Case Report

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Anterior Nutcracker syndrome in a young male patient: a case report and review of literature

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Introduction and importance: The left renal vein (LRV) is affected by a venous compression syndrome called Nutcracker syndrome (NCS). This syndrome is characterized by extrinsic compression of the LRV, which usually occurs between aorta and superior mesenteric artery. It is a rare and under-diagnosed condition, more prevalent in females and that, if left untreated, can lead to severe problems. There are no clear guidelines regarding management. Therefore, the authors report this rare case and its symptoms in male patient and they display current management options.

Case presentation: NCS was observed during computer tomography in a male patient presented with persistent left flank pain and associated haematuria. Ultrasound for left scrotum demonstrated left moderate-sized varicocele. The left varicocele testis unit was 1.6 mm and during the Valsalva manoeuvre in the supine position the testis unit was 2 mm. LRV compression between abdominal aorta and superior mesenteric artery was identified by computer tomography imaging and therefore, diagnosis of NCS was confirmed.

Clinical discussion: The actual prevalence is unclear, and incidence rates have been observed to fluctuate among age group and more prevalent in women. Main symptoms include haematuria, left flank discomfort, varicocele in men, proteinuria and anaemia. Depending on severity of symptoms, management might range from conservative care to surgery.

Conclusion: This treatment strategy was effective in reducing the symptoms of the patients. In young patients, conservative treatment is advised for a fair amount of time. However, more studies on how much the authors should wait before considering surgery is important.

Keywords: computed tomography, left renal vein, vein entrapment syndrome, case report, nutcracker syndrome.

Introduction

The Nutcracker syndrome (NCS) usually happens when left renal vein (LRV) becomes wedged because of extrinsic compression, which alters blood outflow to inferior vena cava[1]. It is called, nutcracker phenomenon when LRV is compressed between normal anatomical structures[2], commonly between the aorta (AO) and superior mesenteric artery (SMA)[3]. Some studies suggest that despite the presence of associated anatomical findings that accompany the syndrome, it is not necessary to cause clinical symptoms[4]. This syndrome may occur at any age and it may be most common in women. Also, it is difficult to know the actual prevalence because of the wide spectrum of different symptoms, and lack of accurate agreed diagnostic criteria[5]. The syndrome cannot be considered as a hereditary disease, although it occurs in siblings[6]. The symptoms are different and it may be asymptomatic[6]. Typical symptoms are: left flank pain, haematuria, left varicocele[7] and proteinuria (particularly orthostatic proteinuria)[3]. The diagnosis is based on symptoms and radiologic findings. Many methods have been adopted such as Doppler ultrasound, computed tomography (CT), MRI, angiography[8]. The management in mild cases based on watchful waiting and conservative approaches, and in severe cases or in case of failure of conservative approaches include a variety of surgical techniques[9,10]. Nowadays many surgical managements are based on endovascular procedures with balloon angioplasty or stent[11]. This case report has been reported in line with the SCARE Criteria[12] and has Research Registry unique identifying number (UIN) of researchregistry9243.

HIGHLIGHTS

- Male.
- Varicocele.
- Conservative treatment is successful.

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Case presentation

A smoker non-alcoholic male patient (23-year-old) self-presented to emergency department with complaint of persistent left flank pain associated with intermittent macroscopic haematuria. On presentation, the patient's BMI was 23.5. The patient has no surgical or medical history of chronic, allergic diseases, drug usage and no familial or genetic history.

The physical examination showed normal vital signs, and he was not passing clots through the urethra. Organomegaly, peritonism or palpable abdominal mass were not seen. Moreover, the results of the cardiovascular and respiratory tests were unremarkable.

Examination

The patient noticed that the pain has begun about 2 months earlier and both of the two symptoms worsened by heavy exertion. The pain was moderate, began gradually, and did not radiate or worsened with position changes but increased with cough and effort. His pain was accompanied with moderate to severe headaches. His blood pressure on admission was 170/100 110 mmHg and reached 220/110 mmHg. Nitroglycerin patch was used to reduce hypertension with very shallow effect in reducing blood pressure. The values fluctuated and were significantly lower as the pain reduced later on. His visual analog scale score was 4. Ophthalmoscopy was performed and showed no abnormal finding.

The patient denied any history of postprandial pain, vomiting, nausea or any other signs or symptoms including fever, bleeding tendency, poor stream, frequency urgency and burning sensation upon micturition. He confirmed no recent trauma. His medical history showed five attacks of colic pain, which followed weightlifting exercises. No history of recurrent urinary tract infections or urinary symptoms and no family history of kidney diseases or hypertension was seen.

Severe tests including laboratory tests and radiological image was requested and performed by multi-disciplinary team of general practitioner, general and vascular surgeon, physicians specialized in heart and kidney diseases.

