



ELSEVIER

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case report

Superior mesenteric artery syndrome (Wilkie Syndrome) with unusual clinical onset: Description of a rare case [☆]

Renato Farina, MD^{a,*}, Pietro Valerio Foti, PhD^a, Maria Coronella, MD^a, Isabella Pennisi, MD^a, Federica Libra, MD^a, Alessia Di Mari, MD^a, Cecilia Gozzo, MD^a, Tiziana Vasile, MD^a, Elena Lamirata, MD^b, Massimiliano Veroux, PhD^b, Guido Basile, PhD^b, Antonio Basile, PhD^a

^a Department of Medical and Surgical Sciences and Advanced Technologies, Radiodiagnostic and Radiotherapy Unit, "GF Ingrassia" Catania, Italy

^b Department of General Surgery and Medical Surgical Specialties, University of Catania, General surgery. Catania, Italy

ARTICLE INFO

Article history:

Received 17 June 2021

Revised 30 June 2021

Accepted 1 July 2021

Keywords:

Wilkie's Syndrome
Doppler Ultrasound
Computed Tomography
Cardiovascular Abnormalities
Superior Mesenteric Artery Syndrome

ABSTRACT

Wilkie's Syndrome is a very rare disease caused by reduction of aorto-mesenteric space with consequent duodenum compression. It can combine with left renal vein stenosis which, when symptomatic, is known as "Nutcracker Syndrome". We describe a clinical onset case with epigastric pain without vomiting in a normal weight patient. 28-year-old woman who came to our observation for intense epigastric pain after a weight loss of 14 kg in 4 months. Multidetector Computed Tomography and Ultrasound revealed gastric and duodenal overdistension with hydro-air levels, severe duodenum stenosis, and left renal vein compression. Wilkie's Syndrome is common in anorexic individuals suffering from recurrent postprandial vomiting, onset with severe epigastric pain, without vomiting, is quite unusual. High-calorie diet must be first therapeutic approach, in case of failure treatment of first choice should be endovascular stenting and, only in selected cases, surgical treatment should be used because it is very invasive and burdened with numerous complications. Failure to diagnose this disease can expose patients to serious health risks.

© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

[☆] Competing Interests: The authors declare that they have no competing interests.

* Corresponding author.

E-mail addresses: radfaro@hotmail.com (R. Farina), pietrofoti@hotmail.com (P.V. Foti), mcoronella@sirm.org (M. Coronella), pennisi.isabella@gmail.com (I. Pennisi), federica.libra@hotmail.it (F. Libra), alessiadimari@hotmail.com (A. Di Mari), ceciliagozzo91@gmail.com (C. Gozzo), tiziana_vasile@hotmail.it (T. Vasile), elena.lamirata@libero.it (E. Lamirata), mveroux@libero.it (M. Veroux), gbasile@unict.it (G. Basile), basile.antonello73@gmail.com (A. Basile).
<https://doi.org/10.1016/j.radcr.2021.07.004>

1930-0433/© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Introduction

Wilkie's Syndrome (WS) also known as Superior Mesenteric Artery Syndrome [1] is a very rare disease caused by aorto-mesenteric space (AMS) reduction resulting in duodenum compression [2]. It can be congenital or acquired. In WS superior mesenteric artery originates with acute angle (<22 degrees) and aorto-mesenteric distance (AMD) is less than 8 mm (Figs. 1a,b,c and d). Acquired form is almost always due to rapid weight loss which causes perivascular adipose tissue reduction and progressive reduction of AMS, common in anorexic patients [3]. When left renal vein (LRV) is also compressed significantly (Nutcracker), it can become clinically symptomatic and is known as "Nutcracker Syndrome" (NCS) [4]. There are two types of NCS: anterior (ANCS) in which LRV runs anterior to aorta [5] and posterior (PNCS) when LRV runs posterior to aorta (retro-aortic LRV) and is compressed between abdominal aorta and spine [6].

