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Case Report

Refractory hypertension and pelvic pain associated with nutcracker phenomenon

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ARTICLE INFO

Article history:

Received 14 October 2019

Revised 12 January 2020

Accepted 25 January 2020

Keywords:

Nutcracker phenomenon

Refractory hypertension

CT angiography

ABSTRACT

We represent a 35-year-old woman, with refractory hypertension and pelvic pain, and with nutcracker phenomenon diagnosed using computed tomographic angiography. Although surgical treatment was not performed because of the patients' nonconsenting, any other cause for the hypertension was not found by extensive work-up, supports the notion that it was either secondary to nutcracker phenomenon or idiopathic.

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Introduction

Compressed left renal vein (LRV), most commonly between the aorta and the superior mesenteric artery, leading to impaired outflow into the inferior vena cava has been defined as nutcracker phenomenon (NCP) [1,2]. If this phenomenon becomes clinically significant, it is called nutcracker syndrome. The most common clinical presentations are hematuria, pain, and varicoceles but owing to these nonspecific manifestations, the diagnosis of NCS is often problematic and is usually postponed [3,4]. Therefore, often multiple investigations and procedures are practiced and even in some cases, no cause is identified unless rare pathologic conditions are suspected. Renin-dependent systemic hypertension due to renal venous entrapment occurring in NCS has previously reported [5]. We describe a case of a 35-year-old woman with abdominal

pain and refractory hypertension independent of the renin-angiotensin system whose NCP was diagnosed by computed tomographic angiography (CTA).

Case presentation

A 35-year-old woman presenting with a history of 4-year hypertension was referred to our hospital for further evaluation. The patient also had a history of mild intermittent abdominal pain in hypogastric region for the past 10 months. She would receive antihypertensive drugs (50 mg spironolactone daily and 5 mg amlodipine at the time of increasing the severity of blood pressure [BP]). The etiology of hypertension had not been identified at that time and the patient was referred to the urologist because of uncontrolled hypertension. The

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<https://doi.org/10.1016/j.radcr.2020.01.040>

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patient had no family history of hypertension and there was no abdominal tenderness in the physical examination. The patient was not pregnant and there was no history of abdominal or pelvic surgery. Also, arterial BP was 150/90 mm Hg. In laboratory examination, thyroid stimulating hormone [TSH] and potassium were within normal limits. In 24-hour urine collection, no proteinuria or hematuria was detected and metanephrine, normetanephrine, and vanillmandelic acid [VMA] levels were reported as within normal limits. Cushing disease evaluating tests such as blood cortisol and aldosterone (in upright and supine position) were found to be normal.

Plasma renin level was lower than normal limit, appearing to be 0.8 mIU/ml in upright position (normal range: 4.4-46.1) and 1.9 mIU /ml in supine position (normal range 2.8-39.9). Then comprehensive cardiovascular evaluations consisting of chest X Ray [CXR], echocardiography, and CTA of the thoracic aorta were done and proved to be normal.

Patient was referred to Farshchian cardiovascular hospital for radiological evaluations including ultrasound of the kidneys as well as color Doppler ultrasonography to rule out renal artery stenosis, as one of the causes of secondary hypertension. Being evaluated by ultrasound, the size, shape, parenchymal echogenicity, and corticomedullary differentiation of both kidneys were normal. There were 2 small stones 2 mm and 4.5 mm in diameter in the upper calyx of the left kidney. Doppler ultrasound showed intrarenal artery resistive index values of 0.55 on the right side and 0.56 on the left side. Acceleration time was measured as 53 milliseconds in the right side and 40 milliseconds in the left side. These values and renal/aortic ratio were found to be in the normal ranges. After gaining no result in color Doppler ultrasound, the patient was taken to CT angiography for more evaluations. Abdomino pelvic CT angiography, with intravenous administration of 90 ml of nonionic water soluble contrast media (visipaque 320 mg/ml), using low dose 128-slice multidetector computed tomography [MDCT] scanner (Siemens, SOMATOM Definition AS, Germany), was performed.

