CASE REPORT

DOI: 10.4274/ijca.55264
Int J Cardiovasc Acad 2023;9(1):20-23

A Rare Cause of Secondary Hypertension: Nutcracker Syndrome

Gülay Uzun, Muhammet Raşit Sayın

Department of Cardiology, University of Health Sciences Turkey, Trabzon Ahi Evren Thoracic and Cardiovascular Surgery Training and Research Hospital, Trabzon, Turkey

Abstract

Left renal vein entrapment syndrome also known as nutcracker syndrome (NCS) is a vascular anomaly that occurs due to compression of the left renal vein from the outside, causing reduced left renal venous blood flow and thus venous congestion. This may be either asymptomatic or present with various clinical scenarios but is rarely associated with hypertension (HT). In this paper, we report an NCS case diagnosed in a young female patient who presented with HT.

Keywords: Nutcracker syndrome, hypertension, left renal vein

INTRODUCTION

Left renal vein (LRV) entrapment syndrome also known as nutcracker syndrome (NCS), is a vascular anomaly that occurs due to compression of the LRV from outside, causing reduced left renal venous blood flow and thus venous congestion. In this paper, we report an NCS case diagnosed in a young female patient who presented with hypertension (HT).

CASE REPORT

A 45-year-old female patient without a history of HT presented with intermittent high blood pressure readings and headache for the last 1 year. Physical examination showed blood pressure readings of 150/90 mmHg in both arms and a pulse rate of 80 bpm. The patient denied taking any herbal product, licorice or any regular medication, nor she had episodes of diarrhea and flushing. The body mass index was 23 kg/m^2. Physical examination of other systems was normal. Transthoracic echocardiographic examination revealed no pathological conditions except for left ventricular diastolic dysfunction. Routine biochemistry, complete blood count, and thyroid function tests were within normal limits. Urinalysis was normal. Ambulatory blood pressure measurement showed episodes of HT during daytime when the patient was in the erect position for long periods. Daytime mean blood pressure reading was 145/90 mmHg. Both kidneys had a normal size and parenchymal thickness on abdominal ultrasonography (USG). Renal Doppler USG showed that the LRV was compressed between the superior mesenteric artery and abdominal aorta. A computerized tomography angiogram showed that the LRV was compressed between the superomedial mesenteric artery and the aorta [Figures 1-3]. The narrowest segment of LRV having a diameter of 2.3 mm and immediately proximal to that point, the renal vein was dilated, and had a diameter of 12.2 mm [Figure 4]. The LRV diameter ratio was 5.3. The aorto-mesenteric angle was 18° [Figure 5]. In addition, there was dilation in the left gonadal vein. There were no other findings or laboratory parameters to suggest other causes of secondary HT in the patient. The patient was diagnosed with NCS. Target blood pressure could not be achieved by lifestyle modifications.

To cite this article: Uzun G, Sayın MR. A Rare Cause of Secondary Hypertension: Nutcracker Syndrome. Int J Cardiovasc Acad 2023;9(1):20-23

Address for Correspondence: MD. Gülay Uzun, Department of Cardiology, University of Health Sciences Turkey, Trabzon Ahi Evren Thoracic and Cardiovascular Surgery Training and Research Hospital, Trabzon, Turkey.
E-mail: drgulayhekimoglu@gmail.com
ORCID ID: orcid.org/0000-0002-7848-5840

Received: 07.02.2023
Revised: 28.03.2023
Accepted: 29.03.2023
Published Online: 14.04.2023

She was started benidipine 8 mg once per day. At the clinical follow-up, her blood pressure was regulated, and no additional problem was observed. Informed consent was obtained from the patient.

DISCUSSIONS

LRV entrapment syndrome is characterized by external anatomic compression of the renal vein causing marked dilation of the hilar portion and narrowing of the para-aortic portion of the latter, which results in altered flow dynamics in the inferior vena cava. When symptoms are absent despite the presence of anatomic hallmarks of the condition, it is called the nutcracker phenomenon. NCS refers to renal vein entrapment with well-known associated symptoms, the severity of which is dictated by the seriousness of anatomic compression and hemodynamic perturbation.[1] LRV is usually entrapped anteriorly, between the superior mesenteric artery and the aorta, or less commonly posteriorly, between the aorta and the vertebral column. Our patient had the more common anterior type.