LRV compression can be clinically asymptomatic (Nutcracker Phenomenon) [7] or symptomatic (NCS) and cause microhematuria, left flank pain, venous congestion with consequent left varicocele; in severe cases it can cause venous thrombosis. In WS duodenal compression generally occurs with recurrent postprandial vomiting and

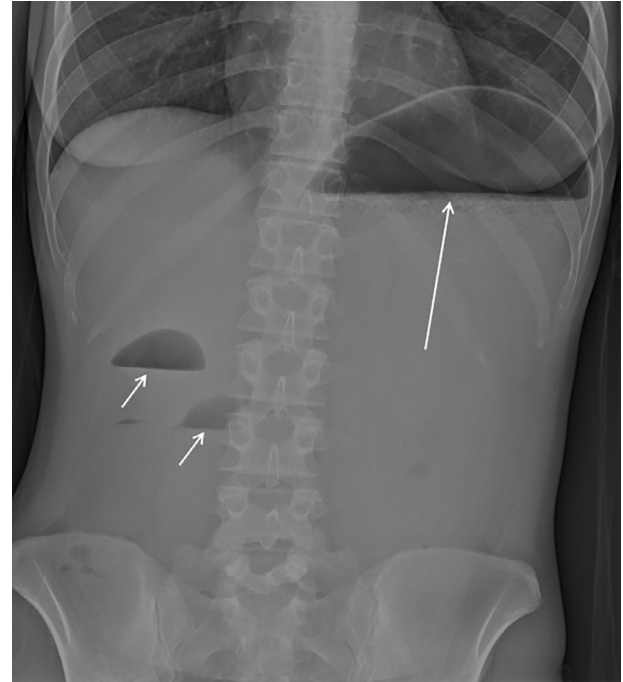


Fig. 2 – Abdominal CR showing: air/fluid level into stomach (long arrow) and duodenum (short arrows), and meteorism absence in left lower abdominal quadrants.

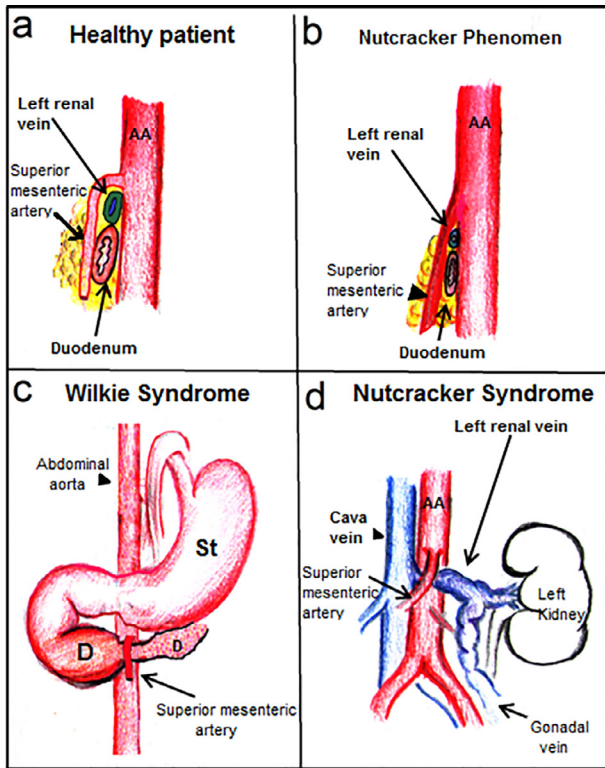


Fig. 1 – Summary diagram of anatomical structures involved in WS and NCS. a: healthy patient with regular aorto-mesenteric angle and distance. b: patient affected by Nutcracker Phenomenon with aorto-mesenteric angle reduction. c: patient with duodenal compression due to MSA (WS). d: Patient with NCS with LRV symptomatic compression.