Lab tests was confirmed the following results: Serum haemoglobin of 14.8g/dl, Haematocrit of 40.6%, kidney function tests including urea and creatinine were normal, Urine analysis showed (30–35) red blood cell, (18–20) white blood cell and (++) blood. His thyroid function, cortisol concentration, and adrenocorticotropic hormone concentration were all in the normal range.

The radiological evaluations started with ultrasound for the urinary system and it did not show abnormalities. Ultrasound for the left scrotum demonstrated left moderate-sized varicocele. Bilateral measurement for diameters of pampiniform plexus were carried out during resting and Valsalva manoeuvre in the supine position. The left varicocele testis unit was 1.6 mm and during Valsalva manoeuvre in supine position the testis unit was found 2 mm. There was no varicocele on the right side.

The patient was further evaluated with CT imaging of abdomen and pelvis with IV contrast. The computed tomography revealed compression of LRV between abdominal AO and SMA. Therefore, the diagnosis of NCS was confirmed.

Follow-up and results from CT

CT of pelvis and abdomen with injections was performed on patient. No overt hepatic masses were observed. Compression of RLV between SMA and abdominal AO causing stenosis was noted. Furthermore, no adrenal or pancreatic masses were seen. Also, omental or peritoneal thickening, abdominal or pelvic ascites, were unmarked or absent. However, medium costochondrosis was observed with rest of the findings in normal range. The patient was admitted in emergency and was followed up at regular intervals. Following are the contrast-enhanced CT images performed on the respective patient (Figure 1).

Treatment

Since the patient is young and in overall good health and Looking at the severity of symptoms, in addition considering that conservative therapy allows for a trial period to assess the patient's response to treatment and symptom improvement without undergoing the risks and potential complications associated with surgery. Treatment was decided to be Rivaroxaban, 10 mg once daily. Ibuprofen (nonsteroidal anti-inflammatory drugs) 400 mg in addition to anti-spasmodic medication was prescribe during the pain attack. Blood pressure was tracked on regular basis to prescribe medication when needed. Patient also got the psychological support form a psychologist to help him process his condition.

Patient perspective and outcomes

The patient is a well-educated individual, studying engineering. He presented to the Emergency room with left flank pain associated with intermittent haematuria. He was scared to see blood in the urine especially and worried about how this condition would affect his life. Phisicians fully explained the case to the patient and he was very understanding and embrace the condition. He understood the treatment options, and prefered to go first on conservative approah. He followed the treatment plan and reported relief of symptoms, improvement of overall health, attacks are less frequent now, Blood pressure is at the normal range and the patient is following up on his condition constantly to detect any changes to modify treatment accordingly.

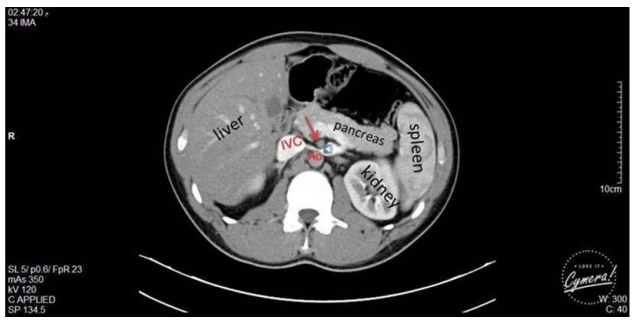

Figure 1. Abdominal with-Contrast computed tomography (CT) scan—Transverse section: shows the inferior vena cava (IVC), aorta, superior mesenteric artery (SMA)—red arrow, The left renal vein (LRV)—white arrow. Nutcracker syndrome is diagnosed as the CT shows the compression of the LRV between the aorta and the SMA.

Discussion

NCS is a kind of venous compression syndrome that affects LRV. It is an uncommon, under-diagnosed, disorder that can cause severe complications, if not treated. The aim of the discussion is to include a review of literature. The following keywords were used "Nutcracker Syndrome", OR "Vein entrapment syndrome" in PubMed and Google Scholar. We scanned the results and included results that cover a range of treatment options, including conservative care, medical treatments, compression socks, weight gain, endovascular treatment, open surgery and regular follow-up.

This disorder is also called as left renal 'vein entrapment syndrome' because it is characterized by extrinsic compression of the LRV, which usually occurs between AO and SMA[13]. Although, variations in anatomy may exist and the condition may also go undiagnosed and untreated[14]. This syndrome causes venous congestion and various other symptoms[15] by restricting flow via the LRV[13]. "Nutcracker phenomenon" defines aorto-mesenteric compression of the LRV while asymptomatic, whereas 'Nutcracker syndrome' identifies individuals with distinctive symptoms linked to Nutcracker anatomical morphology[13].

