Orthostatic proteinuria as a result of venous compression (nutcracker phenomenon)—a hypothesis testable with modern imaging techniques

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Introduction

The nutcracker phenomenon [entrapment of the left renal vein (LRV)] refers to compression of the LRV between the aorta and superior mesenteric artery, resulting in elevation of pressure in the LRV, leading to congestion of the left kidney and occasionally to collateral formation. An abnormal branching of the superior mesenteric artery from the aorta is the underlying pathophysiological principle of the nutcracker phenomenon [1]. The posterior nutcracker, one of the rarer patterns of this phenomenon, is characterized by a retroaortic position of the LRV and its compression between the aorta and the spine [2].

Entrapment of the LRV is an unusual but well-acknowledged cause of left sided gross haematuria, ureteral and peripelvic varices, and unexplained left flank and abdominal pain. Variable degrees of proteinuria has been observed in some patients with the nutcracker phenomenon [3–5].

Case

A well developed 11-year-old boy was admitted to our department for clinical assessment following the detection of proteinuria. When proteinuria was detected, he had an upper respiratory tract infection, but proteinuria persisted after the resolution of the febrile illness. He was asymptomatic, normotensive, and physical examination was normal. Urinalysis was normal except 3+ proteinuria and 24 h urinary protein excretion of 0.4 g/day. His night time urine was protein free, but while he was in an upright and ambulatory position in the daytime, urine specimens contained protein. A very high γ-globulin peak was detected by urinary protein electrophoresis. Serum creatinine, blood urea nitrogen, total protein, albumin levels and creatinine clearance were within normal limits. An intravenous urogram was normal. Doppler sonographic examination showed a retroaortic position of the LRV and a high resistivity index in the left renal vessels. Selective left renal venography and aortography confirmed the presence of a markedly compressed LRV between the aorta and the lumbar vertebral body proximal to the insertion into the inferior vena cava (Figure 1).

Discussion

The nutcracker phenomenon (left renal venous hypertension) is an uncommon cause of non-glomerular left renal bleeding and unexplained left loin or abdominal pain [1–3,5,6]. A rarer pattern of this phenomenon, so-called posterior nutcracker, was also reported as the cause of left sided intermittent gross haematuria [2,5]. In the last decade, some authors reported that proteinuria may also be seen in patients with the nutcracker phenomenon. In 1991, Shintaku et al. suggested that LRV entrapment was the cause of orthostatic proteinuria in children [4]. In another study, it was found that nine of 16 patients with the nutcracker phenomenon had various degrees of proteinuria [5]. In 1997, LRV entrapment was documented by bilateral ureteral catheterization and imaging studies as a cause of massive orthostatic proteinuria. These observations suggested that renal congestion due to LRV entrapment was the source of orthostatic proteinuria [7]. Some long-term follow-up studies revealed that orthostatic proteinuria was a benign condition, had a good prognosis and was not associated with any underlying glomerular disease [8,9].

It was shown that urinary γ-globulin was >6% in patients with orthostatic proteinuria who had no other glomerular disease [10]. Shintaku et al. found a high percentage of urinary γ-globulin in three of nine children with LRV entrapment [4]. A very high percentage of urinary γ-globulin was a remarkable finding in our patient with orthostatic proteinuria. Therefore, a high percentage of γ-globulinuria may be a valuable marker of orthostatic proteinuria in patients with LRV entrapment, as described by Shintaku et al.

We suggest that this uncommon entity should be considered in children with haematuria and proteinuria in the absence of any other detectable pathology. A

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Fig. 1. Selective left renal venography (a) and aortography (b) showing the retroaortic position of the left renal vein and its compression between the aorta and the body of the vertebrae.

high percentage of urinary γ-globulin by electrophoresis in patients with orthostatic proteinuria may raise the suspicion of the nutcracker phenomenon. Since Doppler ultrasound has been used successfully for the diagnosis of the nutcracker phenomenon [5], this non-invasive imaging technique might be performed before invasive diagnostic procedures. Nevertheless, selective left renal venography is the gold standard procedure for exact definition of this syndrome and its variants. However, some authors reported that sagittal planes of magnetic resonance imaging (MRI) clearly indicated the nutcracker phenomenon [11]. Thus MRI and the more versatile MRI angiography, which have provided highly reliable information about vascular anatomy, should also be used to study for the diagnosis of the nutcracker phenomenon.

References