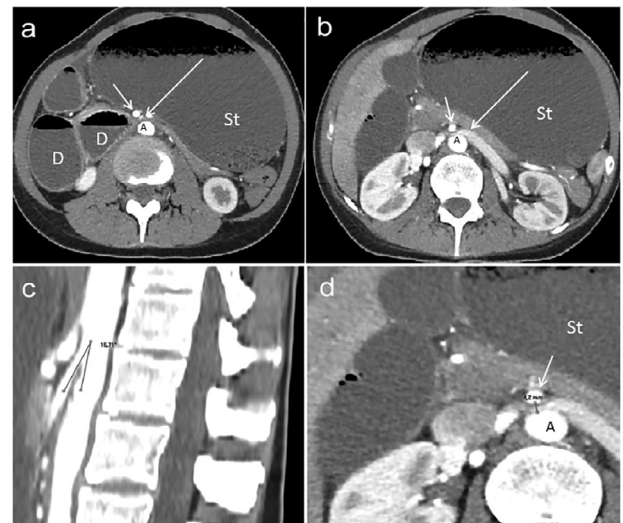


Fig. 3 – Abdomen MDCT. a: Reconstruction on an axial plane shows stomach (St) and duodenum (D) that appears over-stretched upstream of stenosis. MSA (short arrow). Aorto-mesenteric space (long arrow). Abdominal Aorta (A). b: In this reconstruction on an axial plane LRV (long arrow) is compressed in aorto-mesenteric space. MSA (short arrow). Abdominal Aorta (A). c: This reconstruction on an sagittal plane shows MSA originates from AA with an acute angle (beak sign) of less than 22 degrees. d: Reconstruction on an axial plane shows aorto-mesenteric distance measurement which results less than 8mm. Abdominal Aorta (A). MSA (arrow). Duodenum (D). Stomach (St).

