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Combined Superior Mesenteric Artery Syndrome and Nutcracker Syndrome in a Young Patient: A Case Report and Review of the Literature

Authors' Contribution:
Study Design A
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Patient: **Male, 18-year-old**
Final Diagnosis: **Superior mesenteric artery syndrome**
Symptoms: **Abdominal pain**
Medication: —
Clinical Procedure: —
Specialty: **Surgery**

Objective: **Rare co-existence of disease or pathology**





Background: Superior mesenteric artery syndrome is the compression of the third portion of the duodenum between the superior mesenteric artery and the aorta causing abdominal pain and vomiting. Nutcracker syndrome is the compression of the left renal vein between the superior mesenteric artery and the aorta causing symptoms related to renal congestion. Both entities, although well described in the literature, are individually rare, and even though they might share a common etiology, their co-existence has been reported in only a few case reports.

Case Report: An 18-year-old male, previously healthy, presented with postprandial abdominal pain relieved by bilious vomiting that started shortly after he lost weight fasting. Our investigation revealed superior mesenteric artery syndrome as well as a compressed left renal vein. He was started on an enriched fluid diet which was progressed gradually as he regained weight. His left renal vein compression at the time was asymptomatic; it will be followed up for possible resolution after the patient's weight returns to normal.

Conclusions: Superior mesenteric artery syndrome is to be suspected in patients with abdominal pain following weight loss. Conservative treatment with a focus on weight regain will cure most cases. Asymptomatic or mildly symptomatic nutcracker syndrome is treated conservatively. For patients requiring intervention, laparoscopic extravascular titanium stent placement seems to be the least invasive promising option today, awaiting further definitive studies.

MeSH Keywords: **Rare Diseases • Renal Nutcracker Syndrome • Superior Mesenteric Artery Syndrome**

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Background

Superior mesenteric artery (SMA) syndrome (also called Wilkie syndrome) is the compression of the third portion of the duodenum between the aorta and the SMA. It is a rare entity with an incidence between 0.13% and 0.3% and a preference for young thin females [1]. Typical symptoms include abdominal pain and vomiting, more pronounced postprandially. It is most commonly caused by weight loss that decreases the fat plane between the duodenum and the SMA [2]. The diagnosis is made with a correlation between radiologic changes, such as duodenal dilation in the presence of a narrow angle of takeoff of the SMA, and clinical presentation [3]. The management focuses on nutritional support and weight regain which cures most cases [4].

Nutcracker syndrome (NCS) is the compression of the left renal vein (LRV) between the SMA and the aorta in an anterior fashion, or less commonly between the aorta and the vertebral column in a posterior fashion. It is also a rare entity. It usually manifests as flank pain with or without hematuria, proteinuria, or renal insufficiency. The causes are usually linked to anatomical variants [5]. The diagnosis can be made with a correlation of radiologic findings and clinical presentation with or without the need for invasive pressure measurement of the LRV [6]. The management of NCS ranges from watchful waiting, for mild cases, to various reconstructive surgical options [7].

Both entities, although well described in the literature, are individually rare. This is a case report of a young male having both syndromes simultaneously.

Case Report

An 18-year-old male, previously healthy, presented complaining of postprandial abdominal pain relieved by bilious vomiting. The pain was epigastric, crampy, and started shortly after eating. It was more pronounced with solids than liquids. The patient reported associated abdominal fullness and bilious, non-bloody, non-projectile vomiting that relieved his pain. His symptoms started after he was fasting for religious purposes and lost some weight. He later developed the inability to tolerate PO intake that led to a weight loss of 25 kg (32% of his body weight) in 3 months. He had no resting abdominal pain, no major change in bowel habits, and no other symptoms. On physical examination, there was no localized tenderness and the abdomen was soft. There was no bulging and there were no palpable masses; all signs were negative. He was seen by other physicians and his workup, including complete blood count, electrolytes, C-reactive protein, aminotransferases, and a gastroscopy was unremarkable. We ordered a computed tomography angiography (CTA). The first finding



Figure 1. Computed tomography angiography showing the incidental finding of a horseshoe kidney (red arrow).

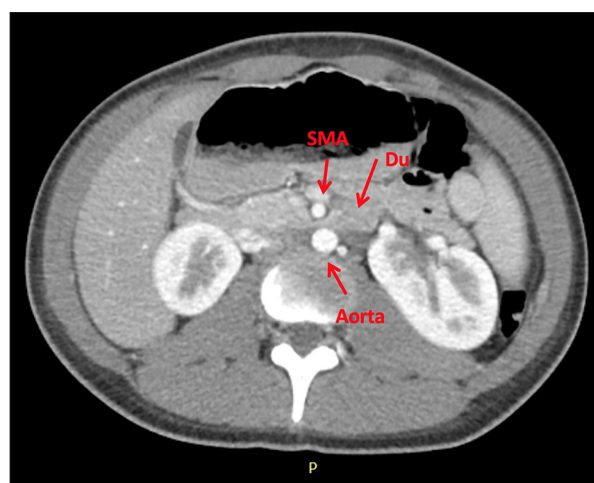


Figure 2. Computed tomography angiography showing the compression of the duodenum (Du) between the superior mesenteric artery (SMA) and the aorta.

