

*Teaching Point*

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**A young woman with intermittent macroscopic haematuria**Olivier Lidove<sup>1</sup>, Rodrigo Orozco<sup>1</sup>, Bruno Guéry<sup>1</sup>, Jean-Michel Correas<sup>2</sup>, Christophe Robino<sup>1</sup>, and Arnaud Méjean<sup>3</sup>Departments of <sup>1</sup>Nephrology, <sup>2</sup>Radiology and <sup>3</sup>Urology, Necker Hospital, Paris, France**Case report**

A 34-year-old black woman was admitted after 1 month of intermittent gross haematuria, initially associated with left loin pain. Physical examination was unremarkable. Renal function was normal and 24-h proteinuria was negative. Haemoglobin level was 11.5 g/dl and haemoglobin electrophoresis was normal. Renal ultrasound and intravenous urography were normal. Urine culture was negative. Schistosomiasis (*S. haematobium*) and tuberculosis were excluded. Two cystoscopic examinations were performed but failed to show the origin of haematuria. The increased haematuria with orthostatism led to the performance of a colour duplex sonography which showed a dilatation of the left renal vein, highly suggestive of Nutcracker syndrome. This diagnosis was confirmed by an enhanced CT scan of renal veins showing left renal vein compression between the superior mesenteric artery and the aorta (Figure 1). In the absence of lumbar pain or anaemia, the patient was discharged without treatment except iron and folate supplementation.

Two months later, the patient was readmitted because of persistent gross haematuria, lumbar pain and anaemia (haemoglobin 9 g/dl). A third cystoscopy revealed bloody urine coming from the left ureteral orifice only. Arterial phase of selective left renal arteriography was normal. Selective left phlebography did not show compression of the left renal vein between the superior mesenteric artery and the aorta, but showed venous collateral circulation with numerous varicosities (Figure 2).

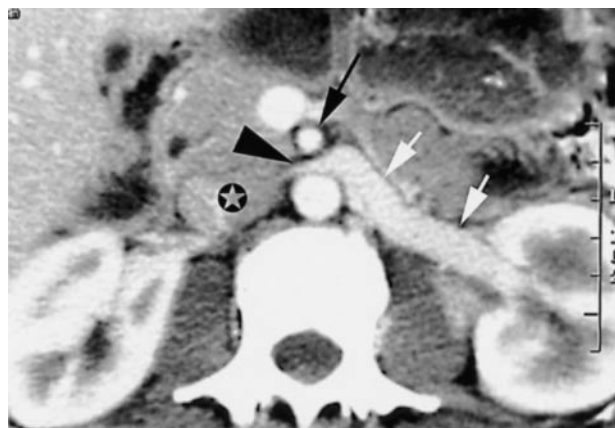
A surgical procedure was therefore performed to relieve the venous compression. The left renal vein was

lysed, divided, brought inferiorly and re-anastomosed to the vena cava (Figure 3). Haematuria completely disappeared on day 3 of follow-up.

Three months later, the patient was in good general condition. She reported no episode of haematuria or loin pain. Haemoglobin level was then 11.7 g/dl.

**Discussion**

Nutcracker syndrome (NS) is a cause of haematuria originating from the left collecting system. It was first described by De Schepper in 1972 [1], who showed that “idiopathic renal bleeding” was often due to NS, also called “left renal vein entrapment syndrome”. NS results from compression of the left renal vein between the superior mesenteric artery and the aorta, leading to

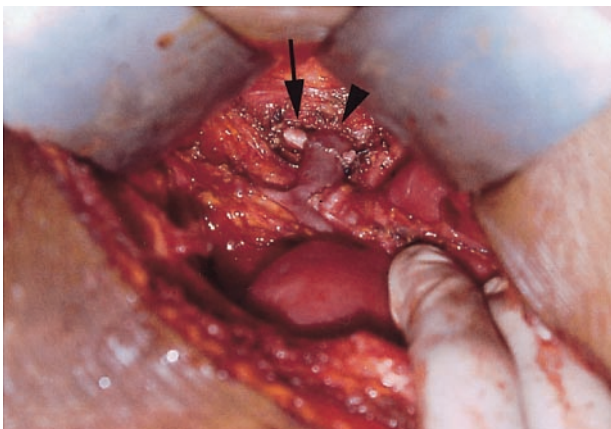


**Fig. 1.** Spinal CT scan of the abdomen. The proximal and middle left renal vein segments were slightly enlarged (white arrows). The distal left renal vein was compressed between the superior mesenteric artery (black arrow) and the aorta, and appeared as a poorly opacified channel (arrowhead) before the inferior vena cava (black star). Of note, pyelic and parapyelic varices were not shown.

