Nutcracker Syndrome Accompanying Pelvic Congestion Syndrome: Color Doppler Sonography and Multislice CT Findings: A Case Report

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Nutcracker syndrome (NCS) is a rare pathology, caused by compression of the left renal vein (LRV) between the abdominal aorta (AA) and the superior mesenteric artery (SMA), due to reduction of the angle between AA and SMA. This leads to LRV varices, left gonadal vein varices and therefore, the pelvic congestion syndrome. For this reason, coexistence of NCS and pelvic congestion syndrome has been described. It manifests by hematuria, proteinuria, and nonspecific pelvic pain secondary to pelvic congestion, dyspareunia and persistent genital arousal. We report a 27-year-old woman who experienced hematuria and left flank pain. The diagnosis of NCS accompanied by pelvic congestion syndrome was missed initially, but later on the diagnosis was made by color Doppler ultrasound, abdominal computed tomography (CT) and CT angiography that were later performed. She refused interventional and surgical treatments, and was lost to follow up.

Keywords: Hematuria; Renal Nutcracker Syndrome; Flank Pain

1. Introduction

The nutcracker syndrome (NCS) is compression of the left renal vein (LRV) by the abdominal aorta (AA) and the superior mesenteric artery (SMA), manifested by pain, hematuria, varicocele and lower urinary tract symptoms. NCS accompanying pelvic congestion syndrome is a rare pathology that can be easily missed by routine diagnostic methods.

2. Case Presentation

A 27-year-old woman was brought to the emergency department because of left flank pain, chronic pelvic pain and hematuria. She had genital complaints. She had no fever, vomiting, or diarrhea. She also denied any trauma to the abdomen or back. Her medical history was otherwise unremarkable. She had left flank tenderness. At that time, urinalysis showed numerous red blood cells. Hematuria persisted on multiple urinalyses.

Color Doppler ultrasound (CDUS) and computed tomography (CT) images showed reduction of the angle between the AA and SMA. The angle between the AA and SMA was 14.5°. Pelvic ultrasonography (US), CDUS and CT images showed multiple pelvic varices including tortuous pelvic veins, dilated arcuate veins in the myometrium communicated with pelvic varicose veins (Figure 1). It was confirmed by CT that demonstrated compression of the left renal vein between the AA and SMA and also dilatation of the left ovarian vein attributable to NCS in axial and coronal oblique images. Axial CT reformatted oblique coronal images and volume-rendered multidetector CT (MDCT) angiography images showed retrograde filling of the left ovarian vein in the arterial phase consistent with reflux in pelvic congestion syndrome (Figure 2). The diagnosis of NCS accompanying pelvic congestion syndrome was established by these findings. With the consent of the patient and her parents, conservative treatment was decided upon.

3. Discussion

NCS is a rare, but treatable clinical condition (1). Because of the variability of symptoms and absence of consensus on diagnostic criteria, the exact prevalence of NCS is unknown, but it may be slightly higher in females (2). NCS is a well-known, but under-recognized cause of hematuria. The LRV normally crosses between the AA and SMA, where it may be compressed, resulting in a variable degree of LRV obstruction. This compression is called NCS (1). The normal angle between the SMA and the abdominal

Implication for health policy/practice/research/medical education:
Nutcracker syndrome accompanying pelvic congestion syndrome is a rare, but treatable clinical condition. Inclusion of this entity in the differential diagnosis of patients with an unknown cause of hematuria and flank pain can achieve efficient management of this condition. In addition, in our case, the angle between the aorta and the superior mesenteric artery was approximately 14.5 degrees, which is almost the lowest angle in comparison with previous studies and reported cases in the literature.

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Figure 1. A 27-year-old woman with NCS and pelvic congestion syndrome. A) CDUS and (B, C) sagittal CT images show reduction of the angle between AA and SMA (14.5°) (arrows). D) Pelvic CDUS shows pelvic varicose veins (arrows).

Figure 2. A) Axial CT image shows narrowing of the left renal vein between AA and SMA (beak sign) (arrow). B) Axial CT and C) Volume-rendered angiography images show retrograde filling of the left ovarian vein in the arterial phase consistent with reflux in pelvic congestion syndrome (arrows).
Pelvic congestion syndrome occurs mostly because of ovarian vein reflux, but it can also occur because of the obstruction of ovarian vein outflow resulting in reversed flow (8, 9).