After manual selection of the field of view, data was reconstructed, keeping slice thickness 5 mm and reconstructed increment: 0.6 mm in dedicated soft tissue kernel setting and it was analyzed in axial, sagittal, and coronal views. Multiplanar reformation, maximum intensity projection, and volume rendering images were reconstructed. The following findings were discerned in the CT angiography of the renal arteries: Both main renal arteries at ostium, main, and segmental parts appeared to be normal without evidence of stenosis, aneurysm, arteriovenous malformation [AVM], arteriovenous fistula [AVF], or abnormal tumor enhancement (Fig. 1). Small caliber left accessory renal artery originating from the abdominal aorta supplying upper pole of the left kidney was seen and proved to be without obvious stenosis (Fig. 2). In sagittal images, aortic-SMA angle was about 16° (Fig. 3A and B) which is lower than normal range. Two stones 2 mm and 4.5 mm in diameter were seen in the upper calyx of the left kidney. Then, delay images were provided for the evaluation of the renal veins and the findings were as follow:

Left gonadal vein dilatation as well as enlarged and tortuous venous structure in the left side of pelvis was seen (Fig. 4). LRV compression between abdominal aorta and superior

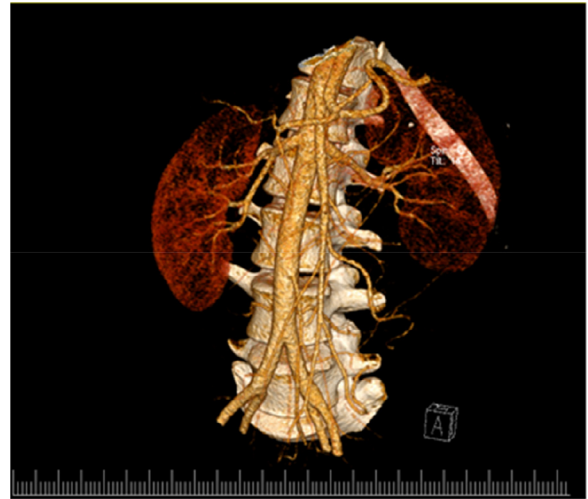


Fig. 1 – Volume-rendered image from renal arteries CTA shows normal caliber without stenosis in the both main renal arteries. CTA, computed tomographic angiography.

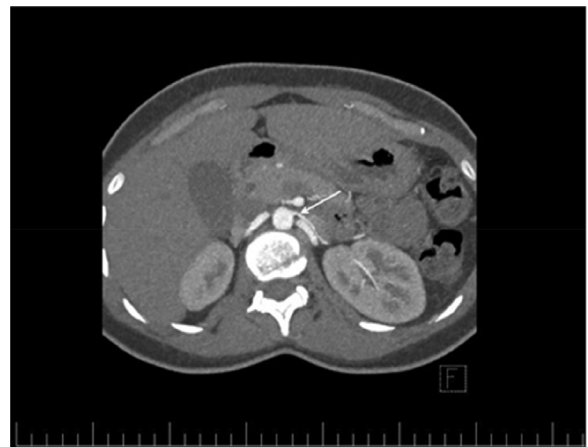


Fig. 2 – Axial maximum intensity projection [MIP] in arterial phase shows small caliber left accessory renal artery, originating from the abdominal aorta and supplying upper pole of the left kidney without obvious stenosis.

mesenteric artery is seen. Compression ratio ($CR = P-C/C$ Diameter of pre-recompressed vein (P) minus diameter of compressed vein (C), divided by the diameter of the compressed vein (C), was 3.4. (Fig. 5). Both adrenal glands show normal size and attenuation and also there was no evidence of superior mesenteric artery [SMA] syndrome on CTA.