was an incidental finding of a horseshoe kidney (Figure 1). We then identified a duodenal obstruction between the SMA and the aorta (Figure 2) as well as a dilated LRV that seemed also to be compressed between the SMA and the aorta without evidence of renal or pelvic congestion (Figure 3). Figure 4 shows the acute takeoff of the SMA at an angle of 12.7 degrees. The patient was diagnosed with SMA syndrome. Because of the radiologic finding of the compressed renal vein, we further investigated the patient's history. He had no history of left flank pain, no left sided varicoceles on physical examination; a urinalysis was ordered and failed to reveal hematuria or proteinuria. The patient was started on an enriched fluid diet which he tolerated; he gained 7 kg in 3 weeks at which

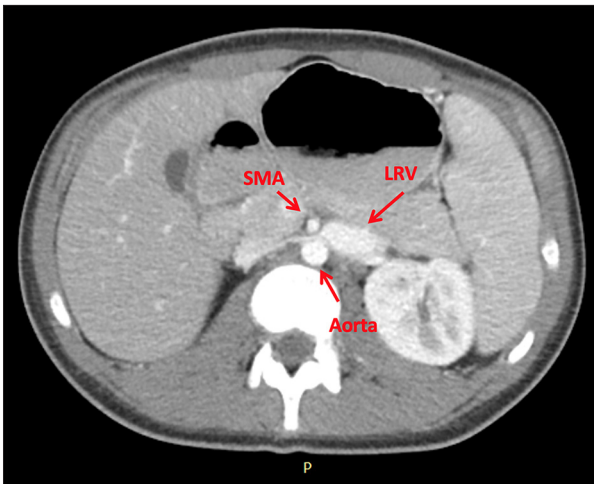


Figure 3. Computed tomography angiography showing the left renal vein (LRV) compression between the superior mesenteric artery (SMA) and the aorta.



Figure 4. Sagittal computed tomography angiography showing the acute takeoff of the superior mesenteric artery from the aorta at an angle of 12.7 degrees.

time he was progressed to mashed diet and 1 week later he was tolerating a regular diet.

Discussion

SMA syndrome is a rare entity with an incidence reported between 0.013% and 0.3% [1]. The typical patient is a young thin female; however, it can be seen in both sexes and all age groups. The mechanism is duodenal obstruction leading to postprandial abdominal pain relieved by vomiting and inability to tolerate PO intake. Considering the anatomical position of the duodenum between the SMA and the aorta, a common cause of the syndrome is weight loss that reduces the fat plane between both vessels leading to the compression that causes further weight loss and thus creates a vicious cycle. Some congenital anatomical changes have also been suggested as causative or aids to the development of the syndrome, those being a low takeoff of the SMA or a high insertion of the ligament of Treitz [2]. Some authors suggest conditions such as burns, bariatric surgeries, and cancers as causes; however, in our opinion, these are conditions that predispose to weight loss but do not themselves cause the syndrome. The diagnosis of SMA syndrome has always been a challenge because there is no single specific test available. Laboratory values may show nonspecific changes of weight loss and electrolyte imbalances, or laboratory values may even be normal. Imaging such as CT scan or MRI may show abnormalities such as a dilated duodenum proximal to the SMA takeoff, a short aorto-mesenteric distance, or a narrow aorto-mesenteric takeoff angle. The normal aorto-mesenteric angle is reported to be between 28 and 65 degrees and the normal aorto-mesenteric distance is between 10 and 34 mm [8]. Patients with SMA syndrome usually have a shorter distance and a narrower angle. A study using multidetector computed tomography (MDCT) to evaluate patients with SMA syndrome found an average angle of 13.5 degrees and an average distance of 4.4 mm. No clear cutoff for diagnosis based on imaging alone can be made; however, some authors have suggested that in the right clinical setting, patients with angles less than 22 degrees and distances less than 8 mm are highly suggestive of the diagnosis [3]. A novel technique was suggested by Guiseppe et al. where they diagnosed 2 patients using magnetic resonance enterography (MRE), a combination of magnetic resonance imaging with dynamic PO contrast. MRE offers the advantage of showing the anatomic variants while identifying the resulting mechanical consequences such as delayed transit and obstruction [9].

As for the management of SMA syndrome, conservative treatment with a focus on weight regain while watching for proper caloric needs with care for refeeding syndrome can cure most cases [4]. Management starts with per OS (by mouth) enriched fluids if the patient can tolerate it, moving to nasojejunal or

jejunostomy feeding tubes. Some authors have suggested positional changes and maneuvers can aid in tolerating enteral feeds; positional changes such as prone position and left lateral decubitus position, and maneuvers such as the knee-chest maneuver, and the Hayes maneuver (compression infraumbilically posteriorly and cephalad) [10,11]. If those measures fail, a trial of parenteral nutrition seems reasonable. If all conservative interventions fail, which is rarely the case, the patient then warrants surgical intervention. Options for surgical interventions are duodenojejunostomy, gastrojejunostomy, and duodenal derotation (also called the Strong's procedure).

