

# Hilar and mediastinal lymphadenopathy in the limited form of Wegener's granulomatosis

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## Abstract

**A patient with the limited form of Wegener's granulomatosis is reported. The case is unusual because of hilar and mediastinal lymphadenopathy, severe ulceration of the respiratory and digestive tracts, and the rapidly fatal outcome.**

Wegener's granulomatosis is a disease characterised by necrotising granulomatous vasculitis of the upper and lower respiratory tracts, glomerulonephritis, and variable degrees of vasculitis, predominantly of small arteries and veins. The limited form of the disease is defined mainly by the absence of glomerular lesions, predominant lesions of the lower respiratory tract, and a better prognosis. We report a fatal case of the limited form of Wegener's granulomatosis associated with hilar and mediastinal lymphadenopathy and severe ulceration of the respiratory and digestive tracts. The diagnosis was confirmed histologically.

## Case report

A 60 year old man who had worked 20 years in a sulphur mine complained of shortness of breath that had lasted for a year. He was admitted with fever, cough, expectoration of mucoid sputum, dyspnoea, gingival and

pharyngeal inflammation, and inspiratory rales, predominantly in both lower lung fields. The chest radiograph showed diffuse bilateral reticulonodular shadowing and moderate prominence of both hilar regions. Sinus films showed mucosal thickening of the right maxillary sinus. The erythrocyte sedimentation rate was 121 mm in the first hour. The leucocyte count was  $24.3 \times 10^9/l$ , with no shift to the left. Concentrations of polyclonal IgG and IgA were increased. The urinary sediment was normal and no proteinuria was found. Rheumatoid factor and antinuclear antibody were present in low titre. Blood, urine, and sputum cultures were sterile. Microimmunofluorescent antibody tests for chlamydia, mycoplasma, rickettsia and legionella gave negative results. Lung function studies showed a restrictive pattern; arterial blood gas analysis when the patient was breathing room air showed an oxygen tension of 10.0 kPa, a carbon dioxide tension of 4.9 kPa, and a pH of 7.48.

Erythromycin was prescribed, with no improvement. Two weeks later multiple large, deep, destructive punched out ulcers developed on the lips, tongue, oral cavity, and nasal septum. Biopsy of the borders of the ulcers of the nasal septum and the tongue showed chronic inflammation only. Fibreoptic bronchoscopy was not tolerated by the patient. Ten days later papular, pustular, and ulcerated lesions developed on the trunk and limbs. Biopsy of the skin lesions showed perivascular multinucleated giant cells. Intravenous methylprednisolone 2 mg/kg a day and cyclophosphamide 2 mg/kg a day were given, but the patient died two days later.

Necropsy showed hilar, mediastinal, diaphragmatic and peripancreatic lymphadenopathy, with glands ranging up to 4.5 cm in diameter. There were no large nodular or cavitating lesions in the lungs. Multiple ulcers in the trachea (5 cm major axis) and main bronchi were seen. There were numerous ulcers of 2 cm diameter in the oesophagus and small and large intestines. Microscopic examination of the lungs and lymph nodes showed nodular cicatricial areas and granulomatous lesions with a necrotic centre surrounded by lymphocytes and epithelioid and multinucleated giant cells (fig 1). Similar lesions were found in the liver, spleen, and gall bladder. Necrotising vasculitis was present in the lungs, lymph nodes (fig 2), larynx, trachea, and intestine. The kidneys appeared normal by both light and electron microscopy. Histochemical staining for detecting fungi, mycobacteria, and other organisms was negative.

## Discussion

Wegener's granulomatosis is characterised by the triad of sinusitis, pulmonary infiltrates, and glomerulonephritis, with less frequent lesions of joints, skin, eyes, ears, nervous system, heart, oral cavity, and testes.<sup>1-2</sup>

Hilar and mediastinal adenopathy has been thought not to be part of the clinical picture. Thoracic lymph node enlargement in a patient with pulmonary infiltrates has usually been held

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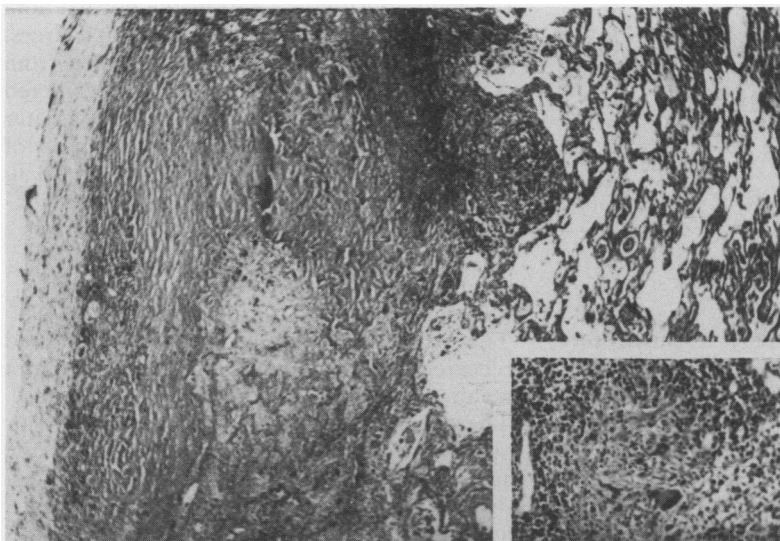


Figure 1 Nodular areas in the lung with necrotic centre surrounded by fibrous scar tissue and multinucleated giant cells, with a higher magnification inset.

