

Nutcracker Syndrome Accompanying With Superior Mesenteric Artery Syndrome: A Case Report

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ABSTRACT

PURPOSE: The duodenum and the left renal vein (LRV) occupy the vascular angle made by the superior mesenteric artery (SMA) and the aorta. When the angle becomes too acute, compression of either structure can occur. Although superior mesenteric artery syndrome (SMAS) and renal Nutcracker syndrome (NCS) share the same pathogenesis, concurrent development has rarely been reported.

CASE REPORT: A 38-year-old female patient with a past history of gastrojejunostomy operated 6 years ago due to SMAS. She referred to vascular clinic with severe intermittent left-sided loin pain during the last 6 years. Computed tomography (CT)-angiogram and selective LRV angiogram with pressure gradient confirmed the diagnosis of NCS. She was treated by LRV transposition with uneventful recovery and considerable relief of symptoms.

CONCLUSIONS: NCS accompanying with SMAS is quite unusual. A patient, who first presents with clinical evidence of SMAS, could also simultaneously or sometime thereafter present with NCS and vice versa.

KEYWORDS: abdominal pain, LRV, NCS, SMA

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Introduction

Nutcracker syndrome (NCS) is characterized by impeded outflow from the left renal vein (LRV) into the inferior vena cava (IVC) due to extrinsic LRV compression.¹ Superior mesenteric artery syndrome (SMAS) (Wilkie's syndrome) is a rare condition whereby external compression of the third part of the duodenum by the SMA results in duodenal obstruction.^{2,3} The duodenum and the LRV occupy the vascular angle made by the SMA and the aorta.⁴ When the angle becomes too acute, compression of either structure can occur.⁴ Although SMAS and renal NCS share the same pathogenesis, concurrent development has rarely been reported.³ This article presents a case of a 38-year-old female with previous history of SMAS treated by gastroduodenostomy, who presented with left loin pain and microscopic hematuria caused by NCS which was treated by LRV transposition. To the best of our knowledge, only few cases in the literature reported NCS associated with SMAS.

Case Report

A 38-year-old female patient with a past history of gastrojejunostomy operated 6 years ago due to SMAS. She referred from urology clinics to vascular clinic with severe intermittent left-sided loin pain during the last 6 years. Physical examination was unremarkable. Laboratory tests were within normal ranges with the exception of a mild hematuria. Renal and abdominal ultrasounds (US) were normal. Esophageal gastro-duodenoscopy revealed patent anastomosis and no obvious pathology in the esophagus, stomach, and duodenum. Computed tomography (CT) abdomen and computed tomography angiography (CTA)

(venous and arterial phases) with three-dimensional reconstruction were performed (CTA examinations were performed using a 64 slices. The scanning parameters were slice thickness of 0.5 mm for 64 slices; pitch factor of 0.828 (helical pitch 53); rotation time of 0.4 s, and standard exposure modulation based on attenuation values of the two scout views (antero-posterior and latero-lateral) which showed decreased aortomesenteric distance and compression of LRV by SMA (Figure 1).

A selective venography of the LRV through femoral vein access revealed significant external compression of the LRV, markedly dilated pre-compression LRV and huge venous collaterals (Figure 2). Pressure gradient between LRV and the IVC was 5 mm Hg. The clinical history, hematuria, and previous history of SMAS were suggestive to establish the diagnosis of NCS confirmed by CTA findings, selective LRV venography, and significant pressure gradient between LRV and IVC.

LRV transposition was performed through mid-line laparotomy. The adrenal veins were scarified and the ovarian vein was preserved (Figure 3). She had significant symptomatic improvement after 6 weeks.

Discussion

SMAS and renal NCS are usually observed in females aged 10 to 40 years. The incidence of SMAS has been found to be between 0.013% and 0.3% of the general population.¹ NCS is a historically more recent clinical phenomenon than SMAS.⁴ Given that both the duodenum and the LRV lie within the aortomesenteric angle, two different types of compression syndrome can arise.⁴ However, both syndromes occurring concurrently are highly



