OPEN

Superior Mesenteric Artery Syndrome and Nutcracker Syndrome as the Presentation of Crohn's Disease in a Young Patient

A Case Report and Review of Literature

*Breanna Belle Lao, MD, †Anna Margaret Pitts, MD, ‡Aby Thomas, MD, and §Sandra M. Camacho, MD

Abstract: Superior mesenteric artery syndrome and nutcracker syndrome are rare vascular complications most often seen after marked weight loss caused by compression of the duodenum and left renal vein between the superior mesenteric artery and the aorta, respectively. The coexistence of superior mesenteric artery syndrome and nutcracker syndrome has been rarely reported. Herein, we present the case of a 16-year-old male with intermittent periumbilical abdominal pain, bilious vomiting, and weight loss who was found to have both of these vascular complications of significant weight loss as the initial presentation of Crohn's disease. This report provides insight into the diagnosis and treatment of these syndromes while highlighting the importance for practitioners to keep vascular complications on their differential diagnosis of vomiting and abdominal pain in patients with Crohn's disease.

Keywords: Crohn's disease, nutcracker syndrome, superior mesenteric artery syndrome

INTRODUCTION

Superior mesenteric artery syndrome (SMAS) is the compression of the third portion of the duodenum between the abdominal aorta posteriorly and the superior mesenteric artery (SMA) anteriorly (1,2). SMAS has an incidence of up to 0.3% (2–4), with symptoms of chronic intermittent abdominal pain, vomiting, nausea, early satiety, and anorexia (2). Risk factors include acute weight loss, rapid growth without weight gain, orthopedic surgical casting, acute gastroenteritis, or neurological injury (1,2). Nutcracker syndrome (NCS) may co-exist with SMAS (5–8). NCS results from compression of the left renal vein, resulting in flank pain, hematuria, proteinuria, calciuria, pelvic vein congestion, and hypertension (9–13).

SMAS has been described in patients with Crohn's disease (CD) (14,15), and both SMAS and NCS were described in pediatric patients (5–8,16). However, none of the reports included patients with both syndromes as the initial presentation of CD. Herein, we report a healthy 16-year-old male who was found to have SMA and NCS syndromes at the time of the diagnosis of CD.

CASE REPORT

A 16-year-old male presented to his pediatrician with several years of periumbilical pain and a new onset of bilious emesis. An abdominal x-ray showed a significant stool burden concerning for constipation and was treated with an enema and osmotic laxatives. However, the pain and bilious emesis progressed, prompting evaluation in the emergency department. He reported weight loss (3.2 kg) the week before presentation with a history of normal weight gain and growth curve (Fig. 1). He denied the use of any medications or supplements. Family history was significant for paternal CD. A computed tomography (CT) scan revealed findings consistent with SMAS and constriction of the left renal vein between the SMA and abdominal aorta (Fig. 2); an upper gastrointestinal series confirmed the findings of SMAS. Sagittal reconstruction on the CT scan demonstrated reduced distance between the SMA and the aorta (4.2 mm, normal range 6.6–16.1 mm) (17) with a narrowed aortomesenteric angle, (17°; normal range 26–65°) (17). Compression of the left renal vein was seen, resulting in a "beak" sign (Fig. 3A). He was referred to our institution for further evaluation. Examination at presentation was significant for ill appearance with thin body habitus, hypertension (133/88 mmHg), hypoactive bowel sounds, generalized abdominal tenderness to palpation, and normal perianal and rectal examination. A nasogastric tube was placed for bowel decompression.

Laboratory testing revealed normal values for complete blood count, liver panel, cystatin C, C-reactive protein, erythrocyte sedimentation rate, thyroid-stimulating hormone, free T4, tissue transglutaminase-IgA, total IgA, and lipase. Significant abnormal laboratory findings included elevated serum creatinine (1.3 mg/dL), proteinuria (10 mg/dL), and elevated fecal calprotectin (881 mcg/gm; normal <50 mcg/gm) (Fig. 4). Due to persistent hypertension, ultrasound with Doppler flow confirmed the diagnosis of NCS (Fig. 3A). Esophagogastroduodenoscopy showed significant gastritis and duodenitis (Fig. 5) with an extrinsic compression in the third portion of the duodenum resulting in a narrowed lumen prohibiting passage of the endoscope. Colonoscopy showed the terminal ileum was normal. The biopsies showed active duodenitis characterized by inflammation within the lamina propria, villous distortion, and focal gastric foveolar metaplasia; the colonic mucosa was visually normal but showed scattered lymphoid aggregates in the biopsies. Magnetic resonance enterography (MRE) showed diffuse circumferential wall thickening, restricted diffusion, and abnormal enhancement of the stomach, first and second portions of the duodenum, and multiple loops of the ileum. He was diagnosed with CD based on history, endoscopy, and image results with Paris Classification of A1bL4a/L4bB3G0.

Received March 30, 2023; accepted July 27, 2023.

From the *Department of Pediatrics, University of Mississippi Medical Center, Jackson, MS; †Department of Pediatrics, The University of Texas, Austin, TX; ‡Division of Pediatric Imaging, Department of Radiology, University of Mississippi Medical Center, Jackson, MS; and §Division of Gastroenterology, Department of Pediatrics, University of Mississippi Medical Center, Jackson, MS.

Correspondence: Breanna Belle Lao, Department of Pediatrics, University of Mississippi Medical Center, 2500 N State St, Jackson, MS 39216. E-mail: blao@umc.edu

The authors report no conflicts of interest.

