Intestinal perforation as a presentation of Wegener's granulomatosis

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INTRODUCTION

Wegener's granulomatosis is a necrotizing vasculitic disease principally affecting upper and lower airways and commonly the kidneys. The disease process may, however, manifest itself in any organ system and cases continue to be reported to substantiate this (Hoffman et al, 1992).

Intestinal involvement has been reported as a less common sequela of the disease process (McNabb et al, 1982; Storesund et al, 1998), and such symptoms usually occur within the first 2 years of onset of the disease. A single case report (Haworth and Pusey, 1984) outlines an 'inflammatory bowel' presentation of Wegener's granulomatosis. Perforation resulting in an acute abdomen has been reported in patients receiving active treatment for Wegener's in the form of systemic steroid therapy (Geraghty et al, 1986), which itself can result in intestinal perforation (Matolo et al, 1976). In this report we present a case of Wegener's granulomatosis presenting acutely as intestinal perforation and subsequently developing a more typical distribution pattern of organ involvement.

CASE REPORT

A 69-year-old man presented to casualty with a history of abdominal pain and progressive abdominal distention for 5 days. This was accompanied by nausea, vomiting, anorexia and altered bowel habit. He reported weight loss amounting to 5 kg predating these acute symptoms. He had no significant past medical history and took no medication.

On examination he was dehydrated, comfortable while laying still, apyrexial at 37°C, blood pressure was 150/80 and pulse rate was regular at 90 beats per minute. His abdomen was distended and tympanic and was tender on deep palpation, principally around the umbilical region. Per rectal examination was normal. Bowel sounds were absent. Auscultation of the chest was clear.

A plain abdominal film revealed distended loops of proximal small bowel, while erect chest X-ray demonstated no free gas under the diaphragm and was otherwise normal. His full blood count revealed a leucocytosis of 33 000/ml and of note was a urea level of 10.4 in the presence of normal serum creatinine.

A laparotomy was undertaken which revealed the presence of free seropurulent fluid in the peritoneum with a variegated contiguous dusky segment of distal jejunum approximately 2 feet in length and containing several perforations of less than 0.5 cm diameter. The remaining small intestine was entirely normal and the superior mesenteric artery was pulsatile along with the arterial cascade to the jejunum. No colonic pathology was apparent. The affected length of intestine was resected and the two ends exteriorized as a double-barrelled enterostomy.

He made an early uneventful recovery. On the second postoperative day he had a sudden and total loss of vision in his left eye. Fundoscopy revealed a central retinal artery occlusion. Following heparinization and low dose aspirin there was partial recovery of sight in the following 48 hours. At this time he developed a purpuric rash on his left hand followed closely by the right hand (*Figure 1*) and both feet. The impression was that of either embolic or vasculitic disease. Urgent echocardiography demonstrated left ventricular dilatation and an apical aneurysm without any evidence of endocarditis. Liver function became deranged. Renal function started to deteriorate with creatinine now becoming elevated. Urinalysis revealed microscopic haematuria and proteinuria. No casts were seen.

Increasing dyspnoea prompted repeat chest X-ray, which revealed patchy infiltration of the left upper lobe and subsequent computed tomography examination revealed upper lobe fibrosis and cavitation in the left lung. He developed extensive sacral pressure sores within days that extended into the natal cleft and rapidly became necrotic and malodorous. While investigating these manifestations, the possibility of Wegener's disease was highlighted and a positive cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) along with a negative rheumatoid factor was obtained. He was commenced on cyclophosphamide and methylprednisolone. Renal function continued to deteriorate and following a cerebrovascular accident, no further aggressive management was undertaken. He subsequently died 20 days after admission.

DISCUSSION

This case fulfills the diagnostic criteria of Wegener's granulomatosis. This is a necrotizing vasculitic phenomenon

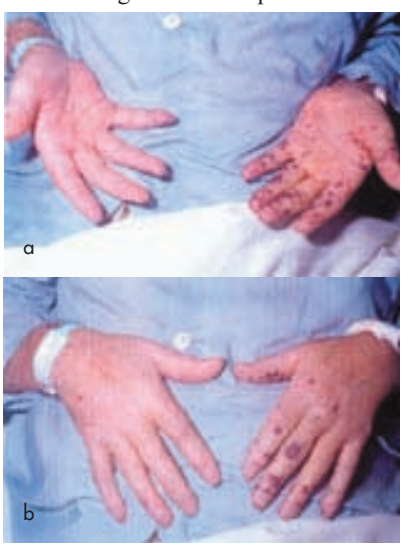

Figure 1. Purpuric rash predominantly on (a) palmar and (b) dorsal aspect of left hand.

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which varies from a mild indolent form to an aggressive, often fatal illness, and demonstrates a preponderance for the respiratory and renal systems such that the airway is almost always initially involved in the disease process (Hoffman et al, 1992).

