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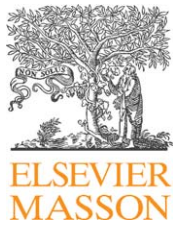

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CLINICAL CASE

Multiple intestinal perforation in a patient with Wegener's granulomatosis: A case report and review of the literature

Perforations intestinales multiples chez un patient atteint de granulomatose de Wegener : cas clinique et revue de littérature

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Summary Wegener's granulomatosis is a necrotizing vasculitis of unknown etiology characterized mainly by inflammation of the small- and medium-sized arteries and veins that affect any viscera. It may rarely involve the gastrointestinal tract. Only a few cases of multiple focus ileal perforation due to ulcers associated with Wegener's granulomatosis have been reported. Herein we report a case of a 32-year-old man with extensive intestinal small bowel ischaemic perforation due to Wegener's granulomatosis.

Introduction

Wegener's granulomatosis (WG) is a complex, immune-mediated disorder in which tissue injury results from the interplay of an initiating inflammatory event and a highly specific immune response. It often involves the upper

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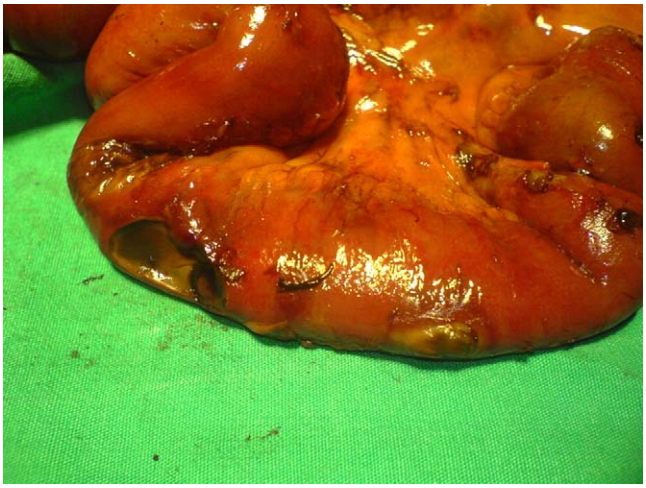

Figure 1 Multiple full thickness necrosis and perforation in the antimesenteric area of the ileum.

respiratory tract, lung and kidney but nevertheless the inflammatory destructive lesions may develop in the gastrointestinal tract [1,2]. During the course of WG, small and large bowel perforations are uncommon occurrences. Herein we present a case of WG with multiple intestinal small and large bowel perforation. We have also reviewed the pertinent literature regarding gastrointestinal tract perforation in cases with WG.

Case report

A 32-year-old man was admitted to the hospital with complaints of abdominal pain, melena and hematemesis. He had had a 2-week history of WG with serum antineutrophil cytoplasmic antibody (c-ANCA) positivity and multiple pulmonary cavities. Microscopic examination of biopsy specimens obtained from the lungs had shown necrotizing granulomatous vasculitis. He had been treated with high-dose steroid and cyclophosphamide. On admission, physical examination revealed increased bowel sounds. Laboratory findings were as follows: serum hemoglobin 9.2 g/dl, white cell count 12,000/mm^3 and platelet count 220,000/mm^3. Biochemical tests were within normal limits. Upper endoscopic examination demonstrated gastritis and red blood clots throughout the stomach but the source of the hemorrhage was unclear. In order to explore the bleeding site, computerized tomography (CT) angiography was performed but no bleeding origin was detected. Thereafter, he was followed-up in the emergency care unit in order to stabilize his hemodynamic state. On the 3rd day of hospitalization, nausea, vomiting and abdominal pain developed. Physical examination revealed abdominal distention and rebound tenderness in all quadrants. Complete blood count showed hemoglobin: 9.6 g/dl, white cell count: 22,500/mm^3, platelet count: 450,000/mm^3. Signs of peritonitis were present and his general condition deteriorated rapidly so that emergency surgery was performed.

During laparotomy there was inflammation of the entire small bowel and numerous necrotic perforated foci on ileal segments (Fig. 1). In the caecum and ascending colon there were also arterial circulatory problems. Therefore right hemicolectomy and ileal resection with jejunostomy were performed. Histopathological examination of the resected specimens showed inflammatory infiltration associated with necrotizing and granulomatous vasculitis of small-medium sized vessels, which was consistent with WG. In the postoperative course, he deteriorated and died as a result of sepsis and multiorgan failure.

Discussion

WG is characterized by necrotizing vasculitis and granulomatous inflammation. Therapy has dramatically improved by daily treatment with cyclophosphamide and prednisolone [3]. However, severe and potentially lethal adverse effects are associated with prolonged treatment with cyclophosphamide and prednisolone [4—6].

Intestinal involvement in WG is uncommon and usually detected in autopsy studies [7]. Clinical manifestations of severe intestinal disease have been frequently reported [1,2,8]. Storesund et al. evaluated six patients of WG who presented with severe intestinal involvement. Acute abdominal pain with signs of peritonitis constituted the main clinical picture in five of the six cases. Intestinal perforations (small and/or large bowel) developed in four of the six cases and perforations were the most common cause of emergency surgery. All of the patients with intestinal perforation were treated with prednisolone [9].

The potential link between corticosteroids and intestinal perforation is not clear in the course of WG. Intestinal perforation has been reported as a rare complication of immunosuppressive treatment in patients after renal transplantation [10]. It is suggested that immunosuppressive treatment, especially with corticosteroids, might play a role in development of intestinal perforation [11,12]. However, it is difficult to discriminate whether intestinal perforation was produced by the underlying vasculitis itself or by the administration of high-dose immunosuppressive agents in the present case. McNabb et al. reported a case of intestinal perforation in WG that occurred in the terminal ileum 5 days after treatment with corticosteroids [13]. On the other hand, Haworth and Pusey suggested that severe ileal, caecal, and rectal involvement improved rapidly after treatment with cyclophosphamide, azathioprine, and prednisolone [14].

Patients on systemic steroid therapy are at increased risk of diffuse peritonitis, especially from gastrointestinal perforation. Menegaux et al. suggested that early abdominal exploration should be performed in patients with persisting abdominal pain during the treatment with systemic steroid [15]. In addition, some authors believe that depletion of lymphoid tissue in the submucosa of the damaged intestinal segment is caused by steroid usage and that this depletion and thinning eventually caused the wall to perforate [16].

Reviewing the available medical literature, we evaluated seven cases of WG with intestinal perforation [2,9,13,17—20]. Six of them were males, and the age of the patients varied from 26 to 56. In almost all of the patients, the intestinal perforation occurred within the 1st year of the disease. As shown in Table 1, the most common location of perforation was the ileum. Immunosuppressive therapy was administered in six of the seven patients. Histological exam-