

Constrictive pericarditis and intestinal haemorrhage due to Whipple's disease

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Summary

Two months after a pericardectomy for constrictive pericarditis a 37-year-old man presented with diarrhoea, abdominal pain and weight loss. During the course of investigation he developed brisk rectal bleeding and emergency angiography revealed a bleeding site related to an area of abnormal vasculature in the caecum. At laparotomy, the small bowel was found to be inflamed and the mesenteric lymph nodes enlarged. The overall histological appearances, confirmed later on endoscopic duodenal biopsies, were those of Whipple's disease. His symptoms resolved promptly after starting tetracycline therapy.

The clinical features of Whipple's disease are protean and often bizarre. We report here a case where the diagnosis was made after the onset of profuse rectal bleeding.

KEY WORDS: pericarditis, haemorrhage: gastrointestinal, Whipple's disease.

Case report

A 37-year-old man presented with breathlessness and ankle swelling for 2 months. Examination revealed peripheral oedema and an elevated jugular venous pressure. Echocardiography and cardiac catheterization confirmed the diagnosis of constrictive pericarditis. He underwent pericardectomy and recovered uneventfully.

Two months later he developed frequent loose stools, colicky lower abdominal pain, vomiting and weight loss. He had complained of flitting pains in the knees, ankles and hands for 5 years but these were not of sufficient severity for him to seek medical advice. On examination he was pale and pyrexial but there were no other abnormalities. Investigations revealed: haemoglobin 9.6 g/dl, white cell count

9.3×10^9 /litre, erythrocyte sedimentation rate (ESR) 19 mm/hr, platelet count and prothrombin time normal, alkaline phosphatase 319 i.u./litre (liver type), plasma proteins, bilirubin, transaminases normal, and cultures of blood, urine, sputum and stool were negative. Chest X-ray, gastroscopy, barium follow through, barium enema and ultrasound examination of the abdomen were normal. Colonoscopy was normal apart from some fresh blood in the caecum. A jejunal biopsy was attempted, but the patient was unable to swallow the Crosby capsule. He then developed torrential rectal bleeding, requiring 12 units of blood. Coeliac and superior mesenteric angiography (Dr M. I. Lavelle) revealed an abnormal vascular pattern in the caecum and contrast medium in the bowel lumen.

At laparotomy, the serosal surface of the small bowel was reddened and the mesenteric lymph nodes were enlarged. Lymph node biopsy revealed foreign body giant cells, an abundant chronic inflammatory cell infiltrate and macrophage granulomata. Macrophages contained PAS-positive granules, the overall appearances being those of Whipple's disease. No other surgical procedure was performed, but endoscopic biopsies obtained later from the distal duodenum confirmed the diagnosis. He was treated with tetracycline 250 mg, 4 times daily, with immediate resolution of symptoms and at follow up 12 months later he had gained 10 kg and had no further bleeding episodes.

Discussion

Whipple's disease is a multisystem disorder, but the principal manifestations are gastrointestinal. In a review of the literature between 1950 and 1969, Maizel, Ruffin and Dobbins (1970) were able to identify 114 cases to which they added a further 19. Eighty-eight per cent were male with an age range

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20–67 years. Weight loss (95%), diarrhoea (78%), arthralgia (65%) and abdominal pain (60%) were the most important symptoms, but neurological features were present in 8 patients. Anaemia (90%) and steatorrhoea (93%) were common while in 14 patients joint symptoms preceded diagnosis by more than 10 years. Gross gastrointestinal bleeding of undetermined site has been reported in only 2 patients (Gibb, Malpas and Parrish 1964; Smith *et al.*, 1965). In the present case, the onset of severe intestinal haemorrhage led to the diagnosis being made at laparotomy. It is also of interest that bleeding has not recurred since tetracycline therapy was commenced. In retrospect, the initial presentation with constrictive pericarditis may also have been due to Whipple's disease, since small quantities of PAS-positive material have now been identified in the pericardium. This in itself is unusual, since valvular lesions, congestive cardiac failure and non-specific electrocardiographic abnor-

malities are the commonest cardiac manifestations (Bostwick *et al.*, 1981) and pericarditis, when it does occur, is frequently subclinical and serofibrinous rather than constrictive in type (Kraunz, 1969).

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(Accepted 2 July 1982)