CASE REPORT

Is hypertension a manifestation of the nutcracker phenomenon/syndrome? Case report and brief review of the literature

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Abstract
Hypertension has been rarely reported in patients with the nutcracker phenomenon/syndrome. We describe a young male adult where a computed tomography angiography provided evidence of left renal vein dilatation, probably due to its compression through the angle between the aorta and the superior mesenteric artery, during the evaluation for secondary hypertension. As there were no other signs for secondary hypertension, we proceeded with a venography of the inferior vena cava and the renal veins that revealed mild anatomical findings compatible with the so called nutcracker phenomenon/syndrome. Blood levels of renin and aldosterone and renocaval pressure gradient from these sites were between normal limits. As there were coexisting anatomical and clinical findings (hypertension), nutcracker syndrome might have been claimed. However, no causal links could be established and these findings should be considered only as a coincidence. Hippokratia. 2012; 16 (2): 187-189

Key words: aldosterone, hypertension, nutcracker syndrome, renin, venography

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The terms nutcracker syndrome (NCS) or phenomenon (NCP) refer to the compression of the left renal vein, as it passes through the angle between the aorta and the superior mesenteric artery. These anatomical findings were first described by Schepper in 1972[1]. Although these terms have been used quite often interchangeably in the literature, it should be emphasized that NCS must be used when there is evidence of nutcracker anatomy accompanied with clinical symptoms. Otherwise, the term NCP seems more appropriate[2,3].

NCS is a rare clinical condition characterized usually by mild hematuria, orthostatic proteinuria, left flank pain, left-sided varicocele in males, pelvic congestion, mild anemia and chronic fatigue in adolescents or young adults[2, 4-10]. Hypertension is not included in the classical signs of NCS, whereas Hosotani et al have reported a case of NCP accompanied with renin dependent hypertension that was reversed after endovascular stent placement[11].

We describe a young male with hypertension, where an extensive clinico-laboratorial investigation revealed that he also presented elements of the nutcracker phenomenon.

Case Report

A 20-year-old male caucasian with a four month history of hypertension was admitted for further investigation. Hypertension was previously treated by a fixed combination of 150 mg of irbesartan and 12.5 mg of hydrochlorothiazide and 10 mg of amlodipine. Past medical history was remarkable for an episode of pericarditis and myocarditis of viral aetiology in 2006 that had resolved completely. His family history was not contributory for hypertension or renal diseases.

Physical examination was unremarkable and the combination of 150 mg of irbesartan and 12.5 mg of hydrochlorothiazide was stopped in order to undergo further laboratorial investigation. All blood tests were within normal limits. Urinalysis did not reveal any abnormalities. Multiple 24 hour urine collections did not reveal any proteinuria (13-81 mg/24h). Urinary tests for pheochromocytoma were also normal. Immunological investigation including anti-nuclear antibodies (ANA), complement (C3/C4), serum immunoglobulins etc was within normal limits.

Before admission, the patient had undergone radiological examination including a renal ultrasound with a Doppler renal arteries scan that was normal and a Computed Tomography Angiography (CTA) that had revealed signs of left renal vein stenosis and dilation (Figure 1).

As there was no other signs indicative of secondary hypertension, we decided to explore the possibility that hypertension might be related with the abnormal left vein anatomy. Under local anaesthesia, a venography was performed in order: a) to reveal the grade of the left renal

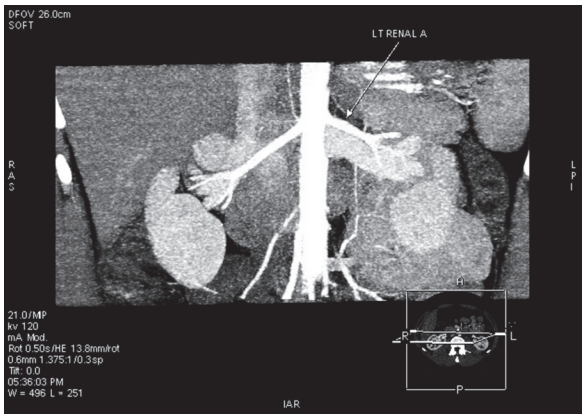

Figure 1: Computed Tomography Angiography (CTA) reconstruction image. The left renal vein appears dilated in her proximal part before crossing the aorta.

vein compression, b) to collect blood samples for Plasma Renin Activity (PRA) and aldosterone determination from both renal veins and the inferior vena cava and c) to measure the pressure gradient between the left renal vein and the inferior vena cava. A very mild stenosis and dilatation of the left renal vein without any signs of varices was recorded. There was a minimal pressure gradient (1 mmHg) between the left renal vein and the inferior vena cava. No further interventions were performed and the patient did not present any complications.

PRA and aldosterone blood levels from the left, the right renal vein and the inferior vena cava did not show any significant differences and were between normal limits. The overall laboratory investigation did not reveal any signs of secondary hypertension and the patient was discharged with the prescription of 300 mg of irbesartan and 12.5 mg of hydrochlorothiazide and 10 mg of amlodipine. After a follow-up of more than 12 months, his blood pressure remains well controlled and he does not present any abnormal laboratorial findings.

Discussion

The terms NCS and NCP are frequently used interchangeably in the literature referring to the known abnormal anatomy of the compression of the left renal vein, as it passes through the angle between the aorta and the superior mesenteric artery. Most symptomatic patients are usually adolescents or young adults[2-5].

Hypertension has not been included in the traditional clinical manifestations of the syndrome that is usually accompanied with abdominal pain, hematuria, orthostatic proteinuria, chronic fatigue and varicocele formation[2,4-10,12].

However, Hosotani et al have reported a case of NCP and hypertension with increased plasma renin activity (PRA) and aldosterone levels in the peripheral blood in a young Japanese woman[11]. The patient underwent selective renal venography that revealed a severe stenosis of the left renal vein and dilated ovarian veins. The authors also reported increased PRA levels in the left renal vein and an increased renocaval pressure gradient that was indicative of NCS. The patient became normotensive and PRA levels normalized after endovascular stent placement in the affected renal vein.

In our patient we were not able to identify any cause that might be related with secondary forms of hypertension. The accidental finding of left renal vein dilatation in our patient led us to investigate a possible relation with hypertension. We were not able to confirm the findings of Hosotani et al, but our patient had some significant differences. In our case, there was only a very mild compression and dilatation of the left renal vein with normal gonadal veins. In addition, the renocaval pressure gradient was also between normal limits (less than 3 mmHg).

NCS/NSP has been also reported concurrently with various clinical entities such as IgA nephropathy[13], membranous nephropathy, idiopathic hypercalciuria, Henoch-Schonlein purpura [2] or familiar mediterranean fever [14], but our patient had no evidence of any glomerular damage.

Treatment is usually conservative for the mild cases, but for more severe symptoms (persistent hematuria or pain) the interventions aim to decrease left renal vein hypertension. Multiple approaches by various surgical techniques, or even renal autotransplantation and endovascular stenting have been applied with rather acceptable results[15-17].

Some authors correctly state that any anatomy compatible with the nutcracker pathology is not always associated with clinical symptoms and these findings might represent just a normal variant[3,18]. Our case, where we were not able to establish any correlation of hypertension with the described nutcracker anatomy supports the above suggestions.

In conclusion, we report a case of radiological evidence of NCP in a hypertensive young adult. As there were no established pathogenetic links, hypertension could not be included in the clinical findings that may commonly accompany NCS and should be considered only as a coincidence.

Conflicts of Interest: None to declare

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