NCS is regarded as an uncommon disorder, while the actual prevalence is unclear, and incidence rates have been observed to fluctuate among age groups[16] including cases ranging from infancy to the seventh decade[17,18]. It is reported more commonly in females in third to fourth decade, while later research found equal frequency in females and males[14,18].

The most prevalent type is anterior NCS, in which LRV compression is seen between AO and SMA. However, the most prevalent cause of posterior NCS is retro aortic or circumaortic LRV compression between AO and spinal column, which is rare[13,16]. Regarding this, various other variants have been described also. They are right-sided NCS, LRV duplication, LRV compression by dilated left inferior vena cava, and LRV compression between SMA and right renal artery[16]. The fundamental mechanism of collateralization and varices is 'venous compression', which leads to outflow blockage and causes LRV hypertension[6,13]. The left gonadal vein and connecting lumbar vein are principally involved in collateralization of venous circulation[14,19].

Majorly the symptoms of the syndrome include haematuria as observed in our patient, left flank pain, varicocele in males, proteinuria and anaemia. Haematuria is the major symptom of this syndrome occurring in 80% of the patients and it is a condition which arises when thin-walled septum separating smaller veins from renal collecting system is disrupted by high blood pressure in collaterals and subsequent venous sinuses close to renal calyces[13,16]. Patients with properly developed collateral circulation that lowers LRV hypertension may not experience renal symptoms[16]. Varicocele or pelvic congestion syndrome may result from venous congestion adjacent to LRV compression affecting left gonadal vein. Infertility, abdominal varicose veins, vaginal wall, buttocks or upper thighs, dyspareunia, dysmenorrhoea, orthostatic hypotension, tiredness and stomach discomfort are some additional symptoms seen in patients experiencing this syndrome[13,15,20].

Imaging is used to confirm diagnosis of NCS followed by using approaches such as history and physical examination. CT angiography, Doppler ultrasound, intravascular ultrasound, MRI, venography are all included in the step-by-step process of the diagnosis of NCS. We have first used ultrasound and later CT for our patient which eventually diagnosed the syndrome. Sometimes being invasive and unneeded for diagnosis, venography along with the measurement of the renocaval pressure gradient is considered as gold standard method for the diagnosis of NCS[17]. Recent years have seen a rise in the recommendation of CT scans due to its accuracy and capacity to examine abdominal abnormalities. The "beak sign" and diameter ratio (hilar-aorto-mesenteric) of greater than or equal to 4.9 are most precise CT metrics[18,21]. The choice between Doppler ultrasound and CT should be made after taking into account each patient's circumstances, expense, radiation exposure, and accessibility, a contrast allergy and additional abdominal diagnostic options[17]. Orthostatic proteinuria or microscopic haematuria may be detected during a urine analysis[20]. Patients with persistent or substantial haematuria may experience anaemia[6,18].

Management of this syndrome has been controversial. Due to limited availability of data, there are presently no clear treatment guidelines. Depending on severity of symptoms, management might range from conservative care to nephrectomy. The potential for spontaneous resolution following the development of adipose tissue and the presence of many venous collateralizations, prolonged conservative therapy is strongly advised for the paediatric population group. Orthostatic proteinuria has been thought to be improved by medical treatments like low dose aspirin and angiotensin converting enzyme inhibitors, notably alacepril[6,18,22]. Compression socks with elastic can help with flank or pelvic discomfort[23,24] and patients who are thin can gain weight[23]. If symptoms continue even after conservative treatment, or if the patient has considerable haematuria, severe discomfort/pain, or renal impairment, endovascular treatment is recommended[17,24]. Open surgery is recommended, if the procedure fails. After endovascular treatment if stent issues occur, either endovascular repair or open surgery repair is performed. Regular follow-up should begin when therapy or repair is effective and symptoms have resolved[17]. Our treatment strategy was effective in reducing the symptoms of our patient and he is getting better.

Patient education and dissuasion

Patients should be counselled about the risks, notably kidney dysfunction, if untreated. In individuals with pelvic congestion syndrome or varicocele, the potential risk of infertility should be discussed. Moreover, screening in the family members of the NCS patients is not important as NCS is not a genetic condition Figure 1.

Remember

NCS is clinically equivalent to Nutcracker phenomenon, which is characterized by complex symptoms substantially variable.
Typical NCS feature is the compression of LRV between AO and SMA.
Stenting, surgery and frequent urine testing are common treatments for NCS.

Ethical approval

None.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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None.

Author contribution

A.H. collected the data, drafted and revised the manuscript. S.H. collected the data, drafted the manuscript. G.R. drafted the manuscript. A.R. drafted the manuscript. M.A.-J. drafted and revised the manuscript.

