

## Small intestinal perforation in Wegener's granulomatosis

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### Summary

A case of Wegener's granulomatosis complicated by small bowel ulceration and perforation is described. Following laparotomy, with bowel resection, haemodialysis and immunosuppressive therapy, a full recovery was achieved including reversal of severe renal failure.

### Introduction

Gastro-intestinal involvement in Wegener's granulomatosis is rare although Carrington and Liebow (1966) described a limited form of the disease without renal involvement in which 2 out of 16 cases had haemorrhage from jejunal ulcers. In Walton's (1958) 54 cases, intestinal involvement was listed in 13 although no case of intestinal perforation was described. A case of Wegener's granulomatosis with small bowel perforation is now reported.

### Case report

A 50-year-old company executive presented with a 9-month history of arthralgia, nasal blockage and epistaxis. In the month before admission he had become lethargic, feverish, and developed pleurisy with haemoptysis together with painful finger nodules. Before these symptoms he had been well and on no medication apart from ketoprofen for his arthralgia. There were no abdominal symptoms before admission.

He was anaemic. There were signs of cutaneous vasculitis with tender finger nodules and a purpuric ankle rash. Examination of the nose under local anaesthetic revealed crusting, granulation and ulceration. The chest was clear. Within one week of admission he developed pharyngeal and mouth ulceration.

### Investigations

Haemoglobin 7.4 g/dl; ESR 90 mm/hr; white blood count  $10.9 \times 10^9/l$  (87% neutrophils, 2% eosinophils); chest X-ray showed diffuse alveolar

shadowing spreading from the hilum; barium meal and sinus X-rays normal; urine showed microscopic haematuria and proteinuria 1.67 g/24 hr; blood urea 18 mmol/l; creatinine 582  $\mu\text{mol/l}$ ; creatinine clearance 9.4 ml/min; Rose Waaler positive 1:320, latex positive 1:128; immunoglobulins, complement C3 and C4 normal; nasal biopsy, inflammatory infiltrate including multi-nuclear giant cells with occasional foci of necrosis (Fig. 1); renal biopsy (at laparotomy) suggestive of sub-acute focal glomerulonephritis.

Following admission his general condition continued to deteriorate rapidly. Prednisolone 40 mg and azathioprine 150 mg daily were started. Three days later renal function had deteriorated further with urea reaching 67.5 mmol/l and potassium 6.8 mmol/l necessitating peritoneal dialysis for 12 hr followed by haemodialysis. Within a few hours of commencing haemodialysis he experienced severe abdominal pain and vomiting with signs of peritonitis. Laparotomy revealed 2 perforated ulcers with at least 9 other ulcers within the distal 90 cm of ileum (Fig. 2). A 55-cm resection of terminal ileum was performed leaving 2 proximal ulcers. Histology of the resected ileum showed non-specific ulceration.

Cyclophosphamide was substituted for azathioprine, and prednisolone was continued at 60 mg/day. He required haemodialysis for 3 weeks, and remained free of further abdominal complications with complete regression of oropharyngeal, nasal and cutaneous vasculitic lesions.

Currently, 6 months after presentation, he is asymptomatic with a creatinine clearance of 70 ml/min on treatment with prednisolone 10 mg daily and cyclophosphamide 100 mg daily.

### Discussion

This case satisfies the diagnostic triad for Wegener's granulomatosis with necrotizing granulomatous lesions of the respiratory tract, generalized