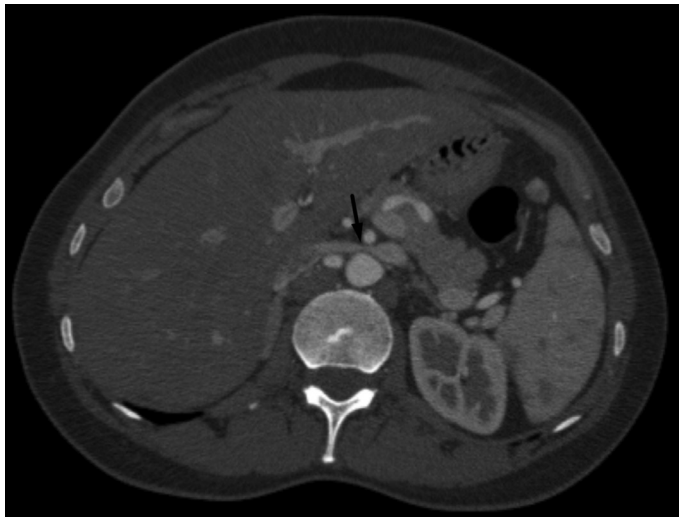

Figure 1: Axial plane of computerized tomographic angiography images of the left renal vein

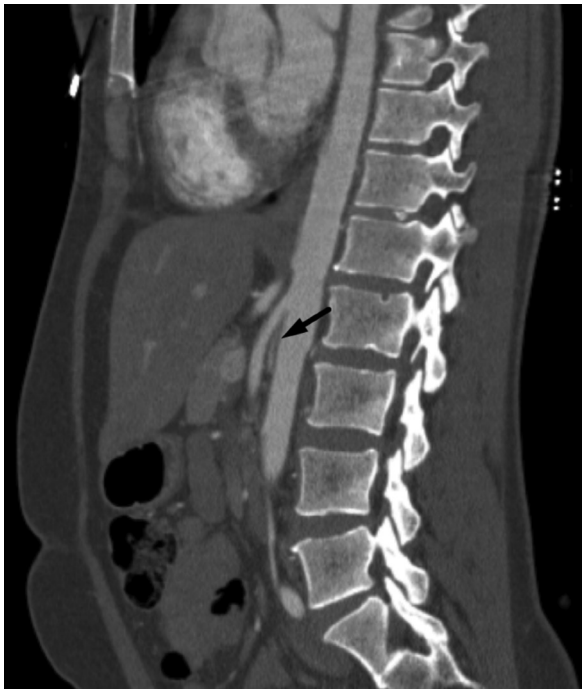

Figure 2: Sagittal plane of computerized tomographic angiography images of the left renal vein

The major clinical signs and symptoms of NCS include mild hematuria, orthostatic proteinuria, flank/abdominal pain, varicocele, and pelvic congestion syndrome.[1] All anatomic variations in the syndrome cause limited outflow in LRV with resultant left renal venous HT. The latter causes hematuria by

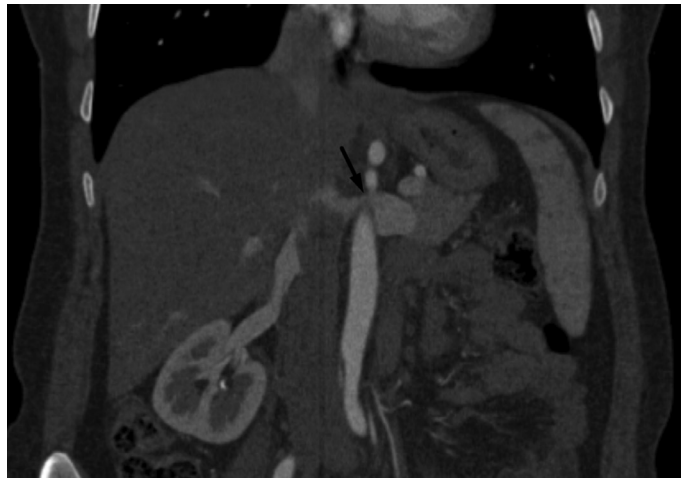

Figure 3: Coronal plane of computerized tomographic angiography images of the left renal vein