In the differential diagnosis of NCS and accompanying pelvic congestion syndrome, pathologies that cause pain and hematuria such as lithiasis, congenital vascular malformations, tumors, infections, parenchymal or urinary tract abnormalities, painful pelvic syndromes and organ or neighboring structure alterations must be excluded (6, 10). Supportive treatment is adequate if the symptoms are mild in NCS. Conservative treatment has been suggested for mild hematuria. Surgical or radiological interventions are indicated for severe pain, significant hematuria and renal functional impairment. Endovascular intervention is now considered the first line of therapy for management of this condition (1, 13, 14).

NCS accompanying pelvic congestion syndrome is a rare pathology that can be easily missed by routine diagnostic methods. It should be considered in the differential diagnosis of patients with hematuria, intermittent flank pain, nonspecific pelvic pain, dyspareunia, and persistent genital arousal without an apparent cause. An inclusion of this entity in the differential diagnosis of patients with an unknown cause of hematuria and flank pain can achieve efficient management for this condition.

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Authors’ Contribution
Mikail Inal and M. Yasemin Karadeniz Bilgili diagnosed the patient and prepared the manuscript. Safa Sahin helped in the diagnosis.

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References
1. Waseem M, Upadhyay R, Prosper G. The nutcracker syndrome: an underrecognized cause of hematuria. Eur J Pediatr. 2012;171(6):1269–71.
2. Kurklinsky AK, Rooke TW. Nutcracker phenomenon and nutcracker syndrome. Mayo Clin Proc. 2010;85(6):552–9.
3. Ahmed K, Sampath R, Khan MS. Current trends in the diagnosis and management of renal nutcracker syndrome: a review. Eur J Vasc Endovasc Surg. 2006;31(4):410–6.
4. Kim KW, Cho JY, Kim SH, Yoon JH, Kim DS, Chung JW, et al. Diagnostic value of computed tomographic findings of nutcracker syndrome: correlation with renal venography and renocaval pressure gradients. Eur J Radiol. 2011;80(3):648–54.
5. Fu WJ, Hong BF, Gao JP, Xiao YY, Yang Y, Cai W, et al. Nutcracker phenomenon: a new diagnostic method of multislice computed tomography angiography. Int J Urol. 2006;13(1):72–80.
6. Dellavedova T, Racca L, Ponzano RM, Minuzzi S, Olmedo JJ, Minuzzi G. Nutcracker syndrome: a case report. Rev Mex Urol. 2010;70(1):53–4.
7. Belenky A, Bartal G, Atar E, Cohen M, Bachar GN. Ovarian varices in healthy female kidney donors: incidence, morbidity, and clinical outcome. AJR Am J Roentgenol. 2002;179(3):625–7.
8. Karcaaltincaba M, Karcaaltincaba D, Dogra VS. Pelvic Congestion Syndrome. Ultrasound Clinics. 2008;3(1):415-25.
9. Cicchiello LA, Hamper UM, Scoutt LM. Ultrasound Evaluation of Gynecologic Causes of Pelvic Pain. Ultrasound Clinics. 2010;5(2):209-31.
10. Mohammadi A, Ghasemi-Rad M, Mladkova N, Masudi S. Varicocele and nutcracker syndrome: sonographic findings. J Ultrasound Med. 2010;29(8):1153-60.
11. Muraoka N, Sakai T, Kimura H, Uematsu H, Tanase K, Yokoyama O, et al. Rare causes of hematuria associated with various vascular diseases involving the upper urinary tract. Radiographics. 2008;28(3):855-67.
12. Karaosmanoglu D, Karcaaltincaba M, Karcaaltincaba D, Akata D, Ozmen M. MDCT of the ovarian vein: normal anatomy and pathology. AJR Am J Roentgenol. 2009;192(1):295-9.
13. Chen YM, Wang IK, Ng KK, Huang CC. Nutcracker syndrome: an overlooked cause of hematuria. Chang Gung Med J. 2002;25(10):700-5.
14. Chang CT, Hung CC, Ng KK, Yen TH. Nutcracker syndrome and left unilateral haematuria. Nephrol Dial Transplant. 2005;20(2):460-1.