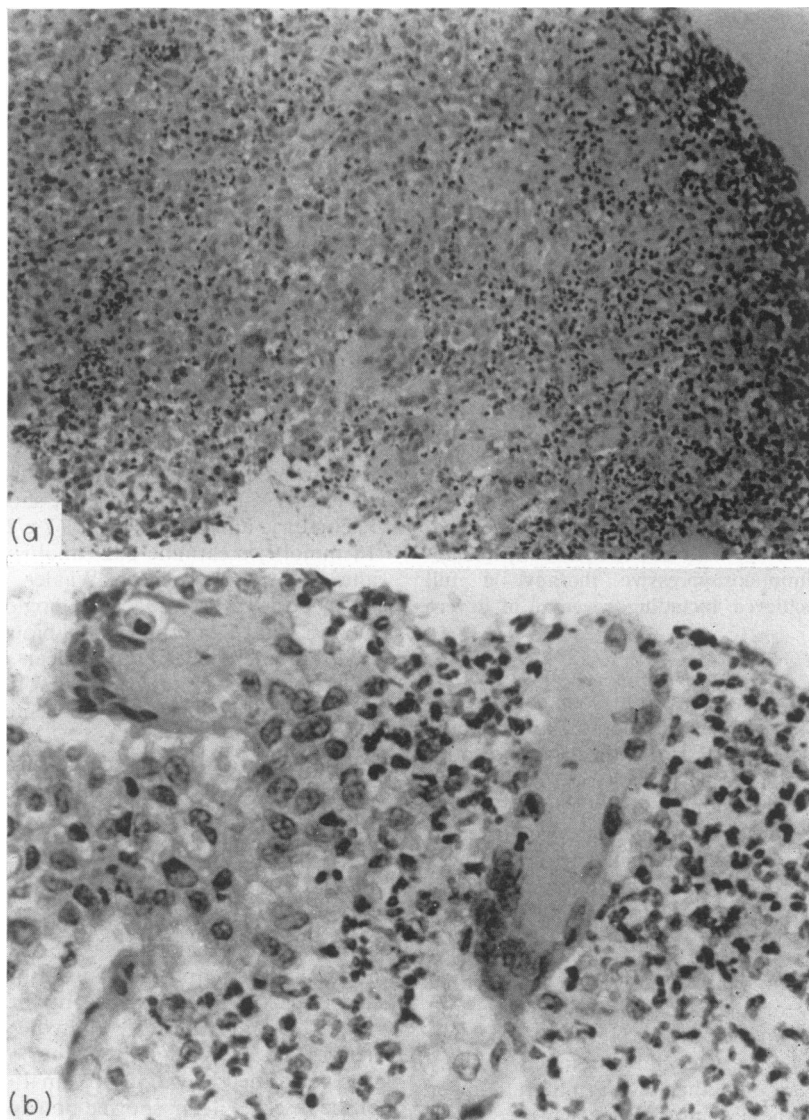


FIG. 1. Nasal biopsy showing chronic granulation tissue and inflammatory reaction including multinuclear giant cells (a) (HE,  $\times 240$ ); (b) (HE,  $\times 800$ ).

focal necrotizing vasculitis, and necrotizing glomerulitis. The dramatic response to immunosuppressive therapy particularly cyclophosphamide is well recognized (Wolff *et al.*, 1974). Unfortunately small bowel vasculitis was not confirmed histologically in the resected specimen. However, there is no evidence incriminating corticosteroids as an aetiological factor in small intestinal ulceration. Although intestinal necrosis and perforation have been

reported with the administration of high dose corticosteroids and azathioprine following renal transplantation (Demling, Salvatierra and Belzer, 1975), no such problems were encountered when this combination was used in the long-term treatment of ulcerative colitis (Jewell and Truelove, 1974). Finally, the continuation of immunosuppressive therapy following bowel resection, together with remission of bowel and multisystem involvement,

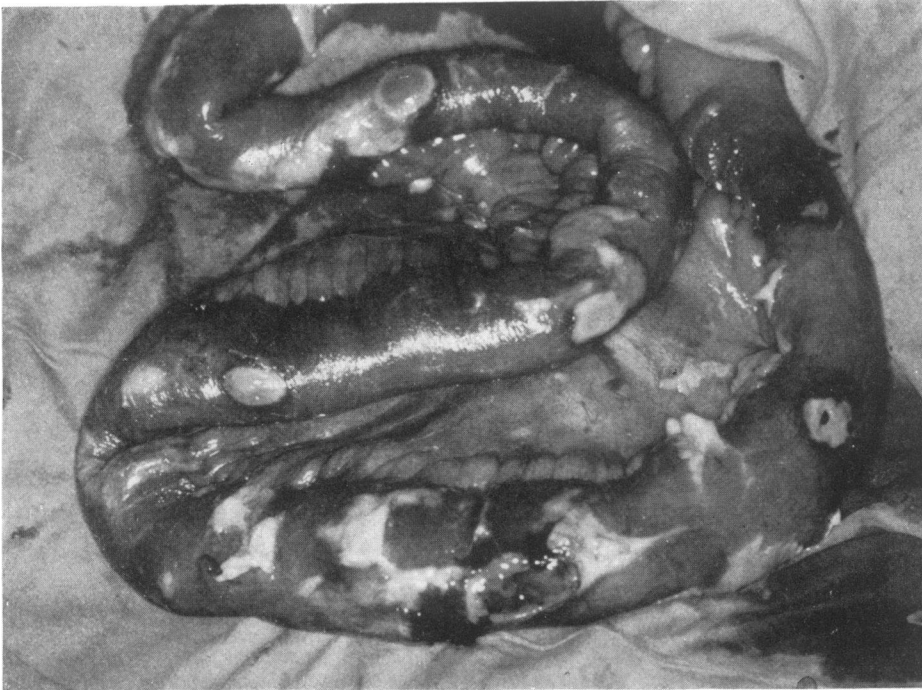


FIG. 2. Distal ileum at laparotomy showing multiple full thickness ulceration.

is further evidence in favour of small intestinal perforation related to Wegener's granulomatosis.

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