Case Report

Nutcracker syndrome: intravascular stenting approach

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Introduction

The clinical syndrome caused by impingement of the left renal vein (LRV) between the superior mesenteric artery (SMA) and abdominal aorta has been termed Nutcracker syndrome [1]. Although often asymptomatic, it may result in varicocele, ovarian vein syndrome, haematuria, flank or abdominal pain, LRV hypertension, and pelviureteral varices. Surgical approaches for Nutcracker syndrome include nephrectomy, nephropexy, renocaval reimplantation, or auto-transplantation. We satisfactorily treated a middle-aged man with Nutcracker syndrome accompanied by renal enlargement, persistent haematuria, proteinuria and hypertension with a new intravascular stent.

Case

A 47-year-old man was referred from a urologist for hypertension, microscopic haematuria and left renomegaly; left varicocelectomy was performed 7 years ago. Physical examination was unremarkable except for a palpable left kidney. Ultrasonography revealed a normal-sized right kidney of 10.5 cm in length and a large left kidney that was 18 cm long. Urinalysis revealed microscopic haematuria and minimal proteinuria. IVP and ultrasonography revealed enlargement of the left renal pelvis and a convolution at the pelviureteral junction. The kidney biopsy specimen showed nonspecific proliferative change. Repeated ultrasonography and Doppler flow analysis revealed a patent LRV, which abruptly narrowed between the aorta and the SMA. Doppler velocimetric study showed that the diameter of the hilar and aortomesenteric portions of the LRV were 13.3 mm and 2 mm respectively (Figure 1).

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Fig. 1. Ultrasonography of LRV in transverse view. (A) Compressed LRV between the aorta (AO) and superior mesenteric artery (SMA). The diameter was 2.0 mm. (B) The dilated LRV lateral to the impingement. The diameter was 13.3 mm.

The peak velocities in the LRV at the hilar and aortomesenteric portions were 8 cm/s and 76 cm/s respectively (Figure 2). Renal angiography demonstrated a normal arterial phase and the venous phase showed varices in the renal hilum with a tortuous collateral vessel at the site of convolution of the ureteropelvic junction. Renal phlebography revealed a compression of the LRV at the level of the aorta. The pressure gradient between the LRV and the inferior
Fig. 2. Doppler ultrasonography of LRV. (A) The spectrum obtained at the lateral LRV. The peak velocity was 0.08 m/s. (B) The spectrum obtained at the aortomesenteric LRV. The peak velocity was 0.76 m/s.

vena cava at the level between the aorta and the SMA was 3 mmHg (Figure 3).

Under fluoroscopic guidance, a Cobran catheter prosthesis was inserted into the LRV and then a 18-mm Niki stent was inserted over the catheter to relieve the compression. No complications occurred and the patient was discharged next day. Two weeks later, no haematuria or proteinuria were observed on follow-up study. All medications, including antiplatelet agents, were stopped and the patient has been free from hypertension for over 4 months.

Discussion

Compression of the LRV was first described in 1950 [1] and named Nutcracker syndrome. The syndrome

Fig. 3. Renal phlebography. Arrowhead shows obstruction of left renal venous outflow due to compression of renal vein by superior mesenteric artery. Arrows demonstrate perirenal and periureteral varices.
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results from compression of the LRV, usually by the SMA or aorta, or by both arteries.

The pathophysiology of Nutcracker syndrome is not fully understood. The passage of the LRV in the fork formed by the aorta and SMA is a normal anatomical finding. It is not understood why compression of the vein occurs in a few patients only. Wendel et al. [2] have proposed that posterior renal ptosis with stretching of the LRV over the aorta may be a factor. In a recent study, Hohenfellner et al. [3] reported that abnormal branching of the SMA from the aorta contributes to the development of Nutcracker syndrome.

The most common clinical manifestation of Nutcracker syndrome is gonadal varices (varicocele or ovarian vein syndrome). However, Nutcracker syndrome has been invoked only recently to explain the less common condition of left flank pain and haematuria associated with LRV hypertension and pelviureteral varices. The present case had suffered from left varicocele, but the underlying cause was not sought. Haematuria may result from increased LRV pressure causing minute rupture of thin-walled veins into the collecting systems or caliceal fornix [4] or communication between dilated venous sinuses and adjacent renal calices [5]. Haematuria from the left ureteric orifice in the absence of any other detectable pathology should raise the suspicion of Nutcracker syndrome.

Diagnosis of Nutcracker syndrome cannot be established with routine diagnostic methods and, therefore, the natural history of this disease is characterized by repeated diagnostic procedures and delayed treatment [3]. Ultrasonography shows narrowing of the LRV between the aorta and SMA and the widening of the distal LRV. The diameter of the distal LRV of the patient was five times or more larger than the narrowed portion. Venography and venous pressure measurements are accepted as the procedures of choice to establish the diagnosis [6]. Pressure measurements alone bear some diagnostic pitfalls. The result depends on the position of the patient (supine or upright) and on the capacitance of the periureteric, adrenal and gonadal venous plexus. Pressure may vary between 4.9 and 14 cm water in patients with Nutcracker syndrome [6]. In this case, the pressure gradient was 3 mmHg and was far higher than that of normal. There is overlap between normal pressures (ranging from 1.3 to 10 cm of water) and those found with this syndrome and selection of a specific cut-off value for the diagnosis of Nutcracker syndrome would be arbitrary. As a consequence, a whole spectrum of diagnostic findings should be considered before diagnosis and further intervention. Volumetric angiographic CT, MRI, and colour Doppler sonography may be helpful for evaluation of Nutcracker syndrome.

The treatment of Nutcracker syndrome is controversial. Conservative treatment has been proposed for cases with mild haematuria [6], while surgery is indicated for massive haematuria and pain. Medial nephropexy with excision of the renal varicosities has been used by Wendel et al. [2], where posterior renal ptosis is believed to contribute to renal vein obstruction. Coolsaet [7] has advocated renal vein bypass to reduce the pressure in the collateral bed. Transposition of the LRV has also been reported by other investigators [8]. Autotransplantation [9] is an alternative treatment which allows better protection of the kidney against ischaemia by proper cooling and irrigation.

When this syndrome leads to clinical symptoms, an intravascular stent should be considered. Despite the pessimism in the past [10] we believe that a well-designed stent placed with minimal invasiveness may offer physiologic relief as in the present case. To the best of our knowledge, there was no previous publication on intravascular stenting for this indication.

References


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