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Case Report

Coexistence of superior mesenteric artery syndrome and nutcracker phenomenon☆,☆☆

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ABSTRACT

Superior mesenteric artery (SMA) syndrome is a rare cause of upper gastrointestinal obstruction. Nutcracker syndrome occurs as the result of external compression of the left renal vein. Although they share a similar pathophysiology, SMA and nutcracker syndrome occurring simultaneously is rare. In this case report, we discuss the pathophysiology and unique computed tomography findings in a 25-year-old female patient diagnosed with SMA syndrome who was also incidentally found to have a coexisting nutcracker phenomenon. Due to similar pathogenesis, radiologists should consider the possibility of coexistence of these rare syndromes in appropriate patients.

Introduction

Superior mesenteric artery syndrome is due a to decreased angle between the origin of the superior mesenteric artery and the abdominal aorta leading to narrowing of the second portion of duodenum and resulting bowel obstruction [1–4]. Nutcracker syndrome is similar in which the decreased angle between the superior mesenteric artery and the aorta causes external compression of the left renal vein [5–7]. This leads to backflow congestion of the left gonadal vein due its drainage into the left renal vein. These abnormalities can be effectively identified using computed tomography (CT) which can effectively evaluate the abdominal viscera and the vasculature.

Case report

The patient is a 25-year-old female with no significant past medical history who presented to the emergency department due to acute onset of abdominal pain associated with nausea, bilious emesis, and diarrhea. She had had a similar transient episode previously, which resolved spontaneously. On

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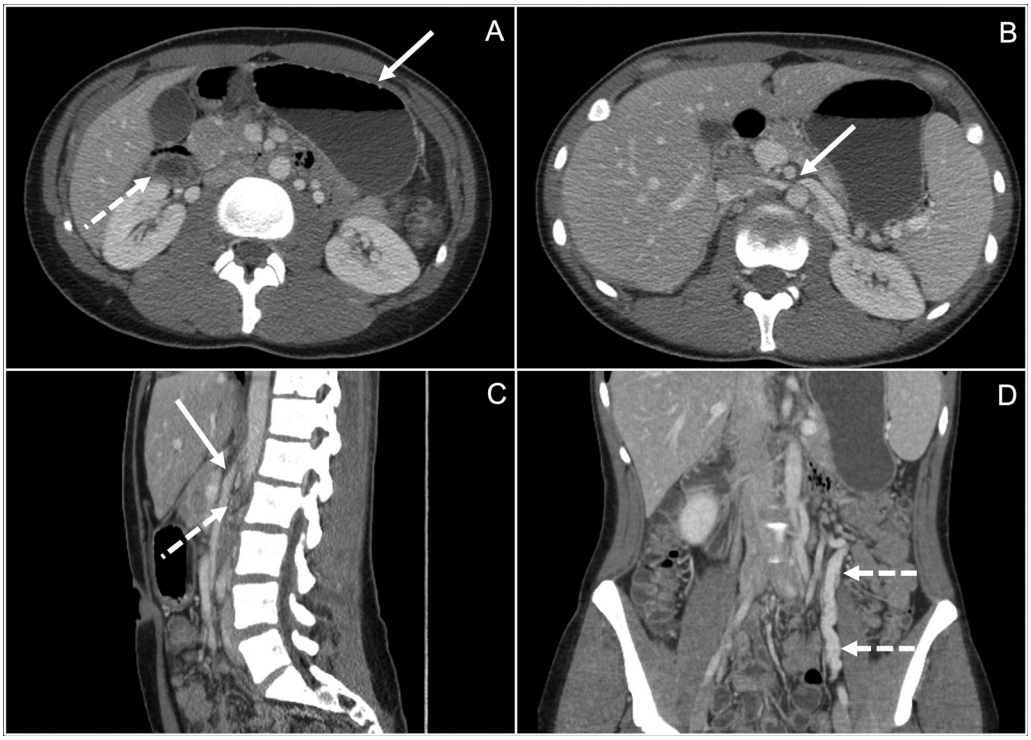

Fig. 1 – Axial CT image (A) at the level of the kidneys showing a distended and dilated stomach (solid arrow) and a distended second portion of the duodenum (dashed arrow). Sagittal image (C) at the level of origin of SMA from the aorta showing a narrow angle (solid arrow) and the collapsed third portion of the duodenum (dashed arrow). Axial image (B) is showing narrowing of the left renal vein as it crosses to the left between the SMA and the aorta. Coronal image (D) shows dilated left gonadal vein (dashed arrows).

physical exam, the patient has mild tenderness to palpation in the epigastric region along with findings consistent with dehydration. She had her symptoms quickly evaluated with a CT abdomen and pelvis with contrast. CT findings showed a significantly distended stomach along with dilated first and second parts of the duodenum with abrupt transition to collapsed duodenum across the midline (Fig. 1A and C). This small bowel obstruction was seen as a result of compression of the third part of the duodenum between the aorta and the superior mesenteric artery. Concomitantly, there was narrowing of the left renal vein in the region where it coursed between the superior mesenteric artery (SMA) and the aorta resulting in dilatation of the remaining left renal vein and left-sided pelvic congestion, suggestive of nutcracker phenomenon (Fig. 1B and D). Patient was referred to surgery and conservative management was started promptly.