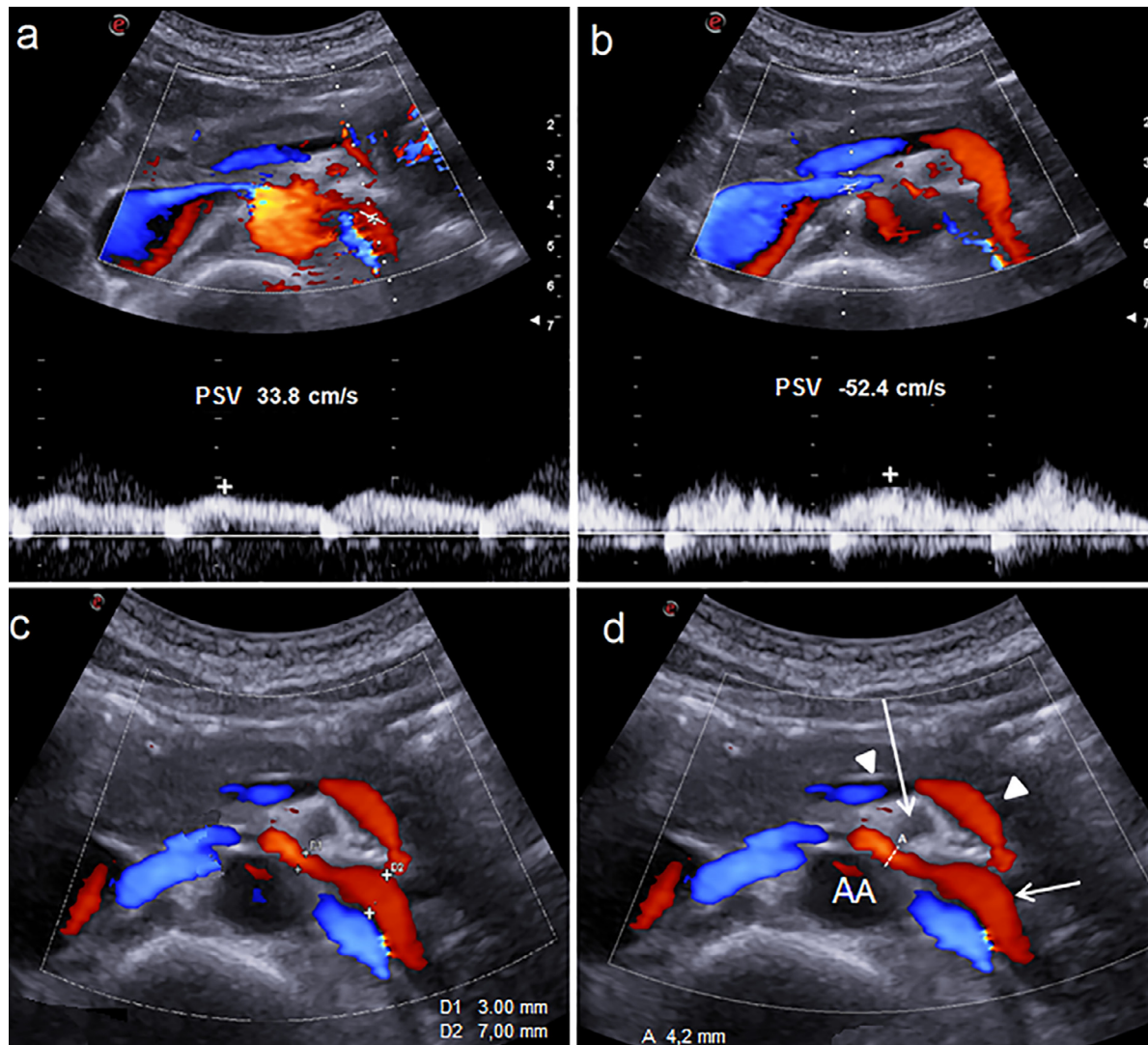


Fig. 4 – a: Duplex Doppler US. Flow sampling performed by landmark positioning in LRV pre-stenotic tract shows low PSV (33.8 cm / s) and **-b:** PSV increase in post-stenotic tract (52.4 cm / s). **c:** Color Doppler US of aorto-mesenteric space shows LRV stenosis (D1) and pre-stenotic dilatation (D2). Left renal vein (long arrow). **d:** Aorto-mesenteric distance measurement (4.2mm). Abdominal Aorta (AA). Mesenteric superior artery (long arrow). Spleno-mesenteric confluence (arrows heads).

sub-occlusive crisis; very rarely it is associated with painful epigastric crises. Treatment depends on stenosis entity and symptomatology and must initially be based on a high-calorie liquid diet aimed at perivascular adipose tissue restoring [9]; if this fails, endovascular LRV stenting or surgery may be indicated. Most common surgical treatment consists in resection of first duodenal loop and retro-vascular duodenum and anastomosis between duodenum and second duodenal loop repositioned anterior to MSA [10]. Endovascular treatment consists in placing a stent in stenotic tract of LRV, restoring normal vascular flow and normal aorto-mesenteric angulation [11]. People with WS are generally very thin and suffer from recurrent episodes of postprandial vomiting and nausea. We describe a rare clinical presentation of disease with severe epigastric pain, without vomiting in a patient with normal body mass index.

Case presentation

28-year-old woman came to our emergency room for intense epigastric pain after taking antispasmodic drugs (Phloroglucine and 1,3,5 Trimethoxybenzene - Spasmex). Patient, of normal weight with a body mass index of 20, had reported rapid weight loss following a low calorie diet with loss of 14 kg in past 4 months. Laboratory tests showed no significant alterations. On clinical examination abdomen was tense and painful and pain increased after compression. Conventional abdomen Radiography (CR) was performed which revealed gastric and duodenal overdistension with hydro-air levels and meteorism absence in remaining abdominal quadrants (Fig. 2). Based on CR results, Multidetector Computed Tomography (MDCT) was performed using a 64-slice Optima

Table 1 – Summary of results obtained by Duplex Doppler US. PSV: Peak Speed Velocity. LRV: Left renal vein.

LRV	PSV	Caliber	
Flow Ratio			1,55
Pre-stenotic tract	33,8 cm / s	7 mm	
Post-stenotic tract	52,4 cm / s	6,9 mm	
Stenotic tract	73,5 cm / s	3 mm	

device (GE Healthcare). MDCT revealed gastric and duodenal overdistension and LRV stenosis (Fig. 3a, b), caused by significant reduction in aorto-mesenteric angle (AMA) and AMD (Fig. 3c, d). In sagittal reconstruction, AMA assumed “beak-sign” appearance due to MSA which in WS originates from aorta at an acute angle (Fig. 3c). MDCT ruled out other abdominal diseases. Subsequently, patient underwent Ultrasound using a MyLab Nine device (Esaote Biomedica) with a convex 3.5 MhZ and linear 7.5 MhZ probe. Ultrasound was performed by an operator with twenty years of experience. Were measured in detail: AMA, AMD (1 cm from MSA origin), LRV peak speed velocity (PSV) in pre-stenotic and post-stenotic tract; Flow Ratio (FR) (post-stenotic tract PSV / prestenotic tract PSV), Resistive Index (RI) of left renal and intrarenal artery, LRV caliber of pre- and post-stenotic tract and gonadal plexus veins diameter. Color Doppler US and Duplex Doppler US showed LRV flow congestion with PSV reduction in pre-stenotic tract (33.8 cm / s) (Fig. 4a), increase in post-stenotic tract (52.4 cm / s) (Fig. 4b) and stenotic tract (73.5 cm / s); LRV diameter reduction in stenotic tract (3 mm), slight dilation in prestenotic (6.9 mm) and post-stenotic tract (7 mm) (Fig. 4c); AMA reduction (15 degrees) and AMD reduction (4.2 mm) (Fig. 4d). FR was 1.55. Color Doppler Ultrasound of gonadal plexus showed no varicocele. Results are summarized in Table.1. On basis of results obtained, diagnosis of WS combined with NCS was made. Patient was discharged after few days with high-calorie liquid diet prescription and Ultrasound follow-up. At first control after three months AMA was significantly increased (28 °) and symptoms completely regressed.

