Wegener’s granulomatosis—an unusual case of colonic haemorrhage

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Case report

A 45-year-old lady presented to her local hospital with a 7-day history of flu like illness, sore throat, myalgia and bilateral polyarthralgia following a period with rhinitis and epistaxis. She had a vasculitic rash on her arms, legs, scalp and oral mucosa. She was tachypnoeic with low oxygen saturations. She had radiological changes in keeping with pulmonary haemorrhage (Figure 1). She was diagnosed with Wegener’s granulomatosis (WG) with a c-ANCA (antineutrophil cytoplasmic autoantibodies) titre of 1 in 320. She was treated with invasive ventilation, pulsed intravenous (IV) methylprednisolone (500 mg) and plasma exchange. She was further commenced on IV cyclophosphamide (5 mg/kg body weight every 21 days).

On Day 10 post presentation she began to have significant fresh per rectal bleeding. She underwent multiple transfusions and bidirectional endoscopies. Gastroscopy did not show any abnormal findings, whereas, limited colonoscopy revealed an oedematous and inflamed mucosa with sigmoid and rectal sparing. Colonic biopsies performed at the time showed generalized vasculitis (Figures 2 and 3). Due to deterioration in renal function she was started on haemofiltration. She continued to have significant bleeding and was requiring large quantities of blood products. She was transfused 92 units of packed red cells over the course of 10 days. A mesenteric angiogram failed to reveal a specific bleeding point and a CT abdomen revealed bowel wall oedema. She continued to have lower gastrointestinal bleeding and underwent a subtotal colectomy and an ileostomy formation on Day 18 post presentation. A colonoscopy done on the table revealed diffuse mucosal haemorrhage. Postoperatively, her weaning period was complicated by bleed from ulcers in the ileum which were treated endoscopically. She recovered her renal function on Day 26 post presentation. The patient was stepped down from ITU to a HDU 40 days post presentation. After a prolonged period of rehabilitation she was discharged with a normal renal function, a well functioning ileostomy and recovering respiratory function.

Discussion

WG is an idiopathic small-vessel vasculitis strongly associated with antineutrophil cytoplasmic autoantibodies (ANCA). Constitutional symptoms such as fever, myalgia, arthralgia and malaise are common. In over 90% of patients there is upper or lower respiratory tract involvement or both. Alveolar capillaritis can cause life-threatening massive pulmonary haemorrhage. About 80% of patients classically develop a pauci-immune necrotizing glomerulonephritis. A high incidence of venous thrombotic events exists. Gastrointestinal disease however is uncommon.

This patient initially presented with clinical features highly consistent with WG and in conjunction with the raised c-ANCA titre, WG was promptly...
diagnosed and immunosuppressive therapy commenced. Initial therapy with cyclophosphamide and corticosteroids induces improvement in 91% of patients, with 75% achieving complete remission. We opted for pulsed IV cyclophosphamide that induces remission at a similar rate to daily oral cyclophosphamide, while having the advantage of a lower cumulative cyclophosphamide dose and a lower rate of leucopenia. Plasma exchange was initiated in addition to the induction immunosuppressive therapy. A retrospective review by Klemmer et al., suggested plasma exchange is beneficial in patients with small-vessel vasculitis and pulmonary haemorrhage, with no complications of the therapy being reported.

Unusually for WG, this patient had severe colonic haemorrhage with significant impact on her morbidity, leading to a protracted ITU stay. Intestinal disease in WG is rare. In one series of 45 patients only 10% had gastrointestinal symptoms such as abdominal pain, diarrhoea and blood loss at presentation or during relapse. In another case series of 87 patients, no gastrointestinal symptoms were reported. In a case series of 6 patients with severe intestinal involvement, both the small and large bowels were affected, with vasculitis, ischaemia, inflammation and ulceration being the main histological findings. There are few reports of massive intestinal haemorrhage in WG. Chow et al. report a case of massive gastrointestinal bleeding of jejunal origin necessitating surgical resection. In addition to conventional immunosuppressive therapy, intravenous immunoglobulin was administered which has been shown to reduce disease activity in persistent vasculitis. Deger et al. report a case of massive gastrointestinal bleeding due to jejunal and colonic involvement requiring surgery and angiographic embolisation. Arhan et al. report a case of severe haemorrhage secondary to duodenal ulceration. In all cases, gastrointestinal bleeding occurred after immunosuppressive treatment. It is thought however, that the severe intestinal manifestations are associated with the disease process rather than related to the immunosuppressive therapy. There is yet to be a randomized controlled trial for the treatment of gastrointestinal involvement in WG.

WG principally affects the upper airways, lungs and kidney. Gastrointestinal manifestations, in particular massive haemorrhage, are rare. Rapid diagnosis, early immunosuppressive therapy and appropriate surgical intervention can be lifesaving.

Conflict of interest: None declared.
References


