SAGE Open Medical Case Reports
Volume 11: 1–4

DOI: 10.1177/2050313X231176395
journals.sagepub.com/home/sco
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An atypical cause of vomiting: Coexisting Wilkie's syndrome and a left renal malformation mimicking a nutcracker phenomenon—A case report

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Abstract

The superior mesenteric artery syndrome and nutcracker phenomenon are rare vascular disorders due to the abnormal development of the superior mesenteric artery stemming from the abdominal aorta with reduced angle (<22°) and resultant compression of the left renal vein and duodenum. It is an underreported entity due to the absence of specific pathognomonic signs. We report the case of a 59-year-old man, admitted for acute bilious vomiting, who underwent a gastroscopy and a computed tomography scan revealing a Wilkie's syndrome associated with a dilated posterior left renal vein communicating with the left ascending lumbar vein without connection with the inferior vena cava mimicking a nutcracker phenomenon.

Keywords

Wilkie, nutcracker, vomiting, imaging, computed tomography scan, endoscopy, double left renal vein

Date received: 2 January 2023; accepted: 27 April 2023

Introduction

The superior mesenteric artery syndrome (SMAS) and nutcracker phenomenon (NCP) are rare vascular disorders due to the abnormal development of the superior mesenteric artery (SMA) stemming from the abdominal aorta with reduced angle (<22°) and resultant compression of the left renal vein (LRV) and duodenum.[1]

It is an underreported entity due to the absence of specific pathognomonic signs.[2]

Moreover, various anatomical abnormalities of the LRV can be found such as circumaortic, retro-aortic or multiple renal vein.[3]

We report the case of a 57-year-old man, admitted for acute bilious vomiting, to which the diagnosis of a SMAS and a double renal vein mimicking a NCP was retained.

Case presentation

A 59-year-old man, with history of chronic smoking for over 30 years, was admitted in the emergency room (ER) for acute bilious vomiting and epigastric pain.

He had a prolonged history of recurrent pain in the epigastrium and multiple episodes of vomiting, with no urinary symptoms, accompanied with asthenia and a weight loss of 10 kg 4 months after the start of the clinical manifestations.

His initial vital signs were normal. The physical examination showed a non-distended abdomen with mild epigastric tenderness. No hepatomegaly or large palpable gallbladder was palpable.

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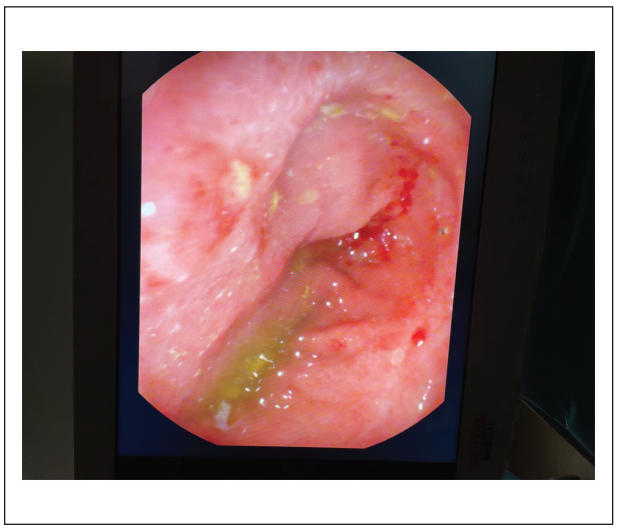

Figure 1. Endoscopic view of the stenosis in the second portion of the duodenum.

Other than a 2.3 mmol/L hypokalemia and a 129 mmol/L hyponatremia, routine blood and urine tests were normal.

The patient proceeded to an upper endoscopy, which revealed an impassable D2 stenosis with D1 dilation and bilious reflux (Figure 1). No overt intraluminal pathology was identified.