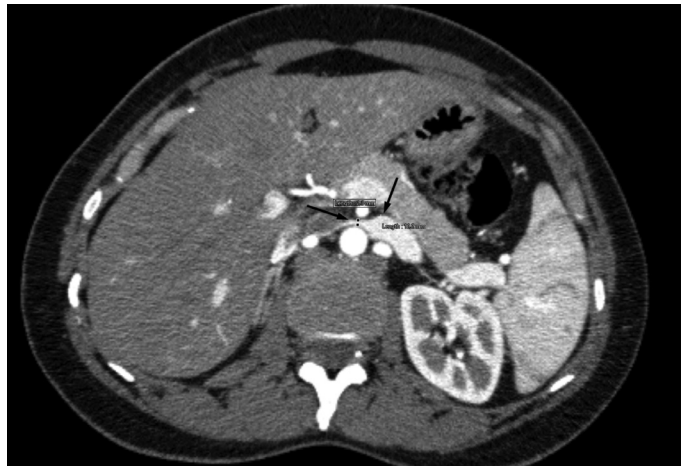

Figure 4: Left renal vein diameter ratio

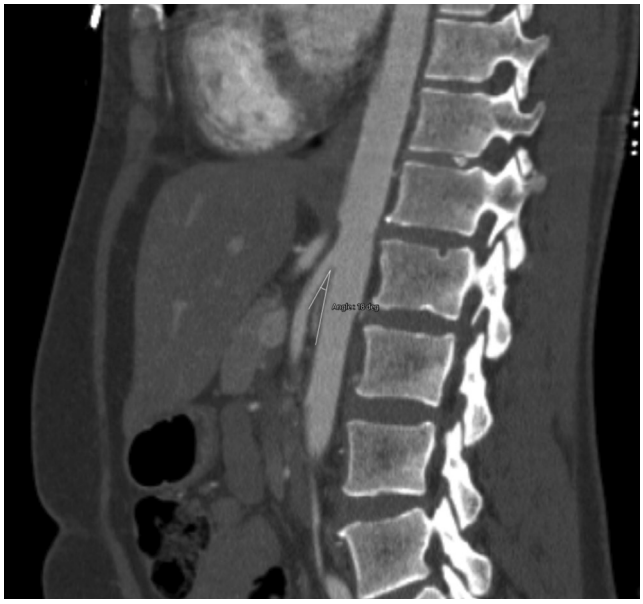

Figure 5: The aorto-mesenteric angle

disrupting the thin septum between small-caliber veins and the collecting system within the parenchyma of the kidney.[1] Patients with fully functional collateral circulation reducing LVR may explain the absence of symptoms. The left gonadal vein draining into the LRV in men may be congested and cause varicocele while women may suffer pelvic congestion syndrome characterized by dysmenorrhea, dysuria, dyspareunia, and pelvic pain.[1]