Discussion

Ultrasound is first-line examination for NCS study [8], it allows to measure AMA and AMD reduction, LRV stenosis and flow congestion, varicocele and LRV thrombosis; furthermore Duplex Doppler Ultrasound (Duplex Doppler US) allows to obtain an estimate LRV stenosis degree thanks to Flow-Ratio (FR) measurement, all very important information for therapeutic planning. Duodenum study represents a Ultrasound limitation and therefore diagnostic integration with Conventional Radiography (CR) or Computed Tomography (CT) are necessary. CR was indicative of high intestinal obstruction. Comparing radiological findings with patient’s clinical history: rapid weight loss, post-prandial vomiting, fever absence and flogosis index alterations absence, WS was suspected. Diagnostic suspicion was confirmed by MDCT that showed gastric and duodenal overdistension extended up to AMS which appeared considerably narrow and in addition to duodenum caused LRV compression.

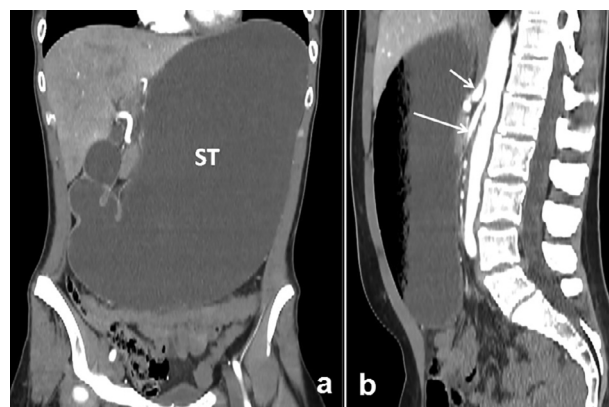


Fig. 5 – Abdomen CT. a: This reconstruction on an coronal plane shows gastric overdistension. Stomach (ST). b: This sagittal plane reconstruction shows over-distended stomach compressing celiac artery (short arrow) and MSA (long arrow). Clip: In this clip are summarized respectively: longitudinal B-Mode US scan of AA showing a reduction of aorto-mesenteric angle; transverse B-Mode US scan of AA showing a reduced distance between AA and MSA; transverse color Doppler US scan showing LRV stenosis; Duplex Doppler US measurement of pre-stenotic and post-stenotic traits of LRV.

After identifying intestinal obstruction, it was necessary to know extent of LRV compression, in order to plan a correct treatment. When compression is not haemodynamically significant and in absence of renal alterations, treatment must be conservative limited to high-calorie liquid diet only, aimed at restoring perivascular adipose tissue and normal AMD. When LRV stenosis is greater than 70%, high calorie diet must be combined with LRV endovascular stenting.

Duplex Doppler US was essential for stenosis degree evaluation, obtained from FR calculation, which in our case was 1.55, corresponding to a stenosis of less than 50%. FR of 2.5 corresponds to a stenosis of 50% and does not require treatment [12], if instead stenosis exceeds 70%, best treatment is vascular stenting associated with long-term anticoagulant drugs [13]. In our case, therefore, we did not consider drug therapy necessary. Intense postprandial epigastric pain, rather unusual in WS, which led patient to Emergency Room, was probably caused by antispasmodic drug-induced gastric overdistension which further aggravated MSA compression (Figs. 4a and b); placement of nasogastric tube caused leakage of about 3.5 liters of gastric contents and regression of painful symptoms. Patient was discharged after a few days with prescription for high-calorie liquid diet and Ultrasound follow-up. At first follow-up after three months, AMA was significantly increased (28 °) and symptoms almost totally regressed Figure 5.