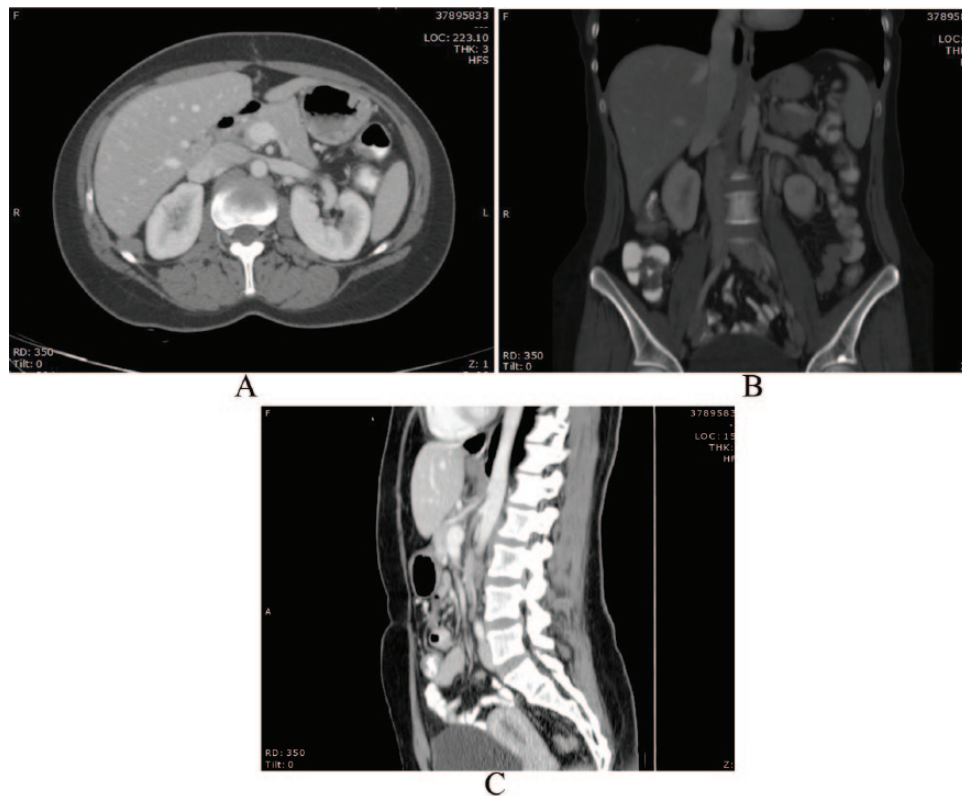


Figure 1. (A) Axial CT scan showing compressed LRV at decreased aortomesenteric angle. (B) Sagittal CT scan showing the reduction of the aortomesenteric angle and compressed LRV. (C) Coronal CT scan showing dilatation of the left gonadal vein. LRV indicates left renal vein.

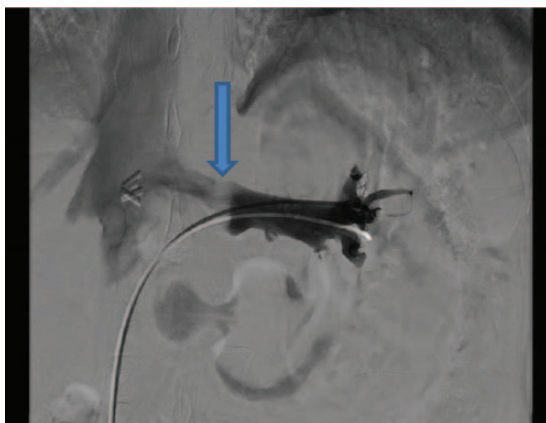


Figure 2. Selective LRV venogram showing severe LRV compression between the abdominal aorta and superior mesenteric artery. LRV indicates left renal vein.

rare.³ Generally, there are two main types of NCS, described as anterior and posterior NCS. In anterior NCS, the LRV is compressed between the abdominal aorta and SMA. The second (posterior) type results in the narrowing of the LRV compression between the aorta and the vertebral column.⁵

Each type of compression is associated with specific clinical symptoms that constitute a rare disorder.⁴ SMAS is a rare cause of abdominal pain, nausea, and vomiting.¹ NCS could be asymptomatic or could give rise to various manifestations.⁶ The most common clinical symptoms are hematuria, left flank abdominal pain, pelvic congestion syndrome, and left-sided varicocele.^{2,7,8} Hematuria

is a typical symptom occurring due to rupture of the thin walled septum separating the urinary collecting system veins. Orthostatic proteinuria, vague flank pain, and hypertension are other clinical symptoms in patients with NCS.⁷ Notably, symptomatic cases are called NCS, whereas non-symptomatic patients may be described having Nutcracker phenomenon.⁶ Gastro-intestinal (GI) symptoms are not common in cases of LRV compression; therefore, the presence of such symptoms in patients diagnosed with NCS should alert the physician to the possibility of a double compression, prompting the need for further investigation.⁴

The diagnosis of SMAS is difficult, particularly because of its often insidious and non-specific presentation. Blood tests may show electrolyte disturbance. Plain abdominal x-ray (AXR) may show marked gastric dilation. Barium upper GI series may show compression of the third part of the duodenum with proximal dilation, distal collapse, and delay in overall transit time. Upper GI endoscopy may reveal narrowing of the third part of the duodenum. CT, the gold standard, demonstrates the close proximity of the SMA and the aorta, and the caliber change of the duodenum.⁹ An SMA-aorta angle of 22° to 25° or less and a distance of 8 mm have been shown to correlate well with the symptoms of SMAS.¹⁰ However, imaging, such as US, Doppler US, CT, or magnetic resonance imaging (MRI) and invasive selective left renal phlebography with the measurement of the pressure gradient between the LRV and IVC, is required to diagnose NCS.¹