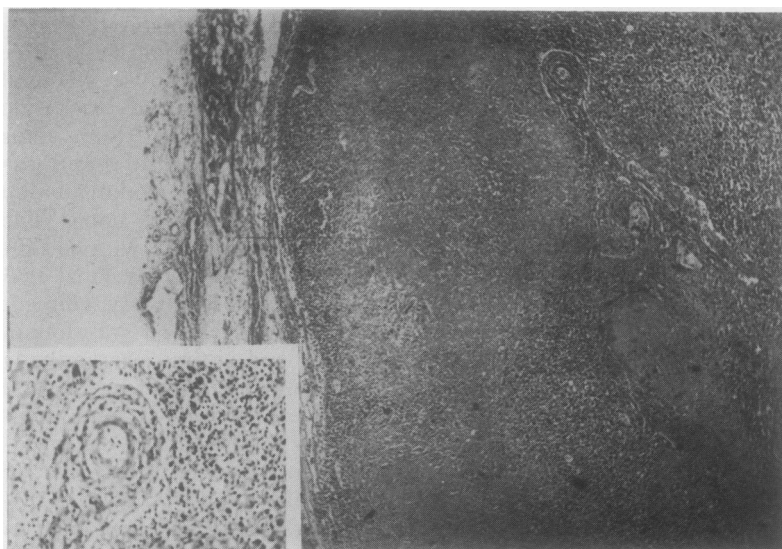


Figure 2 Hilar lymph node with vasculitis and necrotic areas surrounded by fibrous scar tissue and multinucleated giant cells, with a higher magnification inset giving a more detailed view of the vasculitis.

to exclude the diagnosis of Wegener's granulomatosis in favour of other conditions. Fauci *et al.*,<sup>2</sup> in their series of 85 patients with 21 years' follow up, did not see hilar lymphadenopathy. We emphasise the hilar, mediastinal, diaphragmatic, and peripancreatic adenopathy found in our patient. Histologically, the lymph nodes showed both granulomas and necrotising vasculitis, indicating that the lymph node changes were due to Wegener's granulomatosis.

Single or multiple nodular masses, with or without cavitation, are the most common pulmonary manifestations in Wegener's granulomatosis. An atypical diffuse reticulonodular pattern was found in our patient. The endobronchial lesions in Wegener's granulomatosis are usually granulomas and secondary to pul-

monary lesions. They occasionally obstruct the airway, and endoscopic removal is then necessary.<sup>3</sup> Our patient had punched out ulcers in the larynx, trachea, and bronchi.

Gastrointestinal ulcers have been described rarely in Wegener's granulomatosis. None of the published reports<sup>4-5</sup> describes ulcers throughout most of the digestive tract from lips to colon, as in our case.

Renal lesions are present in 83% of patients with Wegener's granulomatosis,<sup>6</sup> focal and segmental glomerulonephritis being usual. The absence of glomerular lesions is characteristic of the limited form of the disease. In our case there were no renal lesions, although this limited form of Wegener's granulomatosis was nevertheless associated with severe multi-system disease leading to the patient's death.

We conclude that Wegener's granulomatosis should still be considered in the differential diagnosis when the chest radiograph is atypical, despite enlargement of hilar or mediastinal lymph nodes.

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## Successful treatment of *Bacillus cereus* infection with ciprofloxacin

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### Abstract

*Bacillus cereus* is rarely a pulmonary pathogen but may cause pneumonia in immunocompromised patients. A patient with bronchiectasis and no recognisable immunodeficiency had this organism isolated during two infective exacerbations, once from respiratory secretions and once by blood culture. Ciprofloxacin treatment was effective on both occasions.

Aerobic Gram positive spore bearing bacilli of the genus *Bacillus* are part of the normal flora of soil, dust, and other environmental sources. Although most *Bacillus* species isolated from clinical material can usually be considered irrelevant contaminants, *B cereus* has a greater pathogenic potential. We report a case of recurrent pneumonia and bacteraemia due to *Bacillus cereus* that was treated with ciprofloxacin.

### Case report

The patient, a 21 year old man, had had recurrent chest infections from childhood as a result of bronchiectasis of unknown cause. There was no history of whooping cough or other important early childhood illness. At the age of 12 years he had been extensively investigated and serum electrophoresis findings, immunoglobulin concentrations, ciliary function, and sweat test results were all normal. The diagnosis was based on the history and