Unusual presentation of more common disease/injury

Polyarteritis nodosa presenting as haematuria following strenuous exercise

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Summary

Polyarteritis nodosa (PAN) can affect many organ systems but usually presents with a systemic illness characterised by malaise, weight loss, myalgia, arthralgia and signs of end-organ damage. Here, we report a case of PAN that presented acutely in a previously well 46-year-old man with visible haematuria and loin pain coinciding with an episode of strenuous exercise. Initially, the patient was thought to have suffered renal trauma, but subsequent investigations revealed intrarenal aneurysms typical of PAN which responded to immunosuppressive therapy. This case illustrates the importance of appropriate imaging for suspected urogenital tract trauma, that a high index of suspicion is required to make the diagnosis and that PAN can present with renal tract bleeding in the absence of obvious systemic features.

BACKGROUND

Polyarteritis nodosa (PAN) is a multisystem disease that presents in a many different ways affecting numerous organ systems. We encountered a case whereby a patient presented with PAN for the first time with haematuria following strenuous exercise. This is a previously undescribed presentation of this interesting disease, of which there are few, if any, examples in current literature.

CASE PRESENTATION

A fit-and-well 46-year-old man was admitted to hospital after passing visible haematuria with clots. The haematuria was first noticed by the patient while voiding immediately after completing a 6-mile hill race. During the race, the patient had slipped and slid down a hill on his left-hand side. Such 'controlled slides' occur regularly to participants in such events and the patient, who was an experienced fell runner, remarked that this particular incident was very much the norm for him in such a race. On presentation to the emergency department, the patient complained of severe left-flank pain and was passing frank haematuria with clots. Systemic enquiry revealed only a non-productive cough for the preceding few days. He had no medical history except for a tonsillo-adenoidectomy as a child. He took no regular medications and had no known drug allergies. He was a non-smoker with minimal alcohol consumption. The patient's initial observations were normal other than a respiratory rate of 24 and temperature of 34.4°C; both of these factors normalised spontaneously over 15 min. The only persistent positive examination finding was tenderness in the left flank of the abdomen.

INVESTIGATIONS

Admission bloods revealed a haemoglobin of 148 g/l, platelet count of 207×10^9 , white cell count of 17.8×10^9 (neutrophils 14.8×10^9), and normal routine biochemistry with the exception of a raised bilirubin of 55 $\mu\text{mol/l}$

(which later proved to be an incidental presentation of Gilbert's syndrome). In view of the frank haematuria and history of suspected trauma of the urogenital tract, a contrast-enhanced CT scan of the abdomen and pelvis was performed. The CT scan (figure 1A) revealed left ureteric obstruction secondary to a ureteric clot and an oedematous left kidney with a dilated renal pelvis but no obvious renal haematoma, laceration, vascular injury or incidental tumour such as an angiomyolipoma (which may bleed on minor trauma). The patient was initially treated with morphine for pain as well as intravenous fluids. He was started on oral ciprofloxacin 500 mg twice daily as his temperature rose to 38°C.

Further imaging in the form of a renal-tract ultrasound scan was performed 2 days following admission to reassess the left-renal collecting system. This confirmed persisting, moderate, left-sided, upper pole, pelvi-calyceal distension and peri-nephric oedema. In light of these findings, continuing pyrexia and ongoing flank pain, a left double-J ureteric stent was inserted under general anaesthetic. The patient was discharged 2 days later after the pain and haematuria settled, with a planned follow-up of a repeat renal-tract ultrasound scan after 6 weeks.

The patient represented 7 days postpresentation with recurrence of his left flank pain, visible haematuria and clot retention of urine. A three-way catheter was inserted, an extensive bladder lavage was required and bladder irrigation was commenced. The patient's haemoglobin fell from 148g/l to 82g/l during the first 48 h of this admission; he was consequently transfused with 2 units of red cell concentrate. The catheter subsequently occluded several times with clot. The patient went on to have a bladder washout in theatre, at which time, the ureteric stent was removed. Cystoscopy confirmed no abnormalities in the urethra, prostate or bladder. During this admission, a further CT scan was performed with an angiographic phase (figure 1B) this showed extensive

Figure 1 (A) Initial delayed CT 2.5 h postcontrast. Coronal MPR (multiplanar reconstruction) image. The left pelvicalyceal system contains a cast of clot. There is a markedly delayed left-sided nephrogram, in keeping with significant obstruction (compare to the unobstructed right kidney). No contrast extravasation was seen. (B) CT angiogram on day 7 postinitial presentation. Oblique maximum intensity projection image showing a false aneurysm at the lower pole of the left kidney as well as the proximal pigtail of the ureteric stent. Despite the stent, the kidney remains hydronephrotic. A cast of clot is seen within the pelvicalyceal system.

haematoma and clots within a dilated left-renal collecting system but no capsular rupture. The scan also showed focal intrarenal arterial dilatation, which was thought to be suspicious of arteriopathy or a systemic vasculitis such as polyarteritis nodosa (PAN). Critically, no angiographic phase was performed in the initial CT scan precluding the diagnosis of PAN at this time point. The patient was referred to rheumatology for further investigation. Routine haematology showed a haemoglobin of 76 g/l, platelet count of 271×10^9 and white cell count of 14.6×10^9 , neutrophils 12.5×10^9 . Eosinophil sedimentation rate was 86, and C reactive protein was 40. Levels of complement were normal and testing for antinuclear antigen, antidouble-stranded DNA, extractable nuclear antigens, anti-neutrophil cytoplasmic antibodies, rheumatoid factor, anticyclic citrullinated peptide antibodies and hepatitis B infection were negative.