Surgical treatment of SMAS includes gastrojejunostomy, duodenojejunostomy, and division of the ligament of Treitz

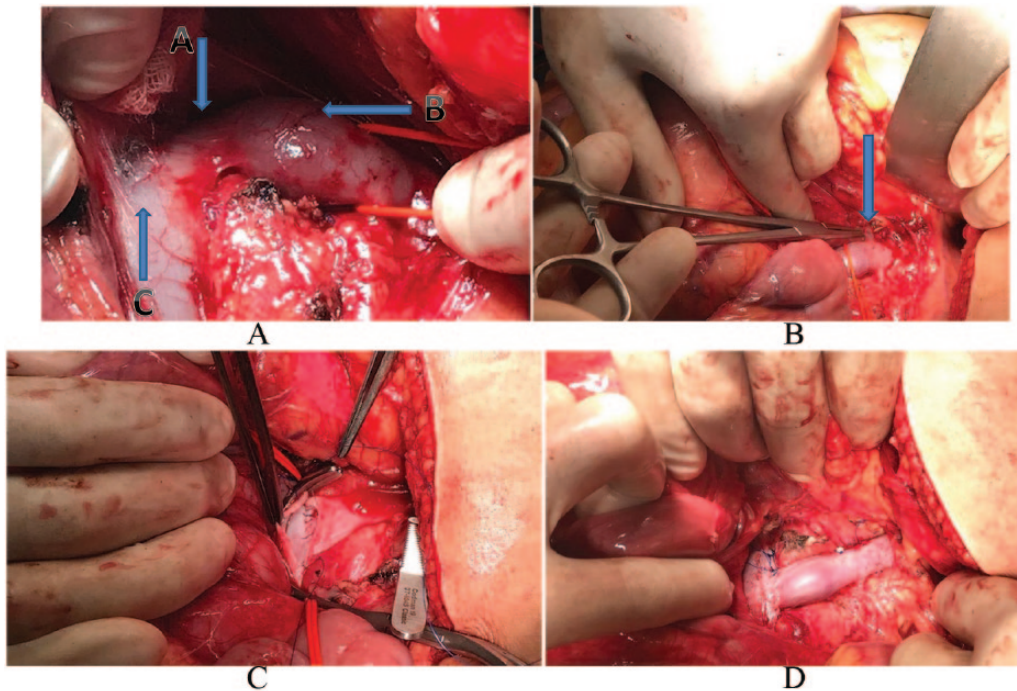


Figure 3. Intra-operative images: (A) Shows the A. aortomesenteric angle, B. LRV and C. the IVC. (B) Shows the adrenal vein. (C) Shows the partial clamp of the IVC and the anastomosis between the LRV and the IVC after transposition. (D) Shows the new site of the LRV on the IVC. IVC indicates inferior vena cava; LRV, Left Renal Vein.

with duodenal mobilization.⁹ Regarding NCS, both stenting and open surgical intervention for correcting anatomical anomaly, including procedures such as transposition of the LRV or SMA, nephropexy, intravascular and extravascular stent implantation, gonadocaval bypass, renal autotransplantation, and nephrectomy, are indicated for patients with significant pain, renal insufficiency, and severe, persistent life-threatening hematuria. However, selection criteria are not well defined.¹¹

In literature review, similar cases were described. Mikail Inal et al¹ described a 28-year-old man with SMAS and NCS treated conservatively. Rebeca Heidbreder⁴ reported a 20-year-old woman with SMAS and NCS required Roux-en-Y duodenojejunostomy and LRV transposition. Rebecca Nunn et al² reported a 19-year-old female with SMAS with coexisting Nutcracker phenomenon which was treated conservatively. Myung Jin Oh³ described a 23-year-old male treated by laparoscopic duodenojejunostomy.

Conclusions

NCS accompanying with SMAS is quite unusual. A patient, who first presents with clinical evidence of SMAS, could also simultaneously or sometime thereafter present with NCS and vice versa.

Author Contributions

Conceived and designed the experiments: NAA. Analyzed the data: NAA. Wrote the first draft of the manuscript: NAA. Contributed to the writing of the manuscript: NAA. Agree with manuscript results and conclusions: NAA. Jointly developed the structure and arguments for the paper: NAA. Made critical revisions and approved final version: NAA. The author reviewed and approved of the final manuscript.

Patient consent

Patient consent to publish the case study was obtained.

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