, Exeter, Devon

Summary

Right middle lobe collapse as a result of endobronchial involvement with Wegener's granulomatosis is reported in a 60-year-old woman. A review of the literature has revealed 6 cases of lobar collapse, only 2 of which had endobronchial disease.

KEY WORDS: Wegener's granulomatosis, endobronchial disease, lobar collapse.

Introduction

In 1939, Wegener described 3 cases of necrotizing granuloma which involved the upper respiratory tract, the lungs and the kidneys. The renal lesion is the hallmark of generalized disease. A limited form of Wegener's granulomatosis involving only the upper and/or the lower respiratory tract has also been reported (Carrington and Liebow, 1966). The typical manifestation of pulmonary involvement is multiple nodules (Felson and Braunstein, 1958). The nodules may be single or multiple, well or poorly circumscribed, and vary in size from a few mm to 9 cm in diameter (Richards, Razavi and Leftwich, 1962). Rarely, lobar collapse, as a direct manifestation of Wegener's granulomatosis, may be the only pulmonary abnormality (Farrelly and Foster, 1980). A case of Wegener's granulomatosis with such a radiological abnormality is described here.

Case report

A 60-year-old woman presented in March 1973 with a 7-week history of sore throat, pain in the left jaw and ear, excessive sweating, cough and weight loss. Physical examination was unremarkable apart from 2 shallow ulcers on the soft palate. Laboratory studies were non-contributory. A chest X-ray showed loss of volume of the right middle lobe (Fig. 1). Subsequently, there was complete consolidation of the right middle lobe. Bronchoscopy showed narrowing of the right middle lobe bronchus, and there was a reddened ulcerated area beyond the middle lobe orifice. Histology of this showed marked fibrosis; epithelial and giant cell granulomata were also present and there was vasculitis with intimal thickening and elastic disruption (Fig. 2). She underwent right middle lobectomy. The right middle lobe bronchus was infiltrated by granulomatous tissue thereby causing consolidation of the resected lobe. The pathological diagnosis was Wegener's granulomatosis.

She remained well until January 1974 when she started to suffer recurrent epistaxis. There were several ulcerative lesions in both nostrils and in her throat. At this stage she started treatment with azathioprine, and was later changed to cyclophosphamide. She remained in clinical remission until February 1975 when she developed a transient right-sided hemiparesis. Thereafter, her condition deteriorated and she died in July 1975.

Post-mortem showed no abnormality in her nasal cavities, the gums, the fauces and the external ears. The lungs were devoid of granulomata. The kidneys were reduced in size, and the cortices were thinned, showing patchy fibrosis and streaking. There was vasculitis of the renal blood vessels.

Discussion

Roghair and Ross (1970) reported a case of Wegener's granulomatosis with radiologically complete consolidation of the right middle lobe; no endobronchial infiltration was evident at bronchoscopy. Gohel *et al.* (1973) in a review of 20 cases of Wegener's granulomatosis noted segmental involvement resembling consolidation in 2 more cases, but it was not known if these 2 cases had any endobronchial disease. Landman and Burgener (1974), in a review of 10 cases of Wegener's granulomatosis noted pulmonary consolidation in 2 cases, one of which was the subject of a previous report (Roghair and Ross, 1970).

In an attempt to establish the approximate incidence of the more common pulmonary radiological findings, they collected 77 cases from the literature and found that only 5 cases had an atypical

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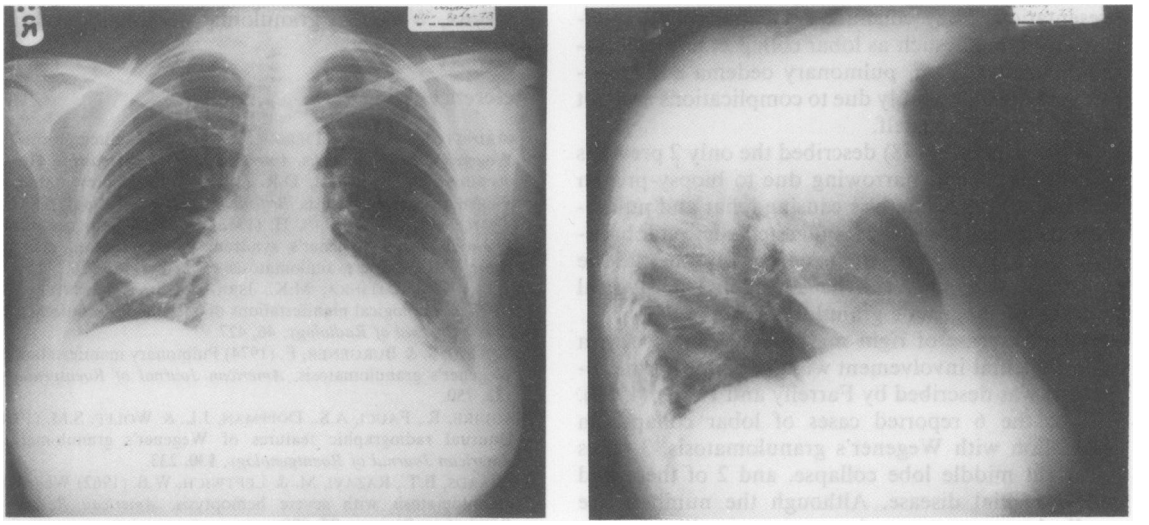

FIG. 1. Posterior-anterior and lateral views of chest showing partial atelectasis of right middle lobe.