An angiography computed tomography (CT) scan was then performed revealing a marked dilation of the stomach and proximal duodenum with an abrupt caliber change in the third portion of the duodenum. The aortomesenteric angle (AMA) and aortomesenteric distance (AMD) were 12° (normal range, 38°–65°) and 3.5 mm (normal range, 10–28 mm), respectively (Figures 2 and 3). The caliber of the colon and the small intestine were otherwise normal. Moreover, a dilated posterior LRV and left lumbar ascending vein, without a dilation of the left gonadal vein was found, suggesting the diagnosis of the posterior NCP (Figure 4).

Based on the clinical history and the imaging findings, a diagnosis of SMAS associated with a NCP was made.

The patient was managed by duodenojejunostomy (done by open surgery) which enhanced his obstruction and appetite.

He was discharged 2 weeks later with a good follow-up and a close observation for his renal nutcracker syndrome.

After 6 months, the patient gained 3 kg and did not experience any pain since surgery.

By reviewing his imaging with more scrutiny, it turned out that it was not a NCP but a double renal vein.

- The anterior one flowing directly toward the inferior vena cava (IVC) with no dilation, even with a very narrowed distance and angle between the SMA and the aorta (Figure 4).

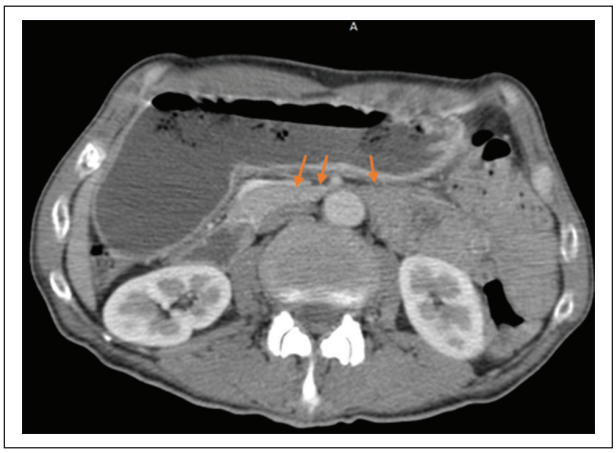

Figure 2. CT scan in axial plan showing the duodenal compression between the aorta and superior mesenteric artery, with a narrowed AMD 3.5 mm.

- The posterior renal vein, which drains directly into the left ascending lumbar vein and empties into the hemiazygos vein, without communicating with the IVC or any other renal vein and without individualizing any retro-aortic segment (Figure 5).

Discussion

SMAS was first described in 1861 by Rokitansky,[4] as a compression of the third part of the duodenum by the narrow angle between the SMA and the aorta.

It is not until the 1920s that a more detailed clinical, anatomical and physiological description was provided by Wilkie,[5] whose name has become an eponym for SMAS.

This potentially life-threatening syndrome is rare as an estimated incidence of less than 0.3% has been reported in the literature.[2]

Normally, the AMA and AMD are 38°–56° and from 10 to 28 mm, respectively. In SMAS, both parameters are reduced with angles less than 15° and a distance from 2 to 8 mm.[1]

Its physiopathology is well recognized in the literature, and it includes any event that further intensifies the narrowing of the aortomesenteric area.

Congenital abnormalities or anatomical defects as duodenal malrotation, a short ligament of Treitz, congenitally low origin of SMA and peritoneal adhesions can likely result in a manifestation of SMAS. This may be exacerbated by some external events like an important weight loss (leading to a loss of retroperitoneal fatty tissues) or an external abdominal compression by a belt or a tumor.[6]

This syndrome can occur as an acute illness or can develop insidiously, depending on its origin and the importance of the duodenal obstruction.