Conclusion

Vascular compression syndromes are very rare, which is why they are still poorly understood. WS should be suspected in

very thin individuals with recurrent postprandial vomiting. Doppler Ultrasound it can highlight with great accuracy AMA reduction and possible LRV stenosis, however it needs integration with MDCT for duodenum study. Failure to diagnose this disease can expose patients to serious health risks.

An optimal therapeutic protocol has not yet been standardized as there are still numerous controversies in literature. Surgical treatment is very invasive and burdened with numerous complications and should only be considered after failure of other therapies.

"Written informed consent was obtained from the patient for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the chief editor of this journal."

Funding

There are no sources of funding for the research.

Author contribution

RF Study design/planning collected data, preparation of manuscript, data analysis/statistics, data interpretation and involved in project development, literature analysis/search PVF, MVC, IP, FL, AD, CG, TV, EL, MV and GB collected data, wrote the manuscript, literature analysis/search. RF and AB: wrote the manuscript.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Statement related to the patient's consent: The consent was obtained from the patient for the publication of this case report and accompanying images.

Declarations

Ethics approval and consent to participate

Not applicable.

Availability of data and materials

All data generated or analyzed during this study are included in this published article and its additional files.

Patient consent

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

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2021.07.004](https://doi.org/10.1016/j.radcr.2021.07.004).

REFERENCES

- [1] Karki B, Pun B, Shrestha A, et al. Superior mesenteric artery syndrome. *Clin Case Rep* 2020 Jul 16;8(11):2295–7.
- [2] Gozzo Cecilia, Giambelluca Dario, Cannella Roberto, et al. CT imaging findings of abdominopelvic vascular compression syndromes: what the radiologist needs to know. *Insights Imaging* 2020 Dec; 11:48.
- [3] Johnson BM, Paladugu G. Superior Mesenteric Artery Syndrome Secondary to Anorexia Nervosa and Methamphetamine Use. *Cureus* 2019 Nov; 11(11):e6121.
- [4] Diab Samer, Hayek Fadi. Combined Superior Mesenteric Artery Syndrome and Nutcracker Syndrome in a Young Patient: A Case Report and Review of the Literature. *Am J Case Rep* 2020;21:e922619 -1–e922619-5.
- [5] De Macedo GL, Alves dos Santos M, Biff Sarris A, et al. Diagnosis and treatment of the Nutcracker syndrome: a review of the last 10 years. *J Vasc Bras Jul-Sep* 2018;17(3):220–8.
- [6] Lourenço de Macedo G, Alves dos Santos M, Biff Sarris A, et al. Venous revascularization to treat posterior nutcracker syndrome by transposition of the left gonadal vein: case report. *J Vasc Bras* 2019;18:e20190037.
- [7] Sawant DA, Moore TF. NC Phenomenon) (An Unusual Course of Segmental Renal Artery Displays a Rare Case of Iilar Nutcracker Phenomenon. *Case Rep Med* 2015;2015:249015.
- [8] Seung Hyup Kim. Doppler US and CT Diagnosis of Nutcracker Syndrome. *Korean J Radiol* 2019 Dec; 20(12):1627–37.
- [9] Farina R, Foti PV, Cocuzza G, et al. Wilkie's syndrome. *J Ultrasound* 2017 Aug 3;20(4):339–42.
- [10] Heidbreder Rebeca. Co-occurring superior mesenteric artery syndrome and nutcracker syndrome requiring Roux-en-Y duodenojejunostomy and left renal vein transposition: a case report and review of the literature. *J Med Case Rep* 2018;12:214.
- [11] Quevedo HC, Arain SA, Abi Rafeh N. Systematic review of endovascular therapy for nutcracker syndrome and case presentation. *Cardiovasc Revasc Med Jul-Aug* 2014;15(5):305–7.
- [12] Labropoulos N, Borge M, Pierce K, et al. Criteria for defining significant central vein stenosis with duplex ultrasound. *J Vasc Surg* 2007;46(1):101–7.
- [13] Avgerinos ED, Saadeddin Z, Humar R, et al. Outcomes of left renal vein stenting in patients with nutcracker syndrome. *J Vasc Surg Venous Lymphat Disord* Nov 2019;7(6):853–9.