Spontaneous pneumothorax in Wegener's granulomatosis

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Wegener's granulomatosis is a disease characterised by necrotising granulomatous angiitis of the upper and lower respiratory tracts, a necrotising glomerulonephritis, and a diffuse small vessel vasculitis that may affect almost any organ system.¹ Various pulmonary lesions have been described, but pneumothorax has seldom been mentioned. We report a patient who developed a spontaneous pneumothorax with a bronchopleural fistula and pyopneumothorax. So far as we are aware this complication has been recorded only once.²

Case report

A 33-year-old man presented with a 12-month history of recurrent pains in his ankles, knees, and wrists. For two months he had had nasal congestion and epistaxes, and finally a spontaneous left periorbital haematoma. Examination showed bilateral episcleritis, vasculitic lesions at the elbows, and an active arthritis of the ankles. The haemoglobin concentration was 12.4 g/dl, WBC $8.0 \times 10^9/l$, platelets $377 \times 10^9/l$, and ESR 105 mm in one hour (Westergren). Screening for hepatitis B, antinuclear factor, and rheumatoid factor gave negative results. The urine contained protein and red-cell and granular casts. The patient subsequently developed a purpuric rash on the extremities, splinter haemorrhages, and a cough with mucoid sputum. Nasal perforation and palatal ulceration followed, as well as a left foot drop. The blood pressure was 140/70 mm Hg. The initial chest radiograph showed two small nodular lesions in the left mid-zone and subsequently a cavitated nodule in the right mid-zone. A biopsy specimen from the hard palate showed severe suppuration, with necrosis, and small granulomatous foci with giant cell formation.

He was treated with cyclophosphamide 150 mg and prednisone 100 mg daily. Although the ESR gradually fell his renal function (blood urea concentration before treatment 14.9 mmol/l (89.8 mg/100 ml)) steadily deteriorated and he developed a chest infection with purulent sputum containing *Proteus mirabilis*, group B haemolytic streptococci, and *Bacteroides fragilis*. Despite the introduction of antibiotics he developed a right pyopneumothorax. Intercostal tube drainage with suction failed to reflate the lung and the pyopneumothorax persisted (fig). Haemodialysis was commenced in view of the severe renal failure. He developed neutropenia ($1.6 \times 10^9/l$) in addition to considerable lymphopenia ($0.15 \times 10^9/l$), necessitating a temporary cessation of cyclophosphamide, which was later reintroduced at a lower dose after improvement of the neutrophil count. Increasing infection, however, with super-

Chest radiograph showing persistent pyopneumothorax despite intercostal tubular drainage. A nodular lesion is present in the left mid-zone.

added growths of *Pseudomonas pyocyaneus*, *Streptococcus faecalis*, and *Candida albicans* in the empyema led to septicæmia, with deterioration and death six weeks after admission. At necropsy there was a small subpleural nodule in the left upper lobe and a cavitated mass 3 cm in diameter in the right upper lobe. Histological examination showed areas of consolidation with central necrosis and necrotising vasculitis. No fistula was seen between the cavity in the right lung and the pleural space. In the kidneys there was a proliferative glomerulonephritis.

Discussion

At presentation this patient had small nodules in the left lung and subsequently a cavitated lesion in the right lung. This is the most common radiographic manifestation of Wegener's granulomatosis but several others have been recorded.³⁻⁵ Pneumothorax is clearly an uncommon feature of the disease, occurring in one of the 20 patients described by Gohel *et al.*,³ one of the 24 cases of Saldana *et al.*,⁶ and one of the 31 cases reviewed by Maguire *et al.*⁵

Information on the pathogenesis of pneumothorax in published reports is scanty. Cavitated nodules might be expected to rupture, as is known to occur with intrapulmonary rheumatoid nodules.⁷ Epstein *et al.*⁸ did not believe that this occurred in either of their two cases because the chest film was normal after treatment of the pneumothorax

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in one, and they found no evidence of nodular rupture in the second. In the case described by Maguire *et al*⁵ there were pathological changes of Wegener's granulomatosis in the bronchopleural fistula and clearly the necrotic granulomatous lesion had ruptured into the pleural space. Unfortunately in our case no fistula was found at necropsy, but one must have existed in view of our failure to reflate the lung despite prolonged suction. Whatever the usual pathogenesis, it has been associated with infection in only one previously reported case.² Our patient had a cavitated nodule and developed a pyopneumothorax during an uncontrolled infection. This strongly suggests that secondary infection in a cavitated nodule precipitated the pneumothorax.

Treatment with immunosuppressive drugs, in particular cyclophosphamide, has undoubtedly improved survival rates in this previously fatal condition.⁶⁻⁹ Corticosteroid treatment alone was by comparison disappointing¹⁰ but may be beneficial for some features of the disease when combined with cyclophosphamide.¹ In our patient, however, there was no response to the combined treatment. He appeared to have a fulminating renal lesion when treatment began and renal failure progressed inexorably.

Wolff and his colleagues¹ did not encounter any infections attributable to cyclophosphamide, which is contrary to the experience of Israel *et al*,⁹ who reported staphylococcal infection in two of 10 patients. Despite careful monitoring our patient developed considerable leucopenia, which during corticosteroid treatment and in the presence of renal failure clearly contributed to the relentless pulmonary and pleural infection.

We have reported this case to draw attention to pneumothorax as a rare complication of Wegener's granulomatosis, to suggest that this may result from secondary infection in a cavitated intrapulmonary nodule, and to emphasise that every precaution should be taken to guard against secondary infection, particularly in patients with

cavitated nodules and particularly during immunosuppressive treatment.

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