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CASE REPORT

Intestinal perforation as an early complication in Wegener's granulomatosis

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Abstract

We present the case of a young man with involvement of the gastrointestinal tract in the early phase of Wegener's granulomatosis. The patient presented at the emergency department with sudden onset of abdominal pain, nausea and vomiting. Radiography work up was negative for free air although ultrasound examination showed extraluminal intra-abdominal fluid. Exploratory laparotomy showed perforation of the jejunum. The bowel was vital except for this small segment of jejunum. A 5-cm long segment of jejunum was resected which revealed ulcerative inflammation accompanied by occluded arteries of the small intestine. Although intestinal perforation in Wegener's granulomatosis is uncommon, several cases have been previously reported. Intestinal involvement in the early phase of the disease is even more uncommon. This case combined with previously reported cases emphasizes the possibility of gastrointestinal manifestation early in Wegener's disease.

Key words: Wegener's granulomatosis; Intestinal tract; Perforation

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INTRODUCTION

Wegener's granulomatosis (WG) is a necrotizing vasculitis of the small to medium-sized arteries characterized by involvement of the upper and lower respiratory tract and the kidneys, although other sites can also be involved[1,2]. The etiology of WG remains elusive. However, previous studies have implicated interplay between genetic susceptibility and environmental triggers as possible risk factors[3,4]. The gastrointestinal tract is rarely involved, occurring in 10%-24% of patients with WG[1]. Only a few cases had histological confirmation of vasculitis at the site of perforation[5-7]. In this case report, we describe a case of WG with intestinal perforation in the early course of the disease.

CASE REPORT

A previously healthy 35 years old Caucasian man with arthralgia of the knees, elbows and hands, skin rash, sinusitis, oral ulcers and renal impairment was admitted to our hospital in June 2009.

Laboratory findings showed: hemoglobin 7.7 mmol/L, C-reactive protein 13.5 mg/L, leucocytes 18000/mm^3, serum creatinine 110 μmol/L and positive anti-neutrophil cytoplasmic antibodies proteinase 3. Urine sediment contained free red blood cells (142/h). Plain chest radiography was normal. Biopsy was performed on the skin lesions and

the microscopic findings showed leukocytoclastic vasculitis. Based on the laboratory and clinical findings, the patient was diagnosed with systemic vasculitis secondary to WG and intravenous prednisolone (60 mg/d, iv) combined with oral cyclophosphamide (200 mg/d) therapy was started.

Later in the disease course, the patient became respiratory insufficient and was intubated. A week after the diagnosis of WG, his clinical condition was determined by a persistent gastrointestinal hemorrhage for which he received a total of 150 units of packed cells. Gastrointestinal endoscopy was performed several times in order to treat the hemorrhage. However, except for ulcerative lesions in the gastrointestinal tract, no explanation for the hemorrhage was found. As the bleeding continued, angiography of the superior mesenteric artery was performed and arterial blood loss was seen in the proximal jejunum. Subsequently coils were successfully placed. In August 2009 the patient was discharged with continued oral medication.

In October 2009 the patient again presented with a sudden onset of severe abdominal pain, nausea and vomiting at the emergency department. On physical examination, his blood pressure was 138/80 mmHg with a pulse rate of 115 beats/min and temperature of 36.5℃. Abdominal examination revealed diminished bowel sounds with diffuse abdominal pain and signs of peritonitis. Laboratory analysis showed a C-reactive protein level of 16 mg/L, erythrocyte sedimentation rate of 17 mm/h and hemoglobin level of 7.0 mmol/L. X-ray analysis of the thorax was negative for abdominal free air although ultrasound examination of the abdomen revealed intra-abdominal extraluminal fluid. Exploratory laparotomy showed ischemia of the proximal jejunum and perforation at this site. Additionally, segmental resection of the jejunum was performed. The procedure could also have been performed laparoscopically.

Histopathological findings were central ulcerative inflammation and occluded small arteries in the intestinal wall. Giant cells were not found. Lamina elastica staining showed impaired arterial wall due to inflammatory processes. Immunohistochemical study for cytomegalovirus antigen was negative. The patient recovered without any additional adverse events and remains in remission on oral prednisolone and cyclophosphamide therapy.

DISCUSSION

WG is a necrotizing vasculitis defined by granulomatous changes of the upper and lower respiratory tract and is frequently associated with glomerulonephritis[8].

Our case meets the diagnostic criteria of WG published by Fauci *et al*[1] which concludes that in order to establish the diagnosis there should be clinical evidence of disease in two of the three principle sites (upper airways, lung and kidney) with histological confirmation in at least one site[1]. Our case shows severe intestinal involvement in an early phase of Wegener's disease. Whether the perforation of the jejunum was due to WG alone or the result of intestinal ischemia due to coiling through angiography remains uncertain.

In our case gastrointestinal hemorrhage developed in the early course of the disease and an endoscopic maneuver was unable to precisely detect the cause of the hemorrhage. The bleeding was managed with coiling side-branches of the superior mesenteric artery through angiography examination. The coiling might have caused ischemia of the intestinal segment resulting in perforation of the intestinal wall. Since endoscopic examination a month after coiling did not show any anomalies of the intestinal wall, ischemia due to coiling through angiography examination seems unlikely. Moreover our patient did not have any clinical symptoms of colonic ischemia[9] prior to the sudden onset of abdominal pain. In our patient, ischemia caused by vasculitis (characteristic for WG) seems to be the most logical explanation for the perforation of the intestinal wall.

Earlier case reports describe intestinal involvement of WG. Very few cases report perforation of the intestinal wall in the early course of the disease[6,10-12].

Use of immunosuppressive therapy has been suggested as an etiological factor for intestinal involvement in WG[6]. Intestinal involvement has however been described in a very early course of the disease in which no medical treatment was started. Also, in multiple cases, histological examination showed extensive vasculitis with fibrinoid necrosis resulting in intestinal perforation[5-10]. Although immunosuppressive therapy is not proven as an etiological factor, it might exacerbate already existing areas of ulceration leading to perforation.

In conclusion, even though uncommon, intestinal involvement may be listed in the clinical symptoms of WG and can occur in the early course of the disease. The diagnosis should be suspected in patients presenting with sudden abdominal pain and symptoms of systemic vasculitis. An early diagnosis is vital and should not be missed since it could result in severe complications and death.

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