Giant cell arteritis presenting as renal vasculitis

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Summary
Giant cell arteritis commonly presents with headache, polymyalgia, and visual signs and symptoms. Other neurological, respiratory or vascular symptoms occur in 10–30% of patients. It is extremely rare for giant cell arteritis to present initially with haematuria. Here we describe a case which presented with fever and haematuria, which emphasise the need to be vigilant about the diagnosis of giant cell arteritis as an underlying cause.

Keywords: pyrexia of unknown origin; giant cell arteritis; renal vasculitis

Giant cell arteritis is characterised by granulomatous inflammation of medium and large arteries and occurs in people over 50 years old. The vessels most commonly involved are those supplying the head and neck (especially the temporal artery) but there are reports of involvement of the coronary arteries, and arteries to the breast and the uterus. The most common manifestations include fatigue, fever, headaches, jaw claudication, loss of vision, scalp tenderness and elevated erythrocyte sedimentation rate (ESR).

Patients can present with nonspecific symptoms including pyrexia of unknown origin. Renal involvement is very rare and has only been described in a few patients who usually presented with transient microscopic haematuria.

We report a case of giant cell arteritis which presented as pyrexia of unknown origin with renal involvement. Renal biopsy showed granulomatous vasculitis. The patient did not have any other signs of giant cell arteritis. She responded to empirical systemic steroid therapy but relapsed, with classical symptoms and signs of temporal arteritis confirmed by temporal artery biopsy, one year later when the steroids were reduced.

Case report
A 75-year-old woman, previously well, presented with fever with rigours, anorexia, malaise and myalgia for 10 days. She had undergone dental treatment three weeks previously. She denied any history of foreign travel and drug intake. She gave a history of insect bites on her legs a few days before the onset of symptoms.

Clinical examination showed a body temperature of 39°C, and no pallor, jaundice, lymphadenopathy, rashes or joint swelling. Temporal arteries were pulsatile and non-tender as were all her other peripheral pulses. There were no arterial bruits. Systemic examination was normal.

Investigations showed a haemoglobin of 10.1 g/dl (normal range 11.5–16.5 g/dl) with a normochromic normocytic picture. White cell count was raised at 13.3 x 10⁹/l (4–11 x 10⁹/l) along with a raised platelet count of 668 x 10⁹/l (150–400 x 10⁹/l), ESR 100 mm/h (1–15 mm/h), plasma viscosity 2.02 (1.5–1.72) and C-reactive protein 276 mg/l (0–30 mg/l). Urine dipstick showed protein + and blood +++ (normal). Liver function tests showed raised liver enzymes with alkaline phosphatase 622 IU/l (30–120 IU/l), gamma-glutamyl transferase 225 IU/l (2–35 IU/l), aspartate transaminase 45 IU/l (13–34 IU/l), alanine transaminase 82 IU/l (4–24 IU/l) and normal lactate dehydrogenase (285 IU/l). Chest X-ray, electrocardiogram and echocardiogram were all normal. Urine microscopy confirmed microscopical haematuria, but was otherwise normal. Repeated blood and urine cultures were negative. An ultrasound scan of the abdomen showed normal liver, spleen, gall bladder, pancreas and kidneys. White cell scan was normal. The titre of antibodies for cytomegalovirus, herpes simplex virus and C Burnett were not raised in paired sera samples. Serum complements were normal. Rheumatoid factor was negative. Auto-antibody screens, including nuclear (HEP2) and ANCA, were negative.

During admission the patient underwent a left kidney biopsy which revealed granulomatous arteritis (figure 1) with focal interstitial infiltrate composed of lymphoid cells and numerous eosinophils. The glomeruli showed a mild degree of mesangial proliferation but were otherwise unremarkable. Differential diagnosis at this stage was thought to include sarcoidosis, Wegener's granulomatosis, drug reaction and temporal arteritis. She declined temporal artery and liver biopsies. As the chest X-ray was normal, she had no ear, nose and throat symptoms, and ANCA was negative, she was treated for nonspecific arteritis with oral steroids (prednisolone 40 mg daily) and made a good recovery. Her ESR came down to 6 mm/h (1–15 mm/h) within a few days. Proteinuria and haematuria disappeared by the end of the first week. Her steroids were slowly tapered and during this period she was reviewed in the clinic at two-monthly intervals. The raised liver enzymes had returned to baseline at the first two-monthly review. No new symptoms and signs were noted.
Giant cell arteritis

Twelve months later she presented with a recurrence of her fever and myalgia affecting her shoulder girdle muscles, jaw claudication, loss of appetite and temporal headaches. The temporal arteries were tender but pulsatile. There was no recurrence of haematuria or proteinuria. A full blood count and renal and liver function tests were normal, plasma viscosity raised at 1.91 (1.5–1.72) and C-reactive protein at 80 mg/l (0–30 mg/l). A left temporal artery biopsy showed active giant cell arteritis (figure 2). She again made a good recovery with an increased dose of steroids.

Discussion

Pyrexia of an unknown origin continues to be one of the most challenging situations facing a physician. In a series of 85 patients with pyrexia of an unknown origin, Barbado et al reported that the most frequent cause was collagen vascular disease (29/85), in particular systemic vasculitis (temporal arteritis 10/85, polyarteritis nodosa 9/85). Other causes of pyrexia of an unknown origin were infections (9/85), tumours (24/85), and miscellaneous conditions such as factitious fever, liver disease, idiopathic granulomatosis and Crohn’s disease (14/85); in 13 cases, no diagnosis was made.

The kidneys are frequently affected by systemic small vessel vasculitides, especially microscopic polyangiitis (microscopic polyarteritis), Wegener’s granulomatosis, Henoch–Schonlein purpura and cryoglobulinemic vasculitis. Large vessel vasculitides such as giant cell (temporal) arteritis and Takayasu arteritis, only rarely injure the kidneys, usually by ischaemia secondary to vasculitic involvement of the renal arteries or abdominal aorta.

When renal involvement does occur in giant cell arteritis, transient microscopic haematuria (as in our patient) is the most common finding. Some patients have normal renal function with intermittent haematuria, red cell casts and minimal proteinuria. Two cases of nephrotic syndrome have been reported, one associated with membranous glomerulonephritis. One patient developed acute renal failure, which recovered completely with steroids, but a biopsy was not undertaken so the cause of the renal failure is unknown.

Biopsy proven renal vasculitis has been described, but only very rarely in patients with temporal arteritis usually at necropsy. Typical findings are giant cell infiltration and a necrotising small vessel arteritis. There are only nine reported cases in the literature of temporal arteritis with glomerular and/or intrarenal vasculitis associated with significantly impaired renal function. Three patients had focal glomerulonephritis, with or without necrotising features and crescents, one patient had ‘lupus’ nephritis, and one patient, who died of renal failure, had giant cell arteritis without an associated glomerular lesion, as seen in our patient. In four cases either renal lesions were not documented by biopsy or the published illustrations are not adequate for accurate interpretation.

Learning point

Giant cell arteritis should be included in the differential diagnosis of an acute systemic vasculitis with renal involvement

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