Case report

Intestinal Perforation due to Vasculitis in a Patient with Wegener’s Granulomatosis

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Abstract

Wegener’s granulomatosis is a small vessel vasculitis that typically involves upper and lower airways and kidneys. In this report, we present a patient with severe gastrointestinal involvement that caused intestinal perforation. A review of the literature and discussion of the pathogenesis of this serious complication is also provided.

Keywords: Wegener’s granulomatosis, crescentic nephritis, intestinal perforation, small vessel vasculitis, acute renal failure

Introduction

Wegener’s granulomatosis is necrotizing small vessel vasculitis with multisystem involvement. The disease typically involves the upper and lower respiratory tracts and kidneys [1]. Gastrointestinal involvement is a very rare manifestation of this disease [2]. We present a Wegener’s granulomatosis patient with crescentic glomerulonephritis and severe gastrointestinal involvement with multiple intestinal perforations.

Case report

A 37 years old man had been followed up with the diagnosis Wegener’s granulomatosis in another hospital for three months. He had been presented with rhinorea, epistaxis, oral ulcers, and lower extremity pain and skin ulcers located in ankles. His C-ANCA test was positive. One month before he started to complain from nausea, vomiting and abdominal pain, his creatinine level was found to be 6.2 mg/dl and a renal biopsy revealed mesangial proliferation. He was then referred to our hospital for further evaluation and management. On admission he was complaining of fatigue, nausea, abdominal, and upper and lower extremity pain. His blood pressure was 130/70 mmHg and heart rate was 92/min. There were multiple purpuric lesions in his lower extremities. On abdominal palpation there were tenderness all over the abdomen and liver was palpable three cm under the costal line. Physical examination of respiratory and cardiovascular systems was unremarkable. In laboratory examination hematocrite was 22%, white blood cells 11.400/mm³, platelets 448000/mm³, erythrocyte sedimentation rate 65mm/h, urea 198, creatinine 7.9mg/dl and albumin 2.4g/dl. C-ANCA test was positive; P-ANCA and Anti nuclear anticores were negative. Chest X-ray was normal. A nasal endoscopic examination was performed and biopsy was taken from some suspected lesions. Pathological examination revealed widespread chronic inflammatory cells infiltration in submucosal area and fibrinoid necrosis in small vessels.

Hemodialysis was started and a second renal biopsy was performed. Pathological examination revealed necrotizing crescentic glomerulonephritis; nine glomerulus were seen in the light microscopy, in six of them there were cellular crescent formation and in the remaining three there were fibro-cellular crescents formation (Figure 1). Immunofluorescent examination did not revealed immune complex deposits. Cyclophosphamide 1g was given and steroid 1g/every other day was stared. On the 4th day of the therapy his general condition deteriorated rapidly, severe abdominal pain and symptoms of peritonitis developed. An emergency laparotomy was performed. There were three-perforation areas in 40, 70 and 110cm below the ilocheclar valve. A 70 cm resection with an ilostomy was performed. However the patient diet due to septic shock, in the first postoperative day in the intensive care unit.

Histological examination of the resection material revealed, vasculitic involvement in submucosal small and middle sized vessels with multiple fibrin thromi (Figure 2).
Discussion

In this case Wegener’s granulomatosis was diagnosed on the basis of skin, upper respiratory tract and kidney involvement, positive C-ANCA and biopsies consistent with small vessel vasculitis. According to autopsy findings gastrointestinal involvement was seen in 24% of the Wegener’s granulomatosis patients [3]. However, clinically apparent gastrointestinal involvement in Wegener’s granulomatosis is a rare event [4]. Cases reports of gastrointestinal involvement in Wegener’s granulomatosis have described cases of enterocolitis, gastrointestinal hemorrhage, intestinal perforation, esophageal involvement, and recurrent pancreatitis [5]. Only a handful of cases with intestinal perforation has been presented so far [2,6-12]. This complication generally occurs in patients with an active disease [2] however to the best of our knowledge at least one case of intestinal perforation was reported in a patient in whom the course of disease was indolent [6].
The use of immunosuppressive therapy has been incriminated as an etiological factor in intestinal necrosis and perforation [12]. The fact that in some cases of perforation no vasculitic involvement was seen in the histological examination favors this hypothesis [6,9]. In our case abdominal symptoms were present before the start of the immunosuppressive therapy and symptoms of peritonitis developed following the start of the immunosuppressive therapy. A possible explanation might be that immunosuppressive agents contribute to the development of perforation in intestinal wall that was weakened by vasculitic involvement [6,9]. On the other hand, the temporal association between the start of the therapy and the development of peritonitis can be a confounder reflecting a more active disease [2]. Indeed, similar to our case, a very active intestinal involvement has been determined in histological examination, in some other cases too. [13]. In a previously untreated patient intestinal perforation was the presenting symptom of Wegener’s granulomatosis [10] and in some cases immunosuppressive drugs were used without any major hazard during the post perforation period [11]. Moreover standard immunosuppressive therapy resulted in resolution in other forms of gastrointestinal involvement of Wegener’s granulomatosis [5]. These observations might suggest that intestinal perforation and necrosis should be regarded as complications of Wegener’s granulomatosis itself rather than as complications induced by its coherent medical therapy [2]. In summary the exact pathogenesis of intestinal perforation in Wegener’s granulomatosis is not fully understood. There is a high incidence of laparotomy and death associated with reported cases of intestinal involvement [2,6-12]. We hope that this case report will point out this rare but serious complication. Early surgical intervention and appropriate immunosuppressive therapy can be life saving.

Conflict of interest statement. None declared.

References