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**Fig. 2.** Selective left phlebography, exhibiting a large venous collateral circulation with periureteral varicosities (arrow) and lumbar anastomoses. Black star, inferior vena cava; white star, left renal vein.



**Fig. 3.** Peroperative view. The left renal vein was long enough to be re-anastomosed (arrow head) in front of the superior mesenteric artery (arrow). Note that the left renal vein was clearly enlarged.

renal venous hypertension and formation of ureteral and renal pelvic venous varicosities. This syndrome causes clinical symptoms such as recurrent episodes of gross haematuria and left loin pain, with increased haematuria with orthostatism, but it may also be asymptomatic, revealed by microhaematuria. Male

patients can develop left varicocele. The frequency of this syndrome seems to be similar in both genders. The incidence of this anatomical anomaly is unknown, and is probably underestimated.

The first steps in the investigation are to exclude the main causes of gross haematuria and to demonstrate unilateral left ureteral bleeding by cystoscopy. Intravenous urography may inconstantly show compression of the left lumbar ureter, due to periureteral varices. Angiographic CT scan (or MRI) is the most reliable way to demonstrate left renal vein entrapment and compression, and the development of the collateral vein network. Renal phlebography and pressure measurements in the left renal vein to establish the pressure gradient between the distal portion of the vein and the inferior vena cava are not necessary for diagnosis. The pressure gradient depends on the intensity of development of the collateral circulation through gonadal, capsular, suprarenal, lumbar, azygos and periureteral veins, and a clear cut-off between normal and pathological pressure gradient values does not exist.

The underlying pathophysiology of the NS is not yet completely clarified. Hohenfellner *et al.* found abnormal configuration of the branching of the superior mesenteric artery from the aorta in three patients studied by MRI [2]. Most probably, bleeding into the upper urinary tract results from rupture of thin-walled varices surrounding the collecting system. Of interest, and not surprisingly, a CT study has shown a 'high' prevalence (>50%) of slight and asymptomatic compression of the left renal vein in the general population [3]. In a study of patients with haematuria related to abnormalities of the renal and perirenal venous system, the left side was found to be affected more frequently, suggesting increased venous outflow resistance on this side [4]. Slower venous washout from the left than from the right kidney may also play a role and explain the predominance (80% of cases) of left renal bleeding in patients with sickle cell trait and gross haematuria [5]. In this regard, it should be recalled that recurrent gross haematuria, probably due to minor episodes of papillary necrosis, may occur in black but also in white patients with sickle cell trait (heterozygotes), and that these patients should be screened for haemoglobin abnormalities [6].

Treatment of NS depends on the severity and consequences of the bleeding. Patients with intermittent and rare episodes of haematuria and no anaemia may only require close follow-up with intermittent iron supplementation, if needed. In contrast, surgery must be considered in the rare cases where gross haematuria is persistent or frequently recurs, causing anaemia (as observed in our patient) and/or is accompanied by left flank pain. Different surgical procedures have been advocated: in the past, nephropexy or excision of peripelvic varices, and in more recent years, left renal venous bypass or transposition of the left renal vein into the vena cava. Kidney autotransplantation is an alternative procedure and has been performed successfully in some patients [7,8]. Balloon angioplasty and

stent implantation were performed recently in a patient by Segawa *et al.*, but this procedure entails the risk of vein occlusion or stenosis in the long-term and thus requires evaluation [9].

In our patient, left renal vein transposition was successful. This effect can probably be ascribed to lowering of renal vein pressure. However, Shaper *et al.* have observed unchanged pressures 6 weeks post-operatively despite resolution of haematuria in four of their five patients [10]. This shows again the poor sensitivity of pressure measurements in the renal vein and that these measurements should therefore be regarded with caution.

### Teaching point

Nutcracker phenomenon can lead to macroscopic haematuria and to anaemia. The bleeding is unilateral. CT scan or MRI demonstrates entrapment of the left renal vein between superior mesenteric artery and aorta. Surgical correction may be indicated and lead to complete cure.

### References

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