Microscopic polyangiitis presenting with bilateral spontaneous perinephric haematoma

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DESCRIPTION

A 50-year-old female patient presented to the emergency room with intermittent fever and bilateral upper/lower limb weakness for 2 weeks and intermittent abdominal pain for 1 week. On general examination there were features of hypotension, tachypnoea and pallor. On nervous system examination there was bilateral foot drop, with decreased power of 3 out of 5 in bilateral upper/lower limbs with absent deep tendon reflex. On respiratory examination there was crepitation in bilateral lower lobes. The rest of the systemic examination was unremarkable.

Routine blood investigations revealed haemoglobin of 50 g/L, creatinine of 1.9 mg/dL, total count of $21.93 \times 10^9/L$ and erythrocyte sedimentation rate of 150 mm/hour. Due to low haemoglobin and hypotension, the patient underwent plain CT of the abdomen, which revealed left perinephric haematoma (figure 1A). After 48 hours the patient underwent contrast-enhanced CT of the abdomen, which revealed bilateral perinephric haematoma compressing the kidneys and multiple aneurysms of the branch vessels of bilateral renal arteries and hepatic artery (figure 1B–F). The patient developed haemoptysis on day 15 of admission. High-resolution CT of the chest revealed alveolar haemorrhage (figure 2A,B). Based on the imaging features, the possibility of a vasculitic cause was raised. Nerve biopsy showed features of vasculitis of medium-sized vessels and axonopathy. The patient had elevated levels of MPO ANCA (Myeloperoxidase Anti-Neutrophil Cytoplasmic Antibody). A final diagnosis of microscopic polyangiitis (MPA) was made based on imaging and laboratory parameters. The patient underwent bilateral percutaneous drainage of haematoma and was initiated on methylprednisolone and cyclophosphamide.

MPA is a rare vasculitis of small-sized vessels with or without involvement of medium-sized vessels and has predilection for capillaries, venules and arterioles.^{1,2} The incidence of MPA ranges between 1 and 9.4 per 100 000 individuals per year and is the most common ANCA-associated vasculitis.^{3,4} MPA most commonly presents with renal involvement (90%), followed by pulmonary involvement (22%).⁵ The most common renal manifestation is Pauci-immune-type, rapidly progressive glomerulonephritis, while alveolar haemorrhage due to alveolar capillaritis is a common pulmonary manifestation.^{5,6} Involvement of medium-sized renal vessels causing formation of microaneurysm and haemorrhage is more common in Polyarteritis Nodosa (PAN).⁷ Hence spontaneous perinephric haematoma formation in patients with

Figure 1 (A) Axial section of plain CT of the abdomen done on admission demonstrates left perinephric haematoma (white arrow). (B) Axial section of postcontrast CT after 48 hours demonstrates bilateral perinephric haematoma (arrows). (C) Axial postcontrast CT on arterial phase demonstrates microaneurysm in the right kidney (white arrow). (D) Delayed image demonstrates no significant change in the density of bilateral perinephric haematoma. (E and F) Coronal Maximum Intensity Projection (MIP) images demonstrate multiple microaneurysms in the liver and bilateral kidneys (arrows).

PAN is not uncommon, but rare in patients with MPA.⁸

According to Zhang *et al*⁹ the most common cause of renal haematoma is neoplasm (61%),

Figure 2 (A and B) Axial section of high-resolution CT of the chest demonstrates few foci of ground glass opacities in bilateral upper lobes (white arrows).

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Images in...

followed by vascular cause (17%). Among vascular causes, PAN is the most common. All patients with bilateral perinephric haematoma had PAN. Other causes of spontaneous perinephric haematoma are renal vein thrombosis, polycystic kidney disease and haemodialysis. Spontaneous haemorrhage due to anticoagulation therapy can occur in patients on haemodialysis.^{9 10}

Pulmonary involvement is commonly seen in MPA and can present as fleeting focal infiltrates to massive lung haemorrhage.¹ According to Ando *et al*¹¹ the most common chest CT findings in patients with MPA are ground glass opacities (94%), consolidation (78%), thickening of bronchovascular bundle (51%) and honeycombing (37%). Occasionally patients with MPA with chronic alveolar haemorrhage may experience pulmonary fibrosis and obstructive bronchitis.⁵ Contrast-enhanced CT can help differentiate the various causes of perinephric haematoma, while angiography can be used to evaluate vascular causes. However, to differentiate between PAN and MPA, biopsy and serum markers are required.

Learning points

- ▶ Spontaneous perinephric haematomas are commonly seen in PAN.
- ▶ Microscopic polyangiitis (MPA) can present with renal and pulmonary involvement; Rapidly Progressive Glomerulonephritis (RPGN) and alveolar haemorrhage are common renal and pulmonary manifestations.
- ▶ Common CT chest findings in patients with MPA are ground glass opacities, consolidation, thickening of bronchovascular bundle and honeycombing.

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