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Severe steroid responsive pneumonitis associated with pyoderma gangrenosum and ulcerative colitis

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Inflammatory bowel disease is associated with a variety of extraintestinal manifestations,[1] including bronchiectasis[2 3] and chronic bronchitis.[2] We report here a patient with quiescent ulcerative colitis who became critically ill with severe pneumonia. Despite intensive investigation, the likely nature of the pulmonary condition became only apparent when after eight weeks she developed pyoderma gangrenosum. Both the pneumonia and the pyoderma responded promptly to high dose corticosteroid treatment only to relapse subsequently at the time of an intercurrent illness. Reinstitution of high dose corticosteroid treatment again resulted in resolution of the lesions. Had we been aware that such pulmonary lesions may occur in patients with inflammatory bowel disease, we would have treated the patient more promptly with corticosteroids.

Case report

A 53 year old non-smoking nursing sister presented in February 1982 with bloody diarrhoea. Rectal biopsy showed active chronic proctitis. Her chest radiograph was normal and a barium enema showed granular mucosa in the rectum and sigmoid colon. She was unable to tolerate sulphasalazine but her symptoms settled rapidly with prednisolone enemas. She remained well until July 1983, when she developed right sided pleuritic chest pain, cough, and purulent sputum. A chest radiograph showed consolidation in the apical segment of the right lower lobe (fig 1). Co-trimoxazole and subsequently doxycycline were prescribed, with no improvement in her symptoms.

She was referred to hospital in August 1983 with the same symptoms. The proctitis was quiescent. Examination showed her to be pale and febrile (38.5°C), distressed by pleurisy but not cyanosed or dyspnoeic at rest. Her pulse was regular at 100 beats/min. There was dullness to percussion and coarse crackles and bronchial breathing were audible at the right base. Examination otherwise showed nothing abnormal. Investigations showed: haemoglobin 8.5 g/dl, mean cell volume 75, white blood cells 75, 14.3×10^9/l (90% neutrophils, no eosinophilia), erythrocyte sedimentation rate 130 mm in one hour; biochemical screen normal; immunoglobulins and serum complement normal; autoantibodies and circulating immune complexes negative; serum iron and iron binding capacity both reduced, serum B_{12} and folate normal; repeated sputum cultures, including anaerobic cultures and culture for tubercle bacilli, sterile; blood and urine cultures also sterile; virus and mycoplasma titres not raised; and precipitins to *Aspergillus*, *M faeni*, and avian antigen not detected. A chest radiograph confirmed consolidation in the apical segment of the right lower lobe (fig 1); a lung scan showed a matched defect of perfusion and ventilation in the right

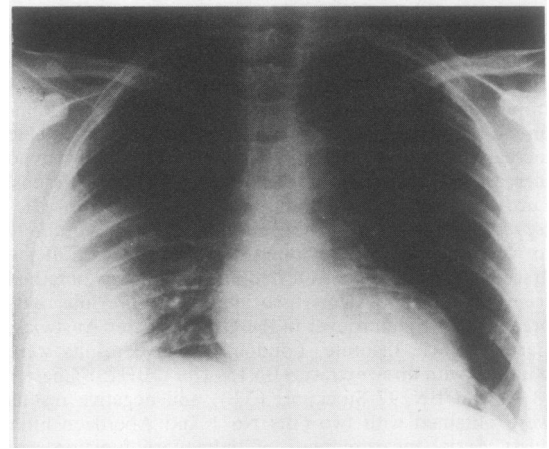

Fig 1 *Chest radiographic appearances during the admission to hospital in August 1983.*

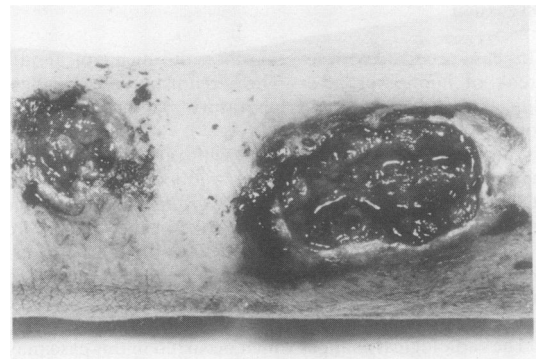

Fig 2 *Right forearm showing pyoderma gangrenosum.*

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mid zone. Bronchoscopy and abdominal ultrasound examination showed normal appearances.