In summary, imaging findings reveal normal renal arteries and NCP with left gonadal vein enlargement and pelvic veins congestion. According to imaging findings and other laboratory data, and considering that there were no other causes for refractory hypertension, nutcracker syndrome is likely to be the cause of hypertension and abdominal pain. Since secondary hypertension had been controlled by renal vein transposition [RVT] in a similar case reported by Narkhede et al [6] and with respect to responsible urolo-

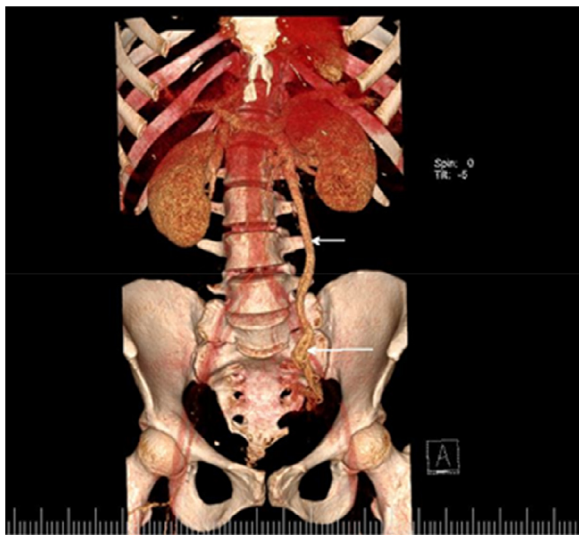


Fig. 3 – Volume-rendered image from renal vein CTA reveals left gonadal vein dilatation (short arrow) as well as enlarged and tortuous venous structure (long arrow) in the left side of pelvis. CTA, computed tomographic angiography.

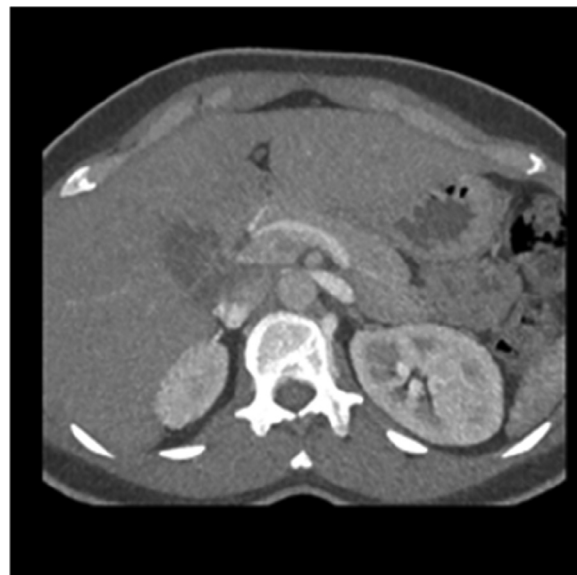


Fig. 5 – Axial MIP view from renal veins CTA shows compression of left renal vein between abdominal aorta and SMA. CTA, computed tomographic angiography.

gist' point of view, the patient was recommended to go under kidney autotransplantation surgery. But the patient did not give consent for surgery and preferred to be followed conservatively. Being closely observed by cardiovascular specialist, the patient started a new set of antihypertensive medications but optimal BP target was not reached in a 3 months period of follow-up.

Discussion

NCP is considered as the LRV anatomic compression between the SMA and aorta (anterior nutcracker) or between aorta and underlying vertebral body (posterior nutcracker). When the increased LRV pressure leads to symptoms, it is called nutcracker syndrome [2] and viewed as more com-