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Authors report no conflict of interest.

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References

[1] Granata A, Distefano G, Sturiale A, *et al*. From Nutcracker phenomenon to nutcracker syndrome: a pictorial review. Diagnostics 2021;11:101.
[2] Karaman B, Koplay M, Ozturk E, *et al*. Retroaortic left renal vein: multidetector computed tomography angiography findings and its clinical importance. Acta Radiologica 2006;48:356–60.
[3] Alaygut D, Bayram M, Soylu A, *et al*. Clinical course of children with nutcracker syndrome. Urology 2013;82:686e690.
[4] Shin JI, Lee JS. Nutcracker phenomenon or nutcracker syndrome. Nephrol Dial Transplant 2005;20:2012.
[5] Shin JI, Lee JS, Kim MJ. The prevalance, physical characteristics and diagnosis of nutcracker syndrome [letter]. Eur J Vasc Endovasc Surg 2006;32:335–6.
[6] Kurklinsky AK, Rooke TW. Nutcracker phenomenon and nutcracker syndrome. Mayo Clin Proc 2010;85:552–9.
[7] Gong XY, Zheng W, Du H, *et al*. Treatment of nutcracker syndrome with spermatic vein ligation and iliac vein anastomosis: case report of three cases. Asian Pacific J Trop Med 2012;5:923–4.
[8] Waseem M, Upadhyay R, Prosper G. The nutcracker syndrome: an underrecognized cause of hematuria. Euro J Pediatr 2012;171:1269–71.
[9] Shin JI, Baek SY, Lee JS, *et al*. Follow-up and treatment of nutcracker syndrome [letter]. Ann Vasc Surg 2007;21:402.
[10] Wang L, Yi L, Yang L, *et al*. Diagnosis and surgical treatment of nutcracker syndrome: a single-center experience. Urology 2009;73:871–6.
[11] Basile A, Tsetis D, Calcara G, *et al*. Percutaneous nitinol stent implantation in the treatment of nutcracker syndrome in young adults. J Vasc Interv Radiol 2007;18:1042–6.
[12] Agha RA, Franchi T, Sohrabi C, *et al*. for the SCARE Group. The SCARE 2020 Guideline: Updating Consensus Surgical CAse REport (SCARE) Guidelines. Int J Surg 2020;84:226–30.
[13] Gulleroglu K, Gulleroglu B, Baskin E. Nutcracker syndrome. World J Nephrol 2014;3:277–81.
[14] Dunphy L, Penna M, Tam E, *et al*. Left renal vein entrapment syndrome: nutcracker syndrome!. BMJ Case Rep 2019;12:1–4.
[15] Maloni KC, Calligaro KD, Lipshutz W, *et al*. Nutcracker syndrome as an unusual cause of postprandial pain. Vasc Endovasc Surg 2020;54:283–5.
[16] Ribeiro FS, Puech-Leão P, Zerati AE, *et al*. Prevalence of left renal vein compression (nutcracker phenomenon) signs on computed tomography angiography of healthy individuals. J Vasc Surg Venous Lymphat Disord 2020;8:1058–65.
[17] de Macedo GL, Dos Santos MA, Sarris AB, *et al*. Diagnosis and treatment of the Nutcracker syndrome: a review of the last 10 years. J Vasc Bras 2018;17:220–8.
[18] Ananthan K, Onida S, Davies AH. Nutcracker syndrome: an update on current diagnostic criteria and management guidelines. Eur J Vasc Endovasc Surg 2017;53:886–94.
[19] Grimm LJ, Engstrom BI, Nelson RC, *et al*. Incidental detection of nutcracker phenomenon on multidetector CT in an asymptomatic population: prevalence and associated findings. J Comput Assist Tomogr 2013;37:415–8.
[20] Berthelot JM, Douane F, Maugars Y, *et al*. Nutcracker syndrome: a rare cause of left flank pain that can also manifest as unexplained pelvic pain. Joint Bone Spine 2017;84:557–62.
[21] Kim KW, Cho JY, Kim SH, *et al*. Diagnostic value of computed tomographic findings of nutcracker syndrome: correlation with renal venography and renocaval pressure gradients. Eur J Radiol 2011;80:648–54.
[22] Ha TS, Lee EJ. ACE inhibition can improve orthostatic proteinuria associated with nutcracker syndrome. Pediatr Nephrol 2006;21:1765–8.
[23] Butros SR, Liu R, Oliveira GR, *et al*. Venous compression syndromes: clinical features, imaging findings and management. Br J Radiol 2013;86:20130284.
[24] Hulsberg PC, McLoney E, Partovi S, *et al*. Minimally invasive treatments for venous compression syndromes. Cardiovasc Diagn Ther 2016;6:582–92.