The syndrome excludes HT as a hallmark sign, which has been reported only rarely. There are several hypotheses why HT develops in this syndrome. These include: 1. Impaired sodium excretion ability of the kidney owing to elevated glomerular pressure resulting from the impedance to blood flow through the LRV; 2. Kidney ischemia and hypoxia secondary to elevated LRV pressure, 3. Renal chemoreceptors and baroreceptors respond to altered glomerular hemodynamic and metabolic milieu and send afferent impulses to the hypothalamus to increase the production and release of norepinephrine, a potent vasoconstrictor.[2,3] Of particular note, our patient presented with an uncommon sign of NCS, i.e, persistently elevated blood pressure and associated headache. As evidenced by normal serum, urinary biochemical tests, as well as imaging studies, she was free of any underlying etiology causing secondary HT such as renal parenchymal disease, hypercortisolism, or adrenal cortical or medullary tumors. She was thus deemed to have no identifiable cause of HT. As a matter of the fact that her LRV was compressed, we made a provisional diagnosis of NCS-associated HT. In the literature, different clinical presentations and treatment modalities exist for NCS cases accompanied by HT. In two reports, NCS was accompanied by HT in two young patients.[4,5] In both these cases, a significant increase was found in blood renin activity, aldosterone concentration, and angiotensin I/II concentrations. They had persistently elevated blood pressure readings despite medical therapy. The patients became normotensive immediately after they received an endovascular stent. In contrast, another case of NCS presented with medically controllable HT despite elevated renin level.[6] Blood pressure control was satisfactorily achieved by medications in the other NCS cases reported in the literature, who had no increased hormone levels.[7-9] Our patient also had normal biochemical parameters. Blood pressure was successfully controlled by medical therapy. Headache was attributed to HT since it improved with blood pressure regulation. However, it should be remembered that NCS associated with headache has also been reported in the literature.[10]

The way a patient with the condition presents and the degree of left renal venous HT usually dictate the management principles. While conservative management is usually employed for mild cases, decompressing the LRV is the main aim in severe cases presenting with persistent hematuria, severe pain, or pelvic congestion syndrome. Satisfactory results have been attained by a variety of treatment modalities including surgical techniques, even renal autotransplantation as well as endovascular stenting to relieve compression. Also in our patient, both blood pressure were brought under control with medical therapy and headache did not recur. Interventional treatments may be considered in cases resistant to medical therapy. NCS may be a difficult-to-diagnose syndrome due to various factors. Decisions regarding treatment should be made on the basis of symptom severity, the reversibility of the condition, patient's age, and disease stage.

REFERENCES

1. Granata A, Distefano G, Sturiale A, Figuera M, Foti PV, Palmucci S, *et al*. From Nutcracker Phenomenon to Nutcracker Syndrome: A Pictorial Review. Diagnostics (Basel) 2021;11:101.
2. Burnett JC Jr, Haas JA, Knox FG. Segmental analysis of sodium reabsorption during renal vein constriction. Am J Physiol 1982;243:F19-22.
3. Abildgaard U, Amtorp O, Agerskov K, Sjøntoft E, Christensen NJ, Henriksen O. Renal vascular adjustments to partial renal venous obstruction in dog kidney. Circ Res 1987;61:194-202.
4. Hosotani Y, Kiyomoto H, Fujioka H, Norihiro Takahashi, Masakazu Kohno. The nutcracker phenomenon accompanied by renin-dependent hypertension. Am J Med 2003;114:617-8.
5. Wang RF, Zhou CZ, Fu YQ, Lv WF. Nutcracker syndrome accompanied by hypertension: a case report and literature review. J Int Med Res 2021;49:300060520985733.
6. Park SJ, Kim SM, Won JH, Lim HS. A case of secondary hypertension associated with the nutcracker phenomenon. Korean Circ J 2014;44:434-6.
7. Mazarakis A, Almpanis G, Tragotsalou N, Karnabatidis D, Fourtounas C. Is hypertension a manifestation of the nutcracker phenomenon/syndrome? Case report and brief review of the literature. Hippokratia 2012;16:187-9.

8. Narkhede NA, Deokar AB, Mehta KP, Kamat NN. Nutcracker syndrome with hypertension as an unusual initial presentation. Indian J Nephrol 2017;27:472-4.

9. Azhar AB, Zeb NT, Shah S, Khalid A. Nutcracker syndrome with hypertension: a case report. Cureus 2019;11:e4781.

10. Scholbach T. From the nutcracker-phenomenon of the left renal vein to the midline congestion syndrome as a cause of migraine, headache, back and abdominal pain and functional disorders of pelvic organs. Med Hypotheses 2007;68:1318-27.