The most characteristic symptoms are post-prandial epigastric fullness, pain, eructation and bilious vomiting. They

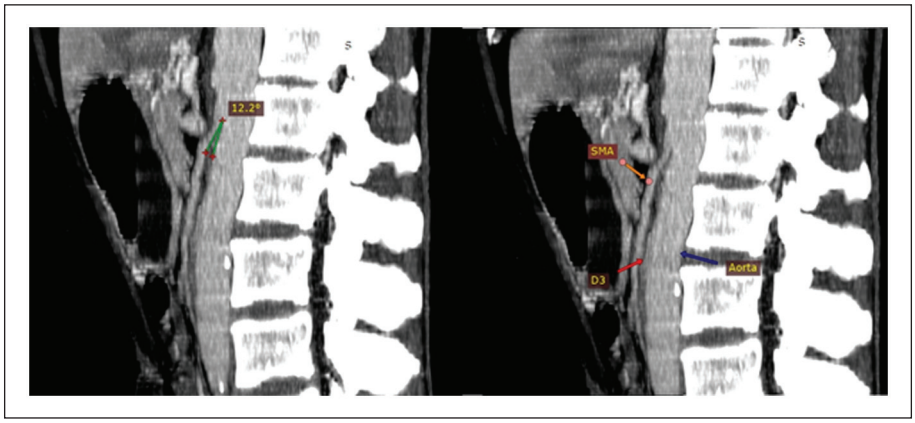

Figure 3. The sagittal reconstruction contrast-enhanced CT image shows an AMA of 12.2°.

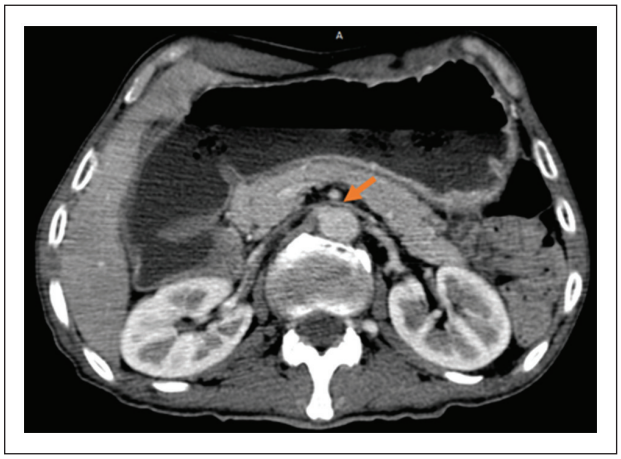

Figure 4. Axial plan showing narrowing between SMA and aorta: distance 3.7 mm, and a normal aspect of the left renal vein.

are worsened by meals and on dorsal decubitus, and can be improved by left lateral deviation and knee–chest postures.[7]

The diagnosis is generally based on clinical findings and radiological evidence of obstruction. Indeed, CT angiography can show the duodenal obstruction and evaluate the decrease in angle and space between the aorta and SMA.[8]

Upper gastrointestinal endoscopy may reveal narrowing of the third part of the duodenum.

Conservative management, as decompression through nasogastric drainage and electrolyte correction, is often successful. However, in unresponsive and long-standing cases or in patients with massive duodenal dilation, definitive surgical therapy can be required.

A number of surgical options have been described such as a gastrojejunostomy, a division of the ligament of Treitz with duodenal mobilization (Strong's operation) and duodenojejunostomy standing as a safe and effective technique, improving symptoms and quality of life.[9]

Among other complications of the aortomesenteric clamp, we can cite a compression of the LRV leading to a nutcracker syndrome or phenomenon.

Although these terms are used interchangeably, it does not refer to the same entity.

Indeed, the syndrome includes not only the anatomical variation, but also the existence of clinical symptoms reflecting the involvement of the LRV, in particular hematuria, proteinuria and left low back pain. This compression will also be responsible of dilation of the gonadal veins, manifested by varicocele in men and pelvic congestion in women.

