Unusual association of diseases/symptoms

Haematuria and the retro-aortic left renal vein

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Correspondence to Dr Katherine E Jones, katherine.jones@ouh.nhs.uk**Summary**

The authors present the case of a 10-year-old boy with a 2-year history of intermittent macroscopic haematuria. After exclusion of other causes of haematuria through history, examination and a number of investigations, MRI revealed the finding of a retro-aortic left renal vein, which appeared to be compressed between the aorta and the L3 vertebral body. The retro-aortic left renal vein or 'posterior nutcracker syndrome' is a rare but important cause of haematuria and other manifestations of left renal vein hypertension. A discussion about the implications of this finding in children with symptoms consistent with left renal vein hypertension, and the potential options for management is undertaken.

BACKGROUND

Anomalies in the venous drainage of the kidneys are uncommon.¹ The occurrence of these venous anomalies can be explained by the complex series of appearances, regression and anastomoses of paired tributaries of the inferior vena cava during the 4th to 8th weeks of embryological development. The dorsal arch of the left renal vein and the intrasupracardinal venous anastomosis usually regress. In rare cases, its persistence gives rise to either the retro-aortic left renal vein (the ventral arch regresses instead), or the circum-aortic left renal vein (the ventral arch also persists, leading to a duplication).²

This anomalous drainage may cause compression of the left renal vein between the aorta and the lumbar spine, leading to manifestations of left renal vein hypertension. The combination of symptoms in association with the retro-aortic left renal vein is rare. It is termed the 'posterior nutcracker syndrome', in contrast to the more common 'anterior nutcracker syndrome', in which compression of a normally situated left renal vein between the aorta and the superior mesenteric artery near its origin gives rise to similar clinical phenomena. Symptoms and syndromes described are varied, and can be explained by the effects of left renal vein hypertension and back pressure on its tributaries, most notably the gonadal veins.

Although the retro-aortic left renal vein is a congenital phenomenon, the posterior nutcracker syndrome has very rarely been reported in paediatric series. We present the case of a 10-year old boy with a history of intermittent haematuria who was found to have a retro-aortic left renal vein on imaging. We discuss the implications of this finding, and the options for its management in children.

CASE PRESENTATION

A 10-year-old boy presented to the urology outpatients' clinic with a 2-year history of intermittent frank, painless haematuria; with superimposed episodes of left flank discomfort. He was previously fit and well, and there was no history of trauma or symptoms suggestive of urinary tract infection. There had been an antenatal finding of left

hydronephrosis, which had remained stable on postnatal follow-up that had subsequently been discontinued. His renal function was normal, and there was no family history of renal or urological disease.

Abdominal examination and blood pressure were normal. Urine specimens showed no evidence of protein, cell casts, abnormal red cell morphology and no growth of micro-organisms. A series of renal tract ultrasound scans performed over an 18-month period revealed stable mild left pelvicalyceal system dilatation, with normal kidneys and bladder. An unenhanced CT scan of the abdomen did not provide evidence of calculi. Diagnostic cysto-urethroscopy did not demonstrate any urethral or bladder anomalies and no bleeding was observed during the procedure.

A MRI of the abdomen was performed to assess for potential anatomical anomalies or vascular malformations. A review of the imaging showed that the left renal vein passed behind the aorta, and appeared to be compressed between the aorta and the L3 vertebral body (figure 1), the so-called posterior nutcracker phenomenon. In view of the exclusion of other potential causes of haematuria, his symptoms were attributed to the posterior nutcracker syndrome.

OUTCOME AND FOLLOW-UP

The patient remains well under regular follow-up, with serial ultrasonography. In view of the infrequent and benign nature of his symptoms, non-operative management has been advocated, with an intention to treat any arising consequences of the haematuria, such as anaemia.

DISCUSSION

The retro-aortic left renal vein is an uncommon venous anomaly. Based on findings in dissected specimens and from abdominal imaging, the estimated incidence may be as low as 0.3%, depending on the series.¹ Symptoms of left renal vein hypertension secondary to compression between the aorta and lumbar spine are even less common. Reported cases in the adult literature reveal manifestations including breathlessness, fatigue, microscopic and

Figure 1 Sagittal CT slice at L3 vertebral level showing the left renal vein passing behind the aorta.

macroscopic haematuria, labile hypertension, proteinuria, pelvic congestion in women and varicocele in men.^{2 3} Notably, the risk of varicoele in men with the retro-aortic left renal vein was 77% compared with 15% in the general male public.³ The significance of this finding in relation to the potential effect on fertility has yet to be explored.

The anterior nutcracker syndrome has been described in children⁴; however, the posterior nutcracker syndrome appears to be a much rarer finding in childhood, with no paediatric case series found.

In patients with symptoms, the diagnosis can be made by abdominal imaging, with multiphase CT urography and MRI.² Digital subtraction angiography and venography have also been employed. The role of Doppler ultrasound with measurement of the peak velocity of blood flow in the left renal vein in children remains uncertain. In our case, ultrasonography did not reveal evidence of left renal hypertension or the diagnosis of a retro-aortic left renal vein.

The natural histories of both the anterior and posterior nutcracker syndromes remain poorly understood. However, the anterior nutcracker syndrome in children appears to resolve spontaneously with age in as many as 75% of patients.⁴ Postulated theories include the likelihood that an increase in body mass index effects changes in the aorto-mesenteric angle, which leads to a reduction in the degree of left renal vein compression. It is currently unknown if a similar phenomenon occurs in the posterior nutcracker syndrome.

The potential treatment options vary depending on the severity of symptoms and likelihood of resolution. Non-operative management should be considered in the first place, with surveillance and treatment of the manifestations of left renal vein hypertension, such as anaemia, hypertension or orthostatic proteinuria and varicocele. Interventions have been described, including endoscopic stenting of the left renal vein, and surgical correction of the anomalous placement of the left renal vein or gonadal vein, with a corresponding improvement in symptoms.⁵ Surgical intervention could potentially include laparoscopic or open techniques, but the safety and feasibility profiles of each warrant further exploration.

Due to the small numbers in the literature, the long-term outcomes in these patients remain largely unknown. The likelihood of resolution of symptoms with age, as in the anterior nutcracker syndrome, should be considered in children. Therefore, the risks of surgical intervention must be carefully weighed against the severity of the patient's symptoms, the likelihood of spontaneous resolution, and the potential benefit of the technique.

Learning points

- ▶ After exclusion of other common causes, renal vascular anomalies should be considered in a patient with intractable haematuria.
- ▶ Imaging (in particular CT and MRI) usually establishes the diagnosis.
- ▶ Treatment is based on a careful assessment of the severity of the symptoms weighed against the potential risks of surgery and other invasive interventions.
- ▶ In paediatric patients, implications for the future should be considered, such as the likelihood of development of varicocele in males.
- ▶ Long-term routine surveillance is advocated in all patients.

Competing interests None.

Patient consent Obtained.

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