Nutcracker Syndrome and Left Renal Scarring in a Boy Presenting with Hypertension

Hipertansiyon ile Başvuran Bir Olguda Nutcracker Sendromu ve Sol Renal Skarlanma

Nutcracker Sendomu, Renal Skarlanma ve Hipertansiyon / Nutcracker Syndrome, Renal Scarring and Hypertension

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Özet

Nutcracker sendromu (NCS), sol renal venin (LRV) aorta ve superior mezenterik arter arasında sıkışması ile karakterizedir. Oldukça nadir görülür ve genellikle hematüri, ağrı ya da proteinüri ile kendini gösterir. Bugüne kadar sadece iki erişkin hastada hipertansiyon tanımlanmıştır. Ancak, NCS'de ipsilateral renal skarlanma (RS) bildirilmemiştir. Biz burada hipertansiyon ile başvuran, NCS ve sol RS saptanan 14 yaşındaki bir olguyu takdim ediyoruz.

Anahtar Kelimeler

Sol Renal Ven Sıkışması; Renal Skarlanma; Hipertansiyon; Hematüri

Abstract

Nutcracker syndrome (NCS) refers to the compression of the left renal vein (LRV) between aorta and superior mesenteric artery. It is very rare and usually manifests with hematuria, pain or proteinuria. Hypertension has been defined in two adult NCS cases. However ipsilateral renal scarring (RS) has not previously been demonstrated in any of NCS patients. Here we report a 14-year-old boy presented with hypertension and found to have NCS and left RS.

Keywords

Left Renal Vein Entrapment; Renal Scarring; Hypertension; Hematuria

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Introduction

Nutcracker syndrome (NCS) refers to compression of LRV (left renal vein) between aorta and superior mesenteric artery (SMA). The impairment of blood flow increases pressure in LRV and leads to development of collaterals in renal pelvis, ureter and gonadal vein [1, 2]. NCS usually manifests with hematuria, orthostatic proteinuria, pain or left sided varicocele. Hypertension has been defined in few adult NCS cases [3, 4]. However renal scarring (RS) has not previously been demonstrated in NCS. A hypertensive boy with NCS and left RS is presented here.

Case Report

A 14-year-old boy was referred for hypertension. He was suffering from chronic headache. There was no fever, hematuria, pain or fatigue in past medical history. Blood pressure was 150/100 mmHg. Anthropometric measurements and physical examination were normal. Urinalysis, serum biochemistry, thyroid hormones, plasma renin activity (PRA) and aldosterone levels were within normal ranges. Abdominal ultrasonography (US) was unremarkable. On color Doppler US, peak systolic velocities of renal arteries were normal whereas the diameter of LRV was expanded. Magnetic resonance (MR) angiography demonstrated the compression of left renal vein between SMA and abdominal aorta (Fig. 1a). 99mTc-dimercaptosuccinic acid (DMSA) scintigraphy revealed scarring on upper pole of left kidney (Fig. 1b). The voiding cystourethrogram (VCUG) imaging was normal. The diagnosis of NCS was established. His hypertension was controlled with triple antihypertensive therapy. The patient has been followed up without any complaint in outpatient clinic.

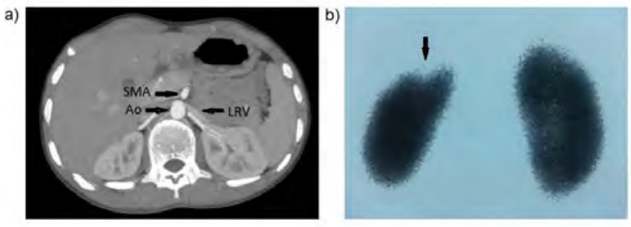


Fig 1. Computed tomographic venogram shows compression of left renal vein (LRV) between superior mesenteric artery (SMA) and abdominal aorta (Ao)(A). Postero-anterior 99mTc-dimercaptosuccinic acid scintigraphy demonstrates scarring on the upper pole of the left kidney.(B).

Discussion

Nutcracker syndrome is very rare and usually late-diagnosed because of the variability of symptoms and absence of consensus on diagnostic criteria. Many of patients, particularly children are asymptomatic [1, 2]. However, the main symptom was hypertension in our patient. Hypertension with NCS has been identified in two case reports in literature [3, 4]. In the first paper, hypertension was found to be associated with increased PRA and aldosterone as the patient became normotensive and PRA normalized after endovascular intervention to LRV [3]. Nevertheless, the authors of the second report were unable to confirm the findings of the first one and concluded that hypertension might be coincidental in NCS [4].

In our patient, hypertension was thought to be associated with RS which is defined by presence of chronic tubulo-interstitial inflammation. Thus, the question was raised whether RS was related to NCS. Although VCUG was normal, the major cause of RS, vesicoureteral reflux (VUR), was not properly ruled out when spontaneous resolution of VUR by older age was considered. On the other hand, previous studies on renal hemodynamic of NCS patients have indicated that congestion of LRV might affect venous return and increase capillary wall permeability. If congestion persists, local tissues may expose to anoxia, followed by oxidative damage, renal arteriospasm resulting in pathological changes such as degeneration, atrophy and necrosis in renal glomerular and tubular cells [5, 6]. Thus, it might be postulated that RS related hypertension in this patient might be a clinicopathological result of NCS.

The treatment of NCS depends on age and severity of clinical symptoms. Conservative approach is recommended for mild cases and also for pubertal children because of high rate of spontaneous resolution probably due to physical development. Patients with serious symptoms may benefit from invasive options including vascular stenting or open surgery [7, 8].

In conclusion, the present case is the first demonstrating the coincidence of NCS and RS in literature. RS might be a clinicopathological consequence of NCS. Further researches might let on the detrimental effects of NCS on kidney.

Competing interests

The authors declare that they have no competing interests.

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