Discussion

Superior mesenteric artery syndrome was first described by Rokitansky in 1842 as external compression of the third segment of the duodenum by the superior mesenteric artery [3]. The incidence of this condition is approximately 0.013%-0.3% in the general population. The commonly affected patient groups are those who undergo recent weight loss, eating disorders or dieting, post-scoliosis surgery, and burns. Females are more commonly affected, with age groups ranging from 10 to 39 years [3,4]. The most common presenting symptoms are vague post-prandial pain, nausea, vomiting and duodenal obstruction. There are several mechanisms described for the pathophysiology of developing SMA syndrome. These include loss of mesenteric fat planes which reduce the cushioning ability to protect from vascular compression [3,4]. Anatomical anomalies include short suspensory ligament of Treitz which results in high suspension of the duodenojejunal flexure, intestinal malrotation, and increased lumbar lordosis. Pressure from external fittings such as body casting also plays its role in rare cases. The most widely accepted computerized tomography and magnetic resonance angiographic imaging findings to describe SMA syndrome is visualization of vascular compression of the duodenum, measurement of aortomesenteric distance (reduced to 2-8 mm compared to typical 10-34 mm) and acute aortomesenteric angle (reduced to 6° to 22° compared to typical 38° to 56°) [5].

Similar pathophysiologic vascular compression and imaging characteristics are also described for nutcracker syndrome (Fig. 2) which includes compression of the left renal vein by the vertically traversing superior mesenteric artery, which results in impaired venous outflow (*anterior Nutcracker*). Compression can also occur between the aorta and vertebral bodies in the case of the anatomical variant of retro-aortic left renal vein (*posterior Nutcracker*). The most common manifestations of this phenomenon include left flank pain, hematuria, and venous hypertension. Radiographic features are similar on CT, MRI,

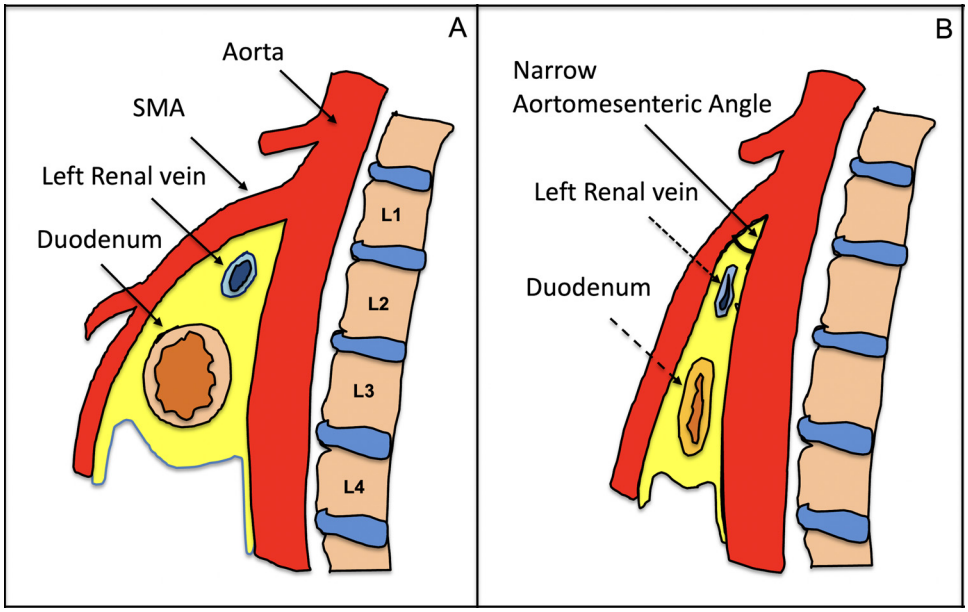

Fig. 2 – Line diagram showing the normal anatomical relationship (A) between the SMA, aorta, duodenum, and left renal vein. A narrow angle of SMA origin can lead to both narrowing of the left renal vein and compression of the third portion of the duodenum, as seen in B.

ultrasound, Doppler ultrasound, and conventional angiography, and include: reduced aortic-SMA angle (normal angle between aorta and SMA is ~45°, with a range of 38-65°), left renal vein stenosis, delayed left sided nephogram, early enhancement of collateral pathway which is left gonadal vein during portal venous phase and rarely formation of left splenorenal shunts [5–7]. Compression ratio (CR) is given by the anteroposterior diameter of the precompressed vein (**P**) divided by that of the compressed vein (**C**); namely, CR = P/C. A compression ratio above 2.25 is highly sensitive and specific for nutcracker syndrome [7]. The terminology of Nutcracker phenomenon/anatomy is used when there is presence of imaging findings in an asymptomatic patient [6].

The coexistence of both SMA and Nutcracker syndrome has rarely been reported and their clinical diagnosis remains challenging [8–10]. Management for both conditions starts with conservative management. For SMA syndrome, focus on weight regain is the priority: duodenal and gastric decompression for symptom relief, fluid resuscitation, and correction of electrolyte imbalances, with aims of optimizing nutrition via either enteral jejunal feed, TPN, or a combination of both [1,2]. Surgical options as a last resort include gastrojejunostomy, duodenojejunostomy, or Strong's operation. Invasive options for nutcracker syndrome include stent placement in the left renal vein for relief of obstruction.

Conclusion

Due to similar pathophysiology involving compression between the SMA and the aorta, coexistence of SMA and nutcracker syndrome should be considered based on imaging findings, in appropriate patients.

Patient consent

A written informed consent was obtained from the patient for the publication of this case report.

Authorship

The authors declare that this is their original work and they all approve the content of this manuscript. They confirm that this manuscript has not been published previously, in any language, in whole or in part, and is not currently under consideration elsewhere.

Ethical clearance

This project did not involve any research and no ethical clearance was required.

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