Given the small number of reported cases in the literature and no large clinical trials reported, there is no level 1 evidence available regarding surgical interventions.

Duodenojejunostomy is considered by many as the procedure of choice for causing minimal enteric bypass; and a number of case series reported high success rates [12–15]. As gastrojejunostomy will cause more nutritional loss, it seems reasonable to keep it as second-line treatment. As for the Strong's procedure, the division of the ligament of Treitz and derotation of the duodenum to the right side, although advocated by some, has been shown to have a high failure rate possibly due to the tense duodenal entrapment proximally between the pancreaticoduodenal arteries [16].

Nutcracker syndrome (NCS) is also a rare entity, with few cases reported in the literature. Considering the anatomical position of the LRV in the same plane as the duodenum, weight loss and subsequent reduction of the fat plane between the SMA and the LRV seems to be the logical cause for the development of this syndrome. However, anatomical variants play a bigger role with NCS than with SMA syndrome, those anatomical variants being posterior renal ptosis, an abnormally high course of the LRV, and an abnormal SMA branching from the aorta [5]. We note that the incidental finding of the horseshoe kidney in our patient did not seem to predispose him to having NCS as the horseshoe kidney takes a lower anatomical position after its entrapment during the embryological ascent. The manifestations of NCS range from asymptomatic to left flank pain, pelvic congestion with left sided varicoceles, hematuria, and proteinuria [5]. A hallmark of NCS is the aggravation of symptoms with position, as standing upright causes the viscera to drop down thus pulling the SMA and aggravating the compression [17]. The diagnosis can be made by Doppler ultrasound, CT scan, or MRI where a dilated vein with a delayed washout and pelvic varicosities in the right clinical setting are highly suggestive. LRV phlebography is an invasive but conclusive method for confirming the elevated pressure in the LRV [6]. As for the management of NCS, conservative treatment seems reasonable for mild pain or mild hematuria as 75% of cases resolve within 2 years [7]. For cases that fail

to resolve or cases with significant symptoms, intervention is warranted. Surgical options are nephropexy, LRV or SMA transposition, gonadocaval bypass, renal to inferior vena cava (IVC) shunt, renal auto-transplantation, or nephrectomy [5]. With advances in endovascular interventions, case reports of successful intravascular stent placement have been published [18–22]. However, these cases of intravascular stent placement were later found to have a high migration rate (6.7%), including migration to the heart [23]. With advances in 3-dimensional (3D) printed stents, Li et al. (2016) reported on laparoscopic placement of an extravascular titanium stent [24]. This approach is appealing given that there would be no need for prolonged anticoagulation, a low risk of migration compared to intravascular stents, and the minimal invasiveness of a laparoscopic procedure. This technique was adopted by Wang et al., who reported their experience with 17 patients, claiming 100% success and no complications after 2-year follow-up [25]. Today, this seems the least invasive, most promising option, awaiting more definitive studies.

Our patient presented with the classical picture of post weight loss abdominal pain and vomiting, thus SMA syndrome was high on our list of differentials. Other possible diagnoses include median arcuate ligament syndrome, congenital malformations like a duodenal atresia or gut malrotation, cholelithiasis, peptic ulcer disease, and less likely view his age an obstructing tumor or a mesenteric ischemia. The CTA confirmed our suspicion of SMA syndrome, but it also revealed a compressed and dilated LRV that was asymptomatic. Our postulation was that this compression was also the result of the recent weight loss and thus no sufficient time had yet elapsed for symptoms to appear. We also predicted it would improve in parallel to the SMA syndrome with weight regain. We therefore plan follow-up of the patient with a CT scan after proper weight regain; if the compression radiologically disappears, no further follow-up will be needed. If, however, it does not disappear, the patient will continue follow-up with yearly physical examinations and urinalysis to monitor for development of symptoms.

What was interesting about this case was that, as discussed, both entities share a common etiology: weight loss. The duodenum and the LRV, present in the same anatomical plane between the SMA and the aorta, are both at risk of compression when this plane is reduced. So, one might assume that as a person loses enough weight to cause one of the syndromes, it should not be uncommon to have the second syndrome develop as well. Looking at the literature, we were surprised to find only a few case reports of concomitant SMA and NCS [1,26–31]; this dual presentation is extremely rare, and our case adds importantly to the scant literature available.

Conclusions

SMA syndrome and NCS are both individually rare entities and although they share a common mechanism, their co-occurrence is extremely rare. SMA syndrome is to be suspected in patients with abdominal pain after weight loss. Conservative treatment with a focus on weight regain cures most cases. Asymptomatic or mildly symptomatic NCS is also treated conservatively. For patients requiring intervention, laparoscopic extravascular titanium stent placement seems to be the least invasive promising option today, awaiting further definitive studies.

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Conflict of interest

None.