CASE REPORT

Colitis: an unusual presentation of Wegener's granulomatosis

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SUMMARY

Wegener's granulomatosis (WG) also known as granulomatosis with polyangiitis (GPA) is an anti-neutrophil cytoplasmic antibody-positive (ANCA) vasculitis which most commonly affects the upper respiratory tract, lungs and kidneys. It is uncommon for colitis to be the primary reason for the first hospital admission related to WG. This case represents one of the few in the literature where colitis is associated with WG and in particular, where colonic involvement was the presenting symptom. The patient presented to hospital with a 3-day history of bloody diarrhoea and was treated for colitis. The disease progressed and during the second admission renal and pulmonary involvement was found. A renal biopsy showed a crescentic change and a CT-confirmed inflammatory changes in the caecum and ascending colon. A diagnosis of WG was made and appropriate treatment initiated. The patient is now in remission.

BACKGROUND

Wegener's granulomatosis (WG) is an anti-neutrophil cytoplasmic antibody (ANCA)-positive vasculitis affecting small- to medium-sized vessels. It is characterised by necrotising granulomas and most commonly affects respiratory tract and kidneys. Postmortem studies have displayed gastrointestinal involvement¹ but colitis as a clinical feature is uncommon. This case represents one of the few where colitis is the primary reason for initial presentation to a hospital.

CASE PRESENTATION

A 29-year-old man presented to the accident and emergency department with a 2-day history of bloody diarrhoea. The blood was red and mixed with the stool. The diarrhoea was watery in nature, occurred 10–15 times a day and contained mucus. At the time of admission the patient had been experiencing symptoms of nasal congestion and lethargy for 2 months. There was no history of gastrointestinal problems and no family history.

There was no shortness of breath, cough or haemoptysis and clinical examination of the respiratory system was unremarkable.

Colitis caused by infection or inflammatory bowel disease was suspected and a series of blood tests were ordered. They showed a slightly raised white cell count ($15.24 \times 10^9/l$) with raised neutrophils ($11.16 \times 10^9/l$) and monocytes ($2.00 \times 10^9/l$). He also had a microcytic anaemia (mean corpuscular volume (MCV) 60.3 fl) (haemoglobin 8.7 g/dl) and raised inflammatory markers (C reactive

protein 301 mg/l, erythrocyte sedimentation rate 109 mm/hour). The urea was within the normal range (urea 5.1 mmol/l), but creatinine was slightly high (creatinine 103 μ mol/l). Stool microscopy and cultures were negative (see Investigations for details).

A cytoplasmic ANCA (cANCA) test was positive. Flexible sigmoidoscopy showed some patchy mild erythema within the rectum but the mucosa in the sigmoid was not obviously inflamed. Biopsies showed no evidence of granuloma formation. No chest x-ray was performed during this initial admission. The patient was started on oral steroids owing to suspected inflammatory bowel disease and the symptoms improved. He was discharged with a gastroenterology follow-up arranged.

Two weeks later the patient developed diffuse abdominal pain with bloody diarrhoea very similar in nature to the previous episode. The patient also had fever, arthralgia and dyspnoea. A CT of the abdomen was performed which showed inflammatory changes in the caecum and ascending colon. At this admission there was also obvious renal involvement with a markedly raised creatinine (388 μ mol/l). Proteinase-3 (PR3) antibodies were high (24 kU/l). The lower respiratory tract was also involved. A chest x-ray showed bilateral costophrenic blunting with some indication of mild pulmonary congestion. A CT scan significantly demonstrated a ground glass appearance in the lung bases.

A renal biopsy showed the presence of a crescentic glomerulonephritis (figure 1) which was consistent with a diagnosis of WG. These results combined with a more detailed history which elicited the fact the patient had experienced epistaxis twice in the last month led to the diagnosis of WG.

The patient was admitted to the renal ward where he received plasma exchange therapy, corticosteroids and pulsed intravenous cyclophosphamide. After 10 days in the ward he was able to go home with him coming back for a last dose of cyclophosphamide.

INVESTIGATIONS**Serology**

cANCA was positive. Proteinase-3 antibodies 24 kU/l. Stool microscopy and cultures: Negative for cryptosporidium, *Escherichia coli* O157, *Campylobacter*, *Salmonella*, *Shigella* and *Clostridium difficile*. Sigmoidoscopy: Small amount of inflammation observed in rectum. No obvious abnormality seen in the sigmoid colon. Rectal biopsy: no granuloma or inflammatory change.

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Figure 1 The arrow highlights a segmental necrotising lesion. The adjacent glomerulus is relatively normal.

CT scan

Inflammatory change in caecum and ascending colon. Ground glass appearance of lung bases was observed. A renal biopsy was performed and the results are shown in figure 1. A segmental necrotising lesion with a crescentic formation is shown. The majority of glomeruli seen on biopsy showed this histological pattern. This is consistent with the expected findings in WG with renal involvement.

