

Advance Access publication 14 September 2011

Nutcracker phenomenon and idiopathic IgA nephropathy

Sir,

Entrapment of the left renal vein between the aorta and the superior mesenteric artery is a documented cause of both postural proteinuria as well as intermittent gross haematuria associated with left flank pain. Idiopathic IgA nephropathy typically presents with recurrent episodes of gross haematuria, usually following an acute febrile illness. Rarely, the disease presents as microscopic haematuria and usually mild proteinuria. We present the first case of postural proteinuria caused by left renal vein entrapment and recurrent gross haematuria caused by an IgA nephropathy.

In an 11-year-old boy with postural proteinuria, renal ultrasonic Doppler disclosed left renal vein entrapment: the anteroposterior diameter at the hilar portion divided by that at the aortomesenteric portion was 13.3 (reference ≤ 4.0) and the peak flow velocity at the aortomesenteric portion divided by that at the hilar portion 15.1 (reference ≤ 4.0).

Two years later, three episodes of acute febrile diarrhoea were followed by gross haematuria that was not associated with flank pain. The kidney biopsy showed mild diffuse mesangial proliferation and matrix expansion. The diagnosis of IgA nephropathy was made on immunofluorescence microscopy, which demonstrated isolated prominent glomerular deposits of IgA, C3 and IgG in the mesangium.

The link between left renal vein entrapment and postural proteinuria is well documented. Furthermore, both left renal vein entrapment and IgA nephropathy are recognized causes of recurrent gross haematuria.

There are five reported patients affected with both left renal vein entrapment and idiopathic IgA nephropathy [1–4]. A 12-year-old German girl with microscopic haematuria was found to have left renal vein entrapment and mesangial deposits of IgA [1]. Similarly, left renal vein entrapment and mesangial deposits of IgA were reported in a 9-year-old Korean girl with isolated microscopic haematuria and recurrent gross haematuria [2] and in a 25-year

old Taiwanese woman with recurrent gross haematuria and left flank pain [3]. Left renal vein entrapment and IgA nephropathy were also disclosed in a 20-year-old woman and in a 22-year-old Japanese man, who, in the context of a pharyngitis, developed gross haematuria and a tendency to microscopic haematuria with persisting proteinuria [4]. In addition, left renal vein entrapment was disclosed in two Korean children a 13-year-old Korean girl with Henoch–Schönlein syndrome and IgA nephropathy [5]. Finally, a 10-year-old Korean boy with a clinically characteristic and rapidly resolving form of Henoch–Schönlein syndrome followed by microscopic haematuria persisting for 26 months was found to have left renal vein entrapment; haematuria subsided in accordance with radiological improvement of renal vein entrapment [6].

Our patient is the first case with concurrent existence of postural proteinuria caused by left renal vein entrapment and recurrent gross haematuria caused by an idiopathic IgA nephropathy. Renal venous congestion may induce both proteinuria and haematuria, which have been implicated as a cause of renal damage. So, a causal relationship between left renal vein entrapment and IgA nephropathy cannot be ruled out. Further investigations are necessary to detect this correlation.

Conflict of interest statement. None declared.

¹Emergency Unit, Clinica Pediatrica De Marchi, Foundation IRCCS Cà Granda Ospedale Maggiore Policlinico, Milan, Italy,

²Pediatric Nephrology and Dialysis Unit, Clinica Pediatrica De Marchi, Foundation IRCCS Cà Granda Ospedale Maggiore Policlinico, Milan, Italy, and

³Division of Pediatrics, Mendrisio and Bellinzona Hospitals, University of Bern, Switzerland

E-mail: yoyobiancorosso@hotmail.com

Marta B. M. Mazzoni¹
Gregorio P. Milani¹
Chiara Persico¹
Alberto Edefonti²
Emanuela A. Laicini¹
Mario G. Bianchetti³
Emilio F. Fossali¹

6. Shin JI, Park JM, Shin YH *et al.* Superimposition of nutcracker syndrome in a haematuric child with Henoch–Schönlein purpura. *Int J Clin Pract* 2005; 59: 1472–1475

doi: 10.1093/ndtplus/sfr108

References

1. Liebl R. Nutcracker phenomenon or nutcracker syndrome? *Nephrol Dial Transplant* 2005; 20: 2009
2. Shin JI, Park JM, Shin YH *et al.* Nutcracker syndrome combined with IgA nephropathy in a child with recurrent hematuria. *Pediatr Int* 2006; 48: 324–326
3. Chen YM, Wang IK, Ng KK *et al.* Nutcracker syndrome: an overlooked cause of hematuria. *Chang Gung Med J* 2002; 25: 700–705
4. Ozono Y, Harada T, Namie S *et al.* The “nutcracker” phenomenon in combination with IgA nephropathy. *J Int Med Res* 1995; 23: 126–131
5. Shin JI, Lee JS. Unexpected superimposition of nutcracker effect in various conditions: is it an unrecognized confounding factor? *Eur J Pediatr* 2007; 166: 1089–1090