CASE REPORT

Cracking haematuria in adolescent with negative work up

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SUMMARY

A 16-year-old girl was referred to our nephrology clinic with persistent visible haematuria (2 months), no urinary tract infection, proteinuria ranging from negligible to 1.5 g/l, normal kidney function and otherwise negative work up including immunology screen. After negative ultrasound scans of the kidneys and bladder, normal renal biopsy and normal cystoscopy, a CT angiogram was obtained. It showed no evidence of arteriovenous malformation but revealed compression of left renal vein (nutcracker syndrome).

BACKGROUND

Haematuria is frequently seen in patients followed by general paediatricians and nephrologists. We would like to present a case of persistent macroscopic haematuria caused by rare, though diagnostically important, nutcracker syndrome. We discuss the step-by-step approach we have taken in this patient, which eventually led us to correct diagnosis.

CASE PRESENTATION

A generally healthy 16-year-old girl was referred to our nephrology clinic with a 2-month history of persistent macroscopic haematuria. Her medical history and family history were unremarkable. She reports no pain or dysuria and was unresponsive to an initial course of antibiotics given for presumed urinary tract infection. Prereferral workup revealed macroscopic haematuria with more than 50 red cells per high-power field and proteinuria ranging from negative to 1.5 g/l on several urine collections, normal blood count, kidney function test (creatinine of 46 µmol/l), ultrasound (US) scans of kidneys and bladder, and cystoscopy.

The patient was 55.1 kg, 172 cm, with a normal blood pressure of 120/80. The cardiovascular, chest and abdominal examinations were all normal and there was no evidence of peripheral oedema. A urine dipstick test found 0.3 g/l of protein, confirmed a 3+ haematuria and was negative for leucocytes and nitrites. The urine protein/creatinine ratio was normal at 18 mg/mmol and C3/C4 complement, antinuclear antibody and antineutrophil cytoplasmic antibody were also normal. There was no evidence of hypercalciuria.

The patient was suspected to have IgA nephropathy, though there was a lack of a preceding upper respiratory tract infection and haematuria was persisting in nature. Kidney biopsy was performed and found to be normal: no changes on light microscopy, negative immunofluorescence and normal glomerular

basement membrane on electron microscopy. A CT angiography was ordered to rule out renal arteriovenous malformations (AVM) and occult tumours.

INVESTIGATIONS

The CT report indicated that an arterial and delayed phase examination of the kidneys was performed. No focal lesions of the kidneys and no vessel tangles suggestive of AVM were found. The renal artery and ureters appeared normal and no free fluid was noted. Adrenals were also reported to be normal and the bladder was found to be within the normal range with no evidence of wall thickening. The initial impression given indicated that it was a normal CT of the kidneys, ureters, bladder and renal arteries. Further review of CT angiogram images revealed evidence of left renal vein (LRV) compression (figure 1). Although the patient had no other symptoms typically associated with nutcracker syndrome such as left flank pain, pelvic discomfort, fullness or varicosities, given the radiological finding and negative haematuria workup, a diagnosis of nutcracker syndrome was confirmed following review by the vascular surgeon.

TREATMENT

As the patient had no risk factors for vascular disease, a conservative treatment approach was recommended as cases of nutcracker syndrome in the paediatric population have been shown to spontaneously resolve with age.^{1 2} The patient was also recommended to gain weight in hopes that increased mesenteric fat deposition between the aorta and the superior mesenteric artery (SMA) would relieve the compression of the LRV.¹

DISCUSSION

Nutcracker syndrome is well described in the literature as a cause of both microscopic and gross haematuria.^{3–7} Also known as LRV entrapment, the nutcracker anatomy occurs when the LRV becomes entrapped between the SMA and the aorta.^{2 8} The prevalence of nutcracker syndrome is not known although it appears to be slightly higher in women and can present at any age, commonly in the third and fourth decade of life.⁹

The most common presenting symptom for nutcracker syndrome is haematuria with left flank pain, often exacerbated by physical activity.^{2 9} Haematuria varies from microscopic to gross and is most likely caused by a rupture of congested renal veins into the collecting system and is often found

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Figure 1 CT angiogram demonstrating compression of left renal vein between aorta and superior mesenteric artery.

to be of left ureteral origin on cystoscopy.^{2–4 6 7} Other common symptoms include gonadal vein syndrome, orthostatic proteinuria, left-sided varicoceles in men and pelvic venous congestion in women.^{2 9} Patients can vary substantially in their presentation and in the involvement of extragenitourinary systems which can include the development of compensatory collateral vasculature and chronic fatigue syndrome.² The natural history of the disease and any potential long-term renal consequence as a direct result of nutcracker syndrome are not well characterised.^{2 9–11}

Owing to a lack of distinct clinical features and diagnostic criteria, the diagnosis of nutcracker syndrome is often made only after other potential causes of haematuria have been excluded, often after multiple investigations.^{2–4} Although it is remarked that the diagnosis of nutcracker is often delayed, no specific average time to diagnosis has been characterised in the literature.^{2 9–11}

Nutcracker syndrome is frequently identified on CT angiograms as happened in our patient. The current gold standard for a definitive diagnosis of nutcracker syndrome is venography and measurement of the renocaval pullback pressure gradient^{2 8 9} which we considered to be too invasive for a patient managed conservatively. Renal Doppler US is a useful non-invasive diagnostic tool to detect a distended LRV with a sensitivity of 69–90% and a specificity of 89–100% though it does not provide a definitive diagnosis for nutcracker syndrome.^{2 12}

The severity of symptoms in nutcracker syndrome is used to guide treatment that can vary hugely with a range from observation to nephrectomy depending on the symptoms.^{2 9} For conservative treatments, an improvement of symptoms is expected within 24 months, more invasive interventions are indicated for unresolved symptoms beyond this point.^{9–11} For more severe cases including extreme pain, extensive haematuria and renal insufficiency, more aggressive interventions including external stenting, LRV bypass, LRV transposition, SMA transposition and even nephrectomy could be performed.^{9–11} The primary treatment option for patients unresponsive to conservative treatment is endovascular stenting that has shown to be effective in relieving symptoms associated with nutcracker syndrome with few risks, although the long-term effects of endovascular stenting are not well known.^{13 14}

In conclusion, it is important for the clinician to remain suspicious of nutcracker syndrome when evaluating a patient with haematuria of unknown aetiology, even in the absence of other symptoms. Careful evaluation of images for evidence of LRV compression is important in early diagnosis of nutcracker syndrome.

Learning points

- ▶ Left renal vein compression (nutcracker syndrome) could be an underdiagnosed cause of haematuria especially in teenagers and should be considered as important differential diagnosis if baseline workup is negative.
- ▶ In our case nutcracker syndrome presented with no other features apart from macroscopic haematuria.
- ▶ A CT angiography should be considered in cases of persistent macroscopic haematuria with particular attention to venous phase.

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