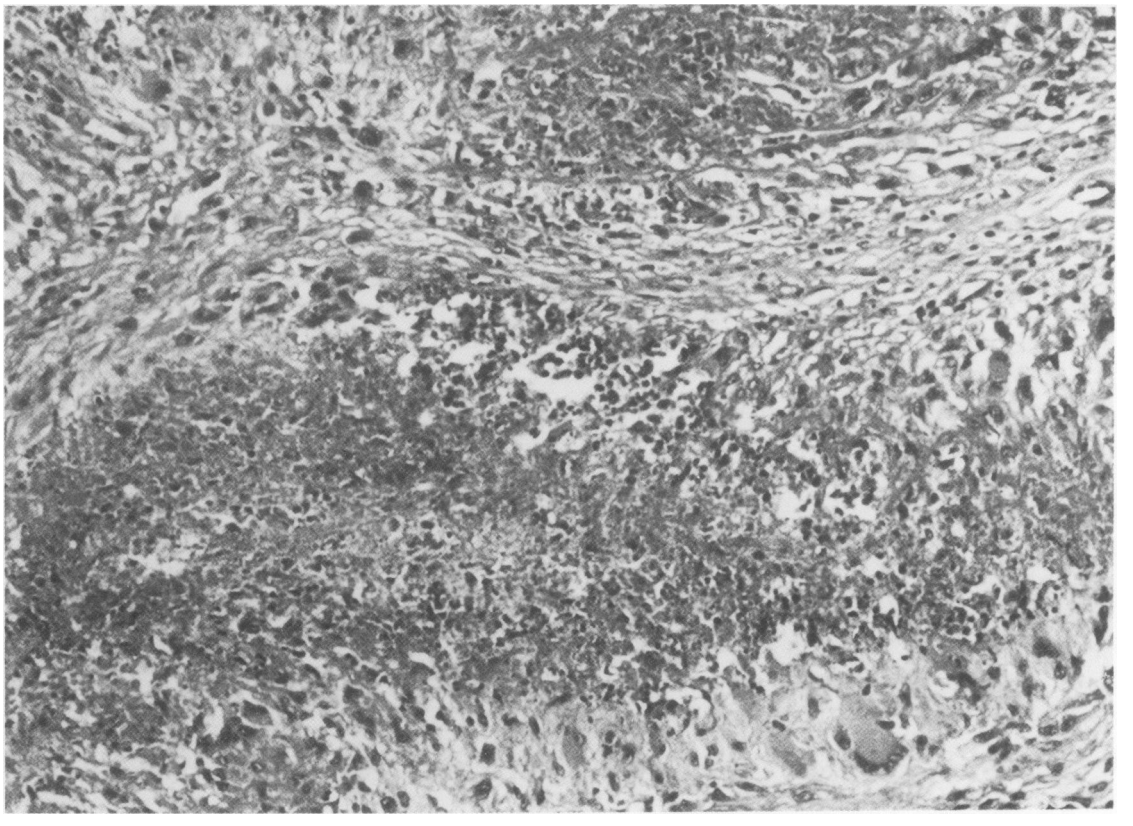

FIG. 2. Granuloma shows a necrotic core circumscribed by a palisade of large cells including occasional multinuclear giant cells (H.E. ×266).

presentation. They concluded that pleural and pulmonary findings such as lobar collapse, bronchiectasis, pleural effusion, pulmonary oedema and pneumothorax were possibly due to complications and not due to the disease itself.

Maguire *et al.* (1978) described the only 2 previous cases of bronchial narrowing due to biopsy-proven Wegener's granulomatosis causing lobar and pulmonary collapse. They even suggested that endobronchial disease, resulting in atelectasis of either a lobe or a complete lung, can be considered as 'typical findings' of Wegener's granulomatosis.

A further case of right middle lobe collapse with endobronchial involvement with Wegener's granulomatosis was described by Farrelly and Foster (1980). Out of the 6 reported cases of lobar collapse in association with Wegener's granulomatosis, 3 cases had right middle lobe collapse, and 2 of these had endobronchial disease. Although the numbers are small, the present case also suggests predilection for right middle lobe collapse in Wegener's granulomatosis. When lobar collapse and consolidation coexists with mucosal changes in the nose, the mouth and the pharynx, Wegener's granulomatosis should be considered.

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