Case Report

ANCA-associated vasculitis complicated by haemoperitoneum

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Abstract

We present a case of myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA)-associated vasculitis with the rare complication of haemoperitoneum due to mesenteric vessel involvement. Initial investigations demonstrated typical renal features including haematoproteinuria and a focal segmental necrotising glomerulonephritis. The patient then developed haemoperitoneum and required an emergency hemicolecotomy. Subsequent histology revealed ileo-colic arteritis with aneurysm formation and rupture. A sustained remission was achieved with corticosteroids and mycophenolate mofetil followed by azathioprine. This case demonstrates the need for a high degree of vigilance in patients with ANCA-associated vasculitis who develop abdominal symptoms.

Keywords: ANCA; haemoperitoneum; myeloperoxidase; vasculitis

Background

Myeloperoxidase-antineutrophil cytoplasmic antibody (MPO-ANCA)-associated vasculitis typically presents as a small-vessel vasculitis, with complications due to coexisting vasculitis in larger vessel types being infrequently reported. This report highlights the rare yet important complication of haemoperitoneum caused by vasculitis, aneurysm formation and rupture of a medium-sized splanchnic artery and describes the successful management of this case.

Case report

A 74-year-old lady, with a history of locally metastatic malignant melanoma affecting her left leg, presented initially with frontal headaches, vomiting and photophobia. Physical examination and biochemistry tests were unremarkable. A computerized tomography (CT) head scan showed a 5-mm Sylvian aneurysm of the left middle cerebral artery with no evidence of bleeding. She refused further investigations to exclude subarachnoid haemorrhage including magnetic resonance imaging and lumbar puncture. Her symptoms improved, and she was discharged from hospital.

A month later, she presented with new symptoms of jaw claudication, scalp tenderness, acheing of the proximal upper limbs, generalized fatigue and anorexia. Differential diagnoses of giant-cell arteritis and polymyalgia rheumatica were considered. Investigations showed a marked inflammatory response, with an erythrocyte sedimentation rate (ESR) of 108 mm/h and C-reactive protein (CRP) of >200 mg/l. Other test results were as follows: total white blood cell count 14.6 × 10⁹/l, serum creatinine 150 μmol/l, alanine aminotransferase (ALT) 100 IU/l and alkaline phosphatase (ALP) 507 IU/l. Urine dipstick revealed 2+ protein and 3+ blood, and urine protein-creatinine index was markedly raised at 2054 (normal <130). Urgent immunology tests revealed a positive perinuclear-antineutrophil cytoplasmic antibody (p-ANCA) with an anti-myeloperoxidase (MPO) titre of >100 U/ml. Anti-proteinase-3, anti-glomerular basement membrane antibody, antinuclear antibody, rheumatoid factor and C3 and C4 complement levels were all within normal limits.

An urgent renal biopsy showed a pauci-immune focal segmental necrotising glomerulonephritis with severe vasculitis and fibrinoid necrosis affecting the small arteries and arterioles, consistent with a diagnosis of MPO-ANCA-associated vasculitis. She was treated with intravenous pulses of methylprednisolone (500 mg daily for 3 days). Her symptoms of jaw claudication, scalp tenderness, anorexia and fatigue improved immediately. Serum creatinine started to decline from a peak value of 174 μmol/l.

Three days later, she complained of severe sudden-onset right upper quadrant abdominal and back pain. She became haemodynamically compromised, and haemoglobin fell to 92 g/l. An abdominal CT scan revealed a solitary bleeding aneurysm of the ileo-colic artery with a large intraperitoneal haematoma (Figure 1). She underwent an emergency right-sided hemicolecotomy. Histological examination showed a ruptured ileo-colic artery aneurysm, with associated patchy segmental necrotising vasculitis (Figure 2).

The patient made an uncomplicated postoperative recovery. After discussion with her dermatologist, a decision...
Fig. 1. Abdominal CT scan showing haemorrhage and the ileo-colic artery aneurysm.

Fig. 2. Section demonstrating ileo-colic artery vasculitis, with segmental necrosis and intramural haemorrhage (double arrow). Single arrow denotes unaffected artery.
was made to commence mycophenolate mofetil with a reducing dose regimen of oral prednisolone. Her MPO titre normalized within 10 weeks, and serum creatinine returned to normal limits. She was converted to azathioprine after 6 months and maintained on 5 mg prednisolone daily and has had no further relapses of her vasculitis.

Discussion

We report an unusual case of MPO-ANCA-associated vasculitis with histologically proven involvement of small- and medium-sized arteries. The initial clinical presentation was dominated by symptoms associated with large-sized blood vessel involvement that responded promptly to steroid therapy, raising the possibility of giant cell arteritis [1]. The diagnosis of MPO-ANCA-associated vasculitis was based on the presence of typical features including haematoproteinuria, positive p-ANCA and MPO antibodies and biopsy findings of a focal segmental necrotising glomerulonephritis and vasculitis with fibrinoid necrosis affecting small arteries and arterioles. These findings were important in selecting an appropriate immunosuppressive regimen. Her symptoms of jaw claudication and scalp tenderness improved immediately after corticosteroid therapy leading us to speculate whether large-sized vessels were involved in this case. A temporal artery biopsy was not performed.

MPO-ANCA-associated vasculitis mainly affects small blood vessels (i.e. capillaries, venules or arterioles), although medium-sized vessels may be involved. Gastrointestinal involvement is usually mild but may involve abdominal pain and bleeding into the gastrointestinal tract, often as a result of bowel ischaemia [1]. Extensive colonic ulceration and haemorrhage have also been reported [2].

Haemoperitoneum associated with MPO-ANCA-associated vasculitis is a rare complication which has been infrequently reported [3–5]. Ravanan et al. describe a patient with typical renal involvement, who subsequently developed multiple episodes of severe intra-abdominal haemorrhage secondary to splanchnic vasculitis despite apparent response of the renal vasculitis to conventional immunosuppression with corticosteroids and cyclophosphamide [3].Histological examination of the splanchnic blood vessels showed a transmural arteritis with intramural haemorrhage. Yahata et al. also describe a case of fatal haemoperitoneum due to rupture of the left gastric artery associated with MPO-ANCA-associated vasculitis [4].

Inflammation leading to arterial aneurysm formation is an uncommon feature of MPO-ANCA-associated vasculitis but is seen more often in other vasculitides, for example affecting 50–60% of cases of polyarteritis nodosa (PAN). In our case, the positive ANCA result and finding of a necrotising crescentic glomerulonephritis were more consistent with a diagnosis of an ANCA-associated vasculitis than PAN [6]. Aneurysm formation is also described in hepatitis virus-associated PAN [7]. However, in the aftermath of a prompt and favourable response to immunosuppression, we did not establish her hepatitis status. Aneurysm formation within mesenteric vessels is described as a rare complication of Behçet’s disease and Wegener’s granulomatosis [8,9].

In conclusion, medium-sized vessel involvement in MPO-ANCA-associated vasculitis causing mesenteric vessel aneurysm formation and haemoperitoneum is a rare but increasingly recognized phenomenon. This case demonstrates the importance of a high degree of vigilance in patients with MPO-ANCA-associated vasculitis who develop abdominal symptoms.

Conflict of interest statement. None declared.

References


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