On the contrary, the NCP corresponds only to a compression of the renal vein without clinical symptoms.[10] The diagnosis of nutcracker syndrome is confirmed by imaging, including Doppler ultrasound of the renal arteries, magnetic resonance imaging (MRI), venography, intravascular ultrasound or an abdominal contrast-enhanced (CE) CT scan, following established criteria such as:[3]

1. Narrowing of the LRV at the aortomesenteric portion (the beak angle $< 32°$);
2. Beak sign: severe form of narrowing of the LRV at the aortomesenteric portion;
3. LRV diameter ratio (hilar to aortomesenteric) $\geqslant 4.9$;
4. Angle between the SMA and aorta $< 41°$;
5. Collateral venous circulation developed in the retroperitoneum and renal hilum.

Conservative management can be sufficient, but endovascular stenting stands as the treatment of choice.[11]

In this case report, we confused a NCP with a double renal vein variation.

The first renal vein was not very dilated and the second was communicating directly with the ascending lumbar vein, also dilated and joining the intrathoracic azygos vein, without any communication with the IVC.

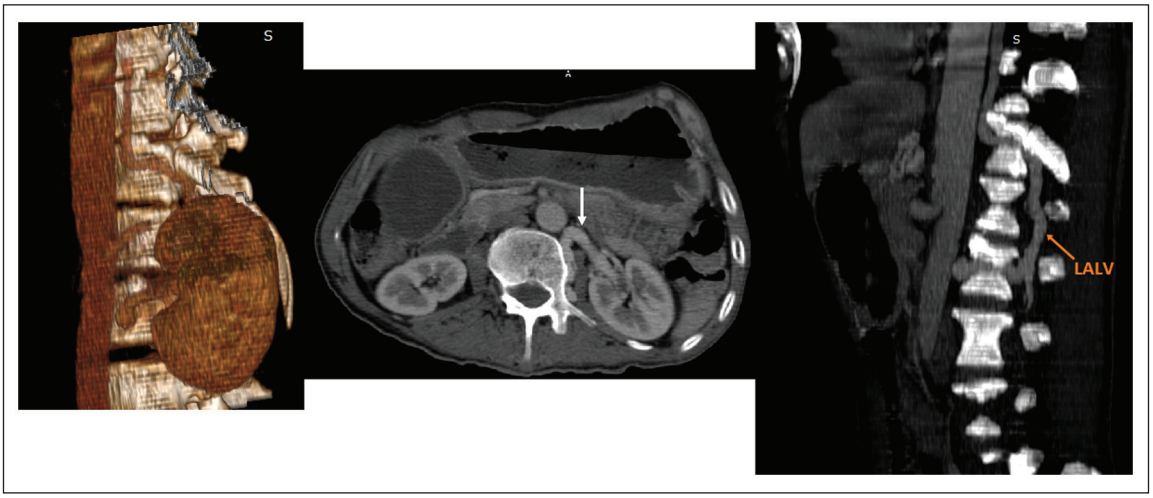

Figure 5. Axial and sagittal reconstruction contrast-enhanced CT scan showing the dilation of the lumbar plexus (white arrow showing the posteriori left renal vein and its communication with the ascending lumbar vein).

Very few cases in the literature described a connection between the LRV and Left ascending lumbar vein without a communication with other LRVs or IVC.

Conclusion

Wilkie's syndrome represents an uncommon diagnosis of upper gastrointestinal obstruction that should not be neglected if no other organic or metabolic cause is detected.

Usually symptomatic, the SMAS can be complicated by a NCP, with or without its own clinical manifestations.

Imaging allows diagnosis, with CT angiography standing as the modality of choice to confirm diagnosis.

Sharing common features resulting from narrowed AMA, this rare association should be looked for by clinicians and considered in cases of acute vomiting.

Author contributions

B.A. and S.E. contributed to manuscript writing and literature review. Y.B. was responsible for patient care and data information. S.B., T.A. and M.T. approved the drafts. L.B. and R.S. were responsible for imaging of the patient. O.B. and A.A.A. were responsible for surgical care.

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship and/or publication of this article.

Ethical approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patient(s) for their anonymized information to be published in this article.

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