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Recurrent hematuria due to nutcracker syndrome: Imaging techniques

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Introduction

Recurrent hematuria in children without UTI is mainly explained by a glomerulopathy, hypercalciuria/nephrolithiasis, Alport syndrome or nutcracker syndrome. The latter results **from compression of the left renal vein** between the superior mesenteric artery and the aorta **(Fig.1)** **Clinical presentation** of **nutcracker syndrome** can vary from asymptomatic hematuria to severe abdominal pain. Mild proteinuria can be associated. Left renal venography is considered to be the gold standard, but is invasive. **Doppler ultrasonography** can be used as first diagnostic test. However, two cases are presented where nutcracker syndrome was diagnosed with MRA, while Doppler ultrasound showed no abnormalities.

Case Reports

CASE 1:
A 16 years old boy presented with 2 identical episodes of macroscopic hematuria with mild flank pain after viral infection. Symptoms disappeared after 3 days. Inter-episode urine samples showed microscopic hematuria and discrete proteinuria. On Doppler ultrasonography there were no arguments for nutcracker syndrome **(Fig. 2)** However, MRA revealed compression of the left renal vein between the aorta and superior mesenteric artery **(Fig. 3)**

CASE 2:
A 7 years old boy presented with a second episode of macroscopic hematuria (without proteinuria) since 1 day. The first episode occurred at the age of 3 years and disappeared after 2 days. Both episodes were identical, with no clinical symptoms besides mild abdominal pain. In both cases the diagnostic work-up for hematuria was performed. Laboratory blood investigations and 24-hour urine collection were within normal ranges. Renal ultrasonography showed no nephrolithiasis. Doppler ultrasonography showed no signs of renal vein compression. MRA did show left renal vein entrapment.

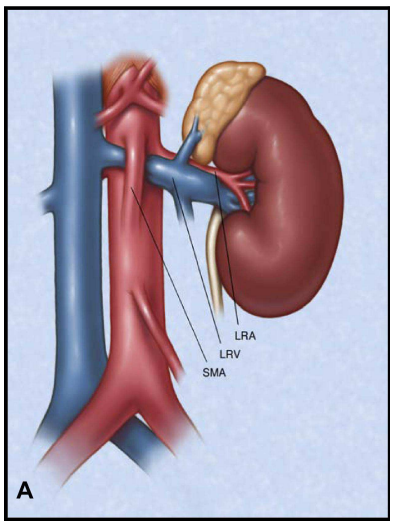

Fig. 1: compression of the left renal vein between the superior mesenteric artery and the aorta
(Source: http://dx.doi.org/10.1016/j.jvs.2008.09.051)

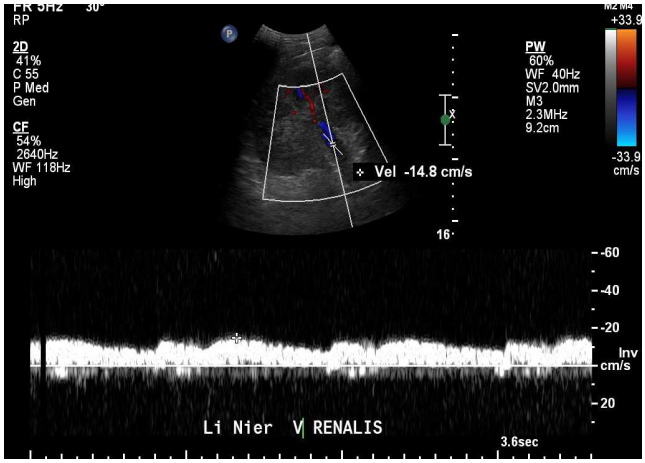

Fig. 2: Doppler ultrasonography showed no decreased diameter of the left renal vein and normal bloodflow rate .

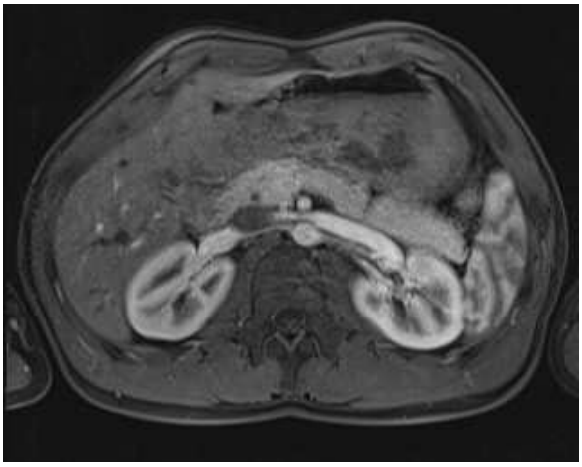

Fig. 3: MRA revealed a dilated left renal vein with compression between the aorta and superior mesenteric artery

Discussion

Doppler ultrasonography can be used as first diagnostic test for renal vein entrapment,but has a limited sensitivity, especially in children.
MRA findings are similar to CT and avoid radiation. In case of recurrent hematuria, a nutcracker syndrome should be excluded before performing a renal biopsy, which is an invasive procedure. Expectation management is appropriate in the great majority of children with confirmed nutcracker syndrome.

Conclusion:

Nutcracker syndrome is a rare cause of recurrent hematuria in children but should always be considered, even when Doppler ultrasonographic assessment shows NO anomalies.
By performing a MRA-scan, unnecessary renal biopsies can be avoided in these patients.

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