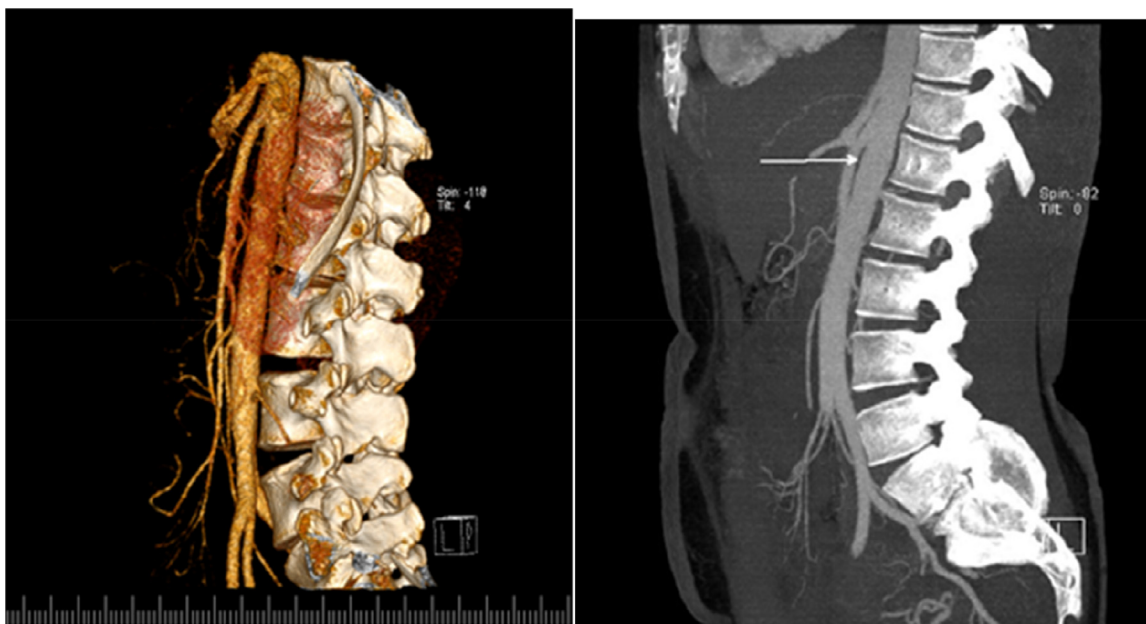


Fig. 4 – (A) Sagittal MIP view and (B), volume-rendered image from abdominal aorta CTA show decreased aortic-SMA angle. CTA, computed tomographic angiography.

mon in women [7]. Nutcracker syndrome symptoms include hematuria (most common), flank pain, abdominal pain, pelvic venous congestion in females, and varicoceles in males. hypertension [HTN] is thought to be a rare symptom of the nutcracker syndrome [8]. In 2014, Park et al reported a case of nutcracker syndrome and hypertension with increased plasma renin activity in the LRV, almost 5 times higher than that in the right renal vein [5]. However, in 2017, Narkhede et al described a young female with hypertension and nutcracker syndrome whose blood level of renin was within normal limits [6]. The plasma rennin level in the laboratory data of the patient chosen for this study was less than normal so possible relationship between nutcracker syndrome and increased plasma renin activity was not supported by the work-up. Diagnostic steps in suspected cases cover laboratory tests, ultrasonography, CTA, and magnetic resonance angiography [MRA] of abdomen and venography with renocaval pressure gradient measurement [1]. Color Doppler ultrasonography should be used for evaluation of the collateral vessels decreasing the blood flow through the narrowed portion of the LRV [1]. Peak systolic velocity at the point of renal vein compression to peak systolic velocity in the hilar renal vein over 4.7 shows high sensitivity and specificity for the diagnosis. CT or MR imaging with intravenous [IV] contrast material reveals focal compression of the LRV, dilatation of prestenotic region, renal hilar and pelvic varices, and dilated gonadal veins [2]. The case chosen for this study showed left gonadal vein dilatation as well as enlarged and tortuous venous structure in the left side of pelvis. CTA or MRA in sagittal reconstruction suggests a small gap between the SMA and anterior wall of the aorta to be less than 5 mm and aortomesenteric angle less than 28° [1,2]. In this case, aortomesenteric angle was 16°, acceptable for the NCP diagnosis. Nutcracker syndrome surgical treatment methods include endovascular intervention (LRV

transposition is less complicated method), renal autotransplantation to the iliac fossa and nephrectomy [2,9]. Owing to severe pelvic vein congestion which had made pelvic pain and also physician trying to control hypertension, the patient was advised to be performed kidney autotransplantation surgery but the patient chose medical treatment strategies.

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