Despite treatment with erythromycin orally for five days, ampicillin intravenously, and subsequently orally, for nine days, and then gentamycin and metronidazole for 10 days, the fever persisted, the sputum remained purulent and the patient's general condition deteriorated. A percutaneous lung biopsy was performed, which showed a chronic non-specific organising pneumonia. No organisms were seen, there was no evidence of vasculitis, and there were no granulomas. Culture of the biopsy specimen was negative. In late September 1983, eight weeks after the onset of the illness, her diarrhoea returned and there was some rectal bleeding. She developed necrotic ulcerating lesions on the right forearm and on both shins (fig 2). The clinical and histological features were typical of pyoderma gangrenosum. She was started on high dose (60 mg/day) prednisolone treatment and she rapidly improved. The purulent sputum disappeared, there was rapid resolution of the pulmonary and cutaneous lesions, and the diarrhoea stopped. She was discharged in October 1983 having maintenance corticosteroid treatment orally and prednisolone enemas.

She continued to improve until January 1984, when she was referred with a right popliteal cyst. Straw coloured fluid (10 ml) was aspirated under local anaesthetic. Seven days later she was readmitted to hospital with severe pain and swelling in the right calf. She had minimal sputum and no diarrhoea. Her chest was clinically clear and the chest radiograph showed only minor residual changes in the right middle lobe. Blood, urine, and sputum cultures were negative. She was treated with intravenous cefuroxime but did not improve. The popliteal cyst was incised and 100 ml of sterile pus was drained. Her clinical condition deteriorated over two weeks, when both the diarrhoea and pyoderma gangrenosum returned. She developed a cough productive of sterile purulent sputum along with clinical signs of left basal pneumonia. A chest radiograph showed peripheral consolidation in the left lower zone. High dose corticosteroid therapy was reinstituted and the pyoderma and pneumonia rapidly settled, as did the diarrhoea. A colonoscopy was performed and the mucosa was normal throughout. Multiple biopsy specimens showed no evidence of colitis, granulomas, or malignancy.

Discussion

Pulmonary complications of inflammatory bowel disease are uncommon and previous reports[1 2] have almost exclusively described bronchiectatic change. The diagnosis of inflammatory bowel disease in the present patient seems clearly established on the basis of the initial enema and rectal biopsy. The diarrhoea was controlled with prednisolone enemas and recurred only in association with pyoderma and pneumonia. Colonoscopy, however, performed while she was receiving high dose corticosteroid treatment, was normal. We were unable totally to exclude a bacterial pneumonia during her first admission because of prior antibiotic treatment and for this reason we hesitated to give corticosteroid treatment. The prompt response of the lesion to corticosteroids and its association with pyoderma gangrenosum suggests, however, that the pneumonia represented a previously underscribed association of inflammatory bowel disease. Further and more positive evidence in favour of this hypothesis came from the subsequent development of pulmonary consolidation in the opposite lung, associated with diarrhoea and pyoderma gangrenosum during an intercurrent illness, and the brisk response of both lesions to the reintroduction of high dose corticosteroid treatment.

The temporal association of the pulmonary consolidation with the pyoderma is intriguing. Pyoderma was first described in association with ulcerative colitis.[4] It can precede colitis or may occur at any stage of the disease, even after the colon has been removed. The exact mechanism is uncertain but there is evidence that it may represent a Schwartzmann type of reaction.[5] It is tempting to speculate that, whatever the cause, a similar mechanism may also have been responsible for the pulmonary lesions. But none of the characteristic histological features of pyoderma (venous and capillary fibrosis, haemorrhage, necrosis, and massive cell infiltration) was observed in the lung biopsy specimen. An alternative hypothesis is that patients with inflammatory bowel disease may have altered immunological defence mechanisms that render them susceptible to bacterial pneumonia, and that this deficiency may be ameliorated by corticosteroid treatment.

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