OUTCOME AND FOLLOW-UP

Once a diagnosis of WG was made upon the second admission, appropriate treatment with corticosteroids was initiated immediately. Owing to the extent of the renal involvement by this time, it was decided that plasma exchange was also appropriate. The patient received pulsed intravenous cyclophosphamide and made a good recovery.

He was then sent home with oral medications and outpatient follow-up. He is currently in remission.

DISCUSSION

WG is a multi-system disease most commonly affecting the upper respiratory tract, kidneys and lungs. A number of studies and case reports have shown that WG has a wide variety of presenting symptoms covering multiple systems.^{2–4} Colitis is a relatively uncommon sign and it is particularly uncommon for colitis to be the primary reason for the first hospital admission. There are a number of cases reported where colitis had been a significant feature of the disease^{5–8} many years after initial diagnosis. However, there have been very few cases reported where colitis seemed to be the only presenting feature.^{9–11} In this case, the patient had experienced sinus congestion and two episodes of epistaxis earlier but colitis was the reason for the first presentation to hospital. Owing to the absence of an infective cause and the temporal nature of the renal and pulmonary involvement leading to diagnosis, this case is likely to represent one of the few where colitis was the primary reason for presentation.

At the second admission the diagnosis of WG was much easier to make because there was renal involvement. It is unfortunate in terms of this case report and scientific curiosity that a complete colonoscopy was not performed. It would have been significant to see the character of the mucosa and examine the histology in the right colon which appeared inflamed on CT.

However, it may be the case that histological findings would have been non-specific.¹²

Steroid and cyclophosphamide therapy was started following the BSR guidelines¹³ and the patient's renal function and symptoms improved. The patient is now in remission.

WG potentially has very serious complications and thus a diagnosis should not be delayed. There should be a high level of suspicion in case of patients with colitis where the cause is unclear and cANCA plus PR3 tests should be performed. A urine dip at admission may have also been useful.

The primary interest in this case is the unusual presentation. WG should be considered in unusual cases of colitis with a positive cANCA. A history of sinusitis or renal involvement should certainly add to suspicion.

Learning points

- ▶ Wegener's granulomatosis (WG) should be considered as a cause for colitis in unusual cases or cases where there are other signs and symptoms such as sinusitis, haemoptysis or impaired renal function.
- ▶ A positive antineutrophil cytoplasmic antibodies (cANCA) in a case of colitis may indicate WG and a PR3 screen should be undertaken, a urine dip be performed and a chest x-ray may be considered.
- ▶ Early diagnosis of WG when it presents before renal involvement may help patients avoid more invasive procedures and treatments.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Wegener F. Über eine eigenartige rinogene granulomatose mit besonderer, beteiligung des arteriensystems und der Nieren. *Beitr Path Anat Pathol* 1939;102:36–68.
- 2 Fauci AS, Haynes BF, Katz P, et al. WG: prospective clinical and therapeutic experience 85 patients for 21 years. *Ann Intern Med* 1983;98:76–85.
- 3 Anderson G, Coles ET, Crane M, et al. WG granuloma. A series of 265 British cases seen between 1975 and 1985. A report by a sub-committee of the British Thoracic Society Research Committee. *Q J Med* 1992;83:427–38.
- 4 Hoffman GS, Kerr GS, Leavitt RY, et al. WG: an analysis of 158 patients. *Ann Intern Med* 1992;116:488–98.
- 5 Deniz K, Ozseker HS, Balas S, et al. Intestinal involvement in Wegener's granulomatosis. *J Gastrointest Liver Dis* 2007;16:329–31.
- 6 Akca T, Colak T, Caglikuleki M. Intestinal perforation in Wegener's granulomatosis. *Turk J Trauma Emerg Surg* 2005;11:348–51.
- 7 Shaikh F, Sabu C, Peirce T, et al. Extensive intestinal ischaemic necrosis in Wegener's granulomatosis. *Gut* 55:1368–9.
- 8 Storesund B, Gran JT, Koldingsnes W. Severe intestinal involvement in Wegener's granulomatosis: report of two cases and review of the literature. *Br J Rheumatol* 1998;37:387–90.
- 9 Schneider A, Menzel J, Gaubitz M, et al. *J Intern Med* 1997;242:513–17.
- 10 Steel C, Bohra S, Broe P, et al. Acute upper gastrointestinal haemorrhage and colitis: an unusual presentation of Wegener's granulomatosis. *Eur J Gastroenterol Hepatol* 2001;13:993–5.
- 11 Qian Q, Cornell L, Chandran V, et al. Haemorrhagic colitis as a presenting feature of Wegener granulomatosis. *J Gastrointest Liver Dis* 2010;19:445–7.
- 12 Muller-Lander U. Vasculitides of the gastrointestinal tract. *Best Pract Res Clin Gastroenterol* 2001;15:59–82.
- 13 Lapraik C, Watts R, Bacon P, et al. BSR and BHPR guidelines for the management of adults with ANCA associated vasculitis. *Rheumatology* 2007;46:1–11.

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