TREATMENT

On day 10 postpresentation, the patient underwent left renal artery angiography (figure 2A). Angiography of the

left renal artery demonstrated a pseudoaneurysm in the lower pole segmental artery as well as abnormal vessels in the upper pole. The vessel supplying the pseudoaneurysm was selectively visualised with a microcatheter and then coiled, which led to good occlusion of the vessel (figure 2B). The frank haematuria settled following embolisation, but the patient still has persistent haematuria on dipstick testing.

In view of the typical appearances of PAN on arteriography, the elevated ESR and CRP, and the persistent leucocytosis, the patient was commenced on treatment with pulsed methylprednisolone and cyclophosphamide. Six pulses were given at 2–3 weekly intervals over the subsequent 16 weeks and the haematuria settled. Following the last pulse, the patient was started on methotrexate 25 mg weekly, folic acid 5 mg weekly and a decreasing dose of prednisolone starting at 20 mg daily.

OUTCOME AND FOLLOW-UP

A CT aortogram performed 3 months later showed no evidence of new disease or recurrence of arterial dilatation

Figure 2 (A) Renal angiogram on day 10 postinitial presentation. Image of right kidney showing beading of the arterial branches in keeping with microaneurysms. (B) Repeat non-selective left-renal angiogram on day 10 postinitial presentation. The vessel feeding the false aneurysm has now been embolised with a nest of platinum coils. Renal arteries are end arteries so there will be no collateral supply that could continue to feed the false aneurysm. The aneurysm is no longer seen. (C) Follow-up CT angiogram 4 months postpresentation. Coronal maximum intensity projection image showing embolisation coils and associated streak artefact in the lower pole of the left kidney. The false aneurysm is no longer seen. The multiple right renal microaneurysms have resolved.

(figure 2C). At 1 year postpresentation, the patient remained well on methotrexate 25 mg weekly and prednisolone 5 mg daily and by 2 years was off all medication.

DISCUSSION

PAN is a systemic vasculitis affecting medium-sized muscular arteries. It is a rare disease, with an incidence of about 3–4.5 cases per 100 000 population annually.¹ According to the American College of Rheumatology (ACR), criteria of the incidence of PAN in Europe and in the USA is between 2 and 9 per million per year.² PAN can occur at any age, but the most common age at diagnosis is 40–60 years. There is no clear gender difference, although most case series report a slight predominance in men.³

PAN may affect many organ systems and, as a result, may initially present in a number of ways (not necessitating vigorous exercise), frequently leading to a delay in diagnosis. As a result, PAN is often a diagnosis of exclusion. The aetiology of this multisystem disease is incompletely understood, but it is thought to be immune-complex mediated in view of the fact that immunoglobulin M and C3 deposits are detectable in lesional biopsies from patients with skin PAN.⁴ Immune-complex formation is also thought to underlie the association between PAN and hepatitis B infection.⁵

Autoimmune damage to the blood vessel walls and consequent destruction of the media and internal elastic lamina produces aneurysms, which are a classic feature of PAN. PAN is diagnosed via biopsy or arteriography, at least 3 of the 10 ACR criteria should be present.⁶

- ▶ Weight loss of 4 kg or more
- ▶ Livedo reticularis
- ▶ Testicular pain or tenderness
- ▶ Myalgia or leg weakness
- ▶ Mononeuropathy or polyneuropathy
- ▶ Diastolic blood pressure greater than 90 mm Hg
- ▶ Elevated blood urea nitrogen or creatinine level (not related to dehydration or obstruction)
- ▶ Presence of hepatitis B surface antigen or antibody in serum
- ▶ Arteriogram demonstrating aneurysms or occlusions of the visceral arteries
- ▶ Biopsy of small-sized or medium-sized artery containing polymorphonuclear neutrophils

The patient whose case is reported above is unusual in that only one of the ACR criteria was present but there seemed little doubt clinically that he had PAN in view of the raised erythrocyte sedimentation rate and C reactive protein, leucocytosis and resolution of the aneurysms with immunosuppressive therapy.

Learning points

- ▶ An angiographic phase is critical when performing a CT for 'renal trauma' or in torrential upper renal-tract bleeding. This patient's initial scan was not optimally protocolled, resulting in the delayed diagnosis of polyarteritis nodosa (PAN).
- ▶ This case report of a patient with only one of the American College of Rheumatology diagnostic criteria highlights the need for a high index of suspicion in order to make a timely diagnosis of this interesting, multisystem disease so that patients can start promptly on appropriate treatment.
- ▶ Extensive haematuria requiring blood transfusion and operative intervention, that is, for clot retention may be necessary with PAN.

Competing interests None.

Patient consent Obtained.

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