Any organ system may become involved but the gastointestinal tract is normally spared. A series of post-mortem examinations, however, has revealed involvement of the gut (Walton, 1958) in nearly a quarter of cases of Wegener's. Perforation resulting from necrosis and peritonitis are the usual features and have been described in both small and large intestine without a predeliction for either. In all cases, the onset of gastrointestinal symptoms occurred within the first 2 years of the disease origin and all occurred in the presence of classical lung and renal involvement (McNabb et al, 1982).

The majority of reported cases describe perforation while the patient is taking immunosuppressive therapy for other sequelae of the condition. The immunosuppressive therapy itself has been shown to predispose to bowel perforation (Matolo et al, 1976). In the case described, small bowel ischaemia and perforation are the sole presenting features of Wegener's granulomatosis in a patient taking no medication before presentation.

Post-mortem examination demonstrates vasculitis in an artery within the affected small bowel (*Figure 2*). Granulomatous disease is evident in the lungs (*Figure 3*) and there is the classical renal appearance of necrosis of loops of capillary tufts (*Figure 4*).

The development of an acute abdomen as a result of Wegener's is a rare event in the course of the disease. This case exhibits such a manifestation as the sole presenting symptom, the patient having no symptoms, signs or radiological evidence of airways disease and a normal serum creatinine on admission. Severe intestinal involvement in Wegener's granulomatosis may therefore be regarded as a part of the disease process and not necessarily a consequence of therapy for it.

Interestingly Storesund et al (1998) have indicated histological evidence of vasculitis to be lacking in half the intestinal specimens resected for perforation. It may therefore be that immunosuppressive therapy plays some part in reduction of the classical histological appearance in such cases. The alternative remains that such therapy itself may be the sole reason for intestinal necrosis and perforation in an organ that is otherwise spared from the disease process. **HM**

Geraghty J, Mackay IR, Smith CD (1986)

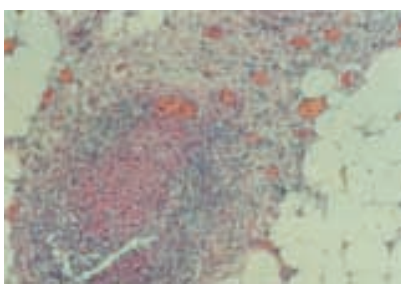

Figure 2. Acute and chronic inflammatory infiltrate in the wall of an artery in the submucosal wall of the small intestine (haemotoxylin and eosin stain x 4).

Intestinal perforation in Wegener's granulomatosis. *Gut* **27:** 450–1
Haworth SJ, Pusey CD (1984) Severe intestinal involvement in Wegener's granulomatosis. *Gut* **25:** 1296–300
Hoffman GS, Kerr GS, Leavitt RY et al (1992) Wegener's granulomatosis: an analysis of 158 patients. *Ann Intern Med* **116:** 488–98
Matolo NM, Garfinkle SE, Wolfman EF (1976) Intestinal necrosis and perforation in patients receiving immunosuppressive drugs. *Am J Surg* **132:** 753–4
McNabb WR, Lennox MS, Wedzicha JA (1982) Small intestine perforation in Wegener's granulomatosis. *Postgrad Med J* **58:** 123–5
Storesund B, Gran JT, Koldingnes W (1998) Severe intestinal involvement in Wegener's granulomatosis: report of two cases and review of the literature. *Br Soc Rheum* **37(4):** 387–90
Walton EW (1958) Giant cell granuloma of the respiratory tract (Wegener's granulomatosis). *Br Med J* **ii:** 265–70

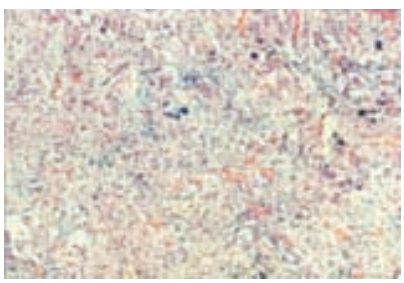

Figure 3. Section of lung demonstrating interstitial fibrosis with many scattered giant cells and focal disruption of the arterioles. There are no classical well-formed granulomas (haemotoxylin and eosin stain x 4).

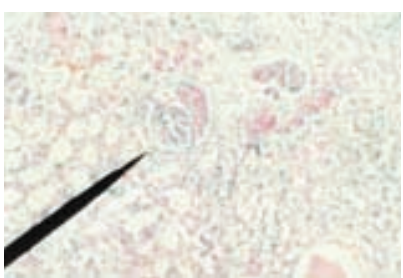

Figure 4. Focal segmental necrosing glomerulonephritis (haemotoxylin and eosin stain x 4).