JPGN Reports (2023) 4:4(e357)
ISSN: 2691-171X
DOI: 10.1097/PG9.0000000000000357

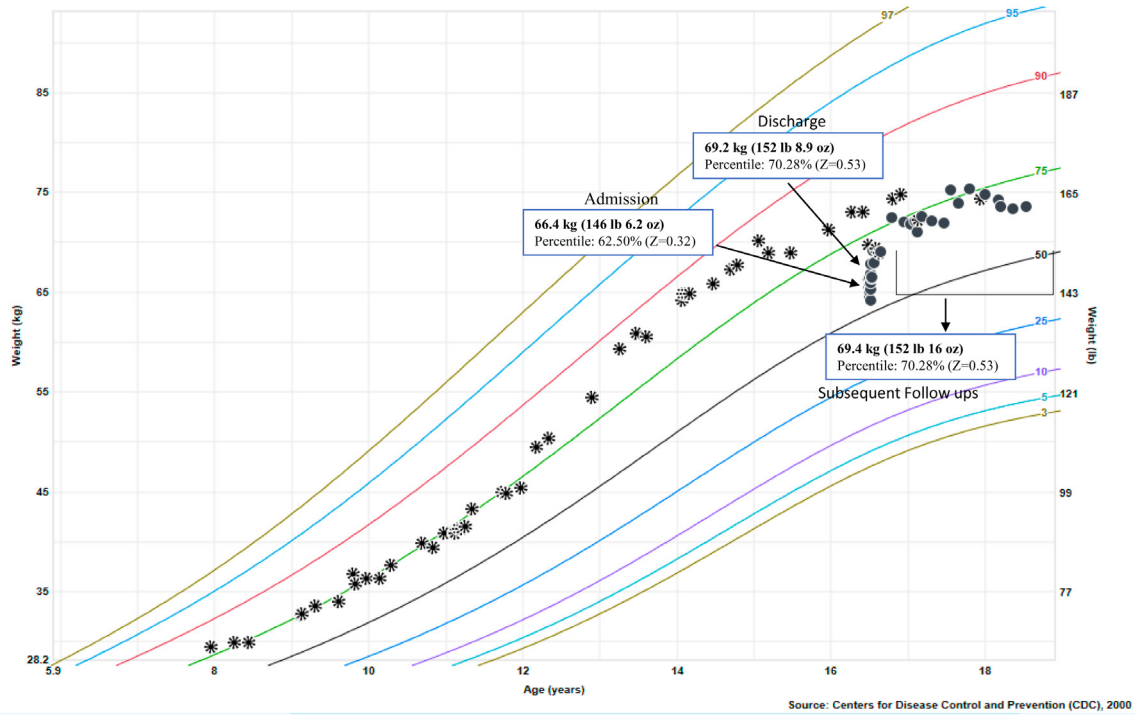

FIGURE 1. Growth chart showing weight before hospitalization, at admission, discharge, and subsequent follow-up visits. Z score is denoted in text. *Outside hospital system data point. •Within hospital system data point.

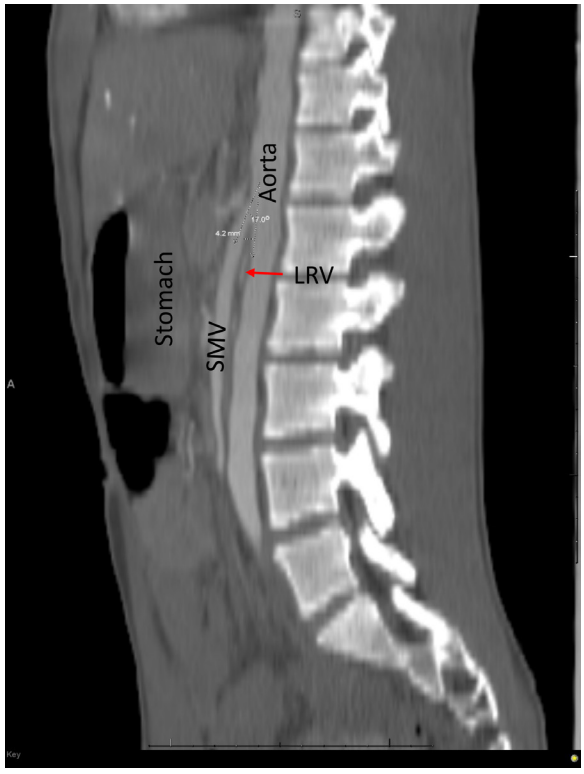

FIGURE 2. Sagittal reconstructed computed tomography (CT) image showing dilated stomach and duodenum with air-fluid level, a severely compressed left renal vein (LRV), narrowed aortomesenteric angle of 17° (normal 26–65°), and reduced aortomesenteric distance at the level of the duodenum of 4.2 mm (normal 6.6–16.1 mm).

A nasal duodenal-jejunal junction tube was used for enteral nutrition, and feeds were started based upon recommendations received from pediatric registered dieticians. CD treatment was started with corticosteroids and infliximab. Serial blood pressures were obtained throughout the stay. After 2 weeks, vomiting, abdominal pain, and hypertension improved, and he was discharged. He was weaned off steroids and continued infliximab infusions. CT scan 1 month later showed resolution of both SMAS and NCS, and the nasal duodenal-jejunal junction tube was removed at that time. By then, he regained the previous weight loss, and abdominal pain and hypertension were resolved (Fig. 1).

DISCUSSION

Concurrent SMAS and NCS have been reported in the pediatric literature with rapid weight loss, being underweight, or postsurgical changes that shorten the aortomesenteric distance and narrow the angle (5–8). The patient described herein showed clinical improvement after treatment for CD despite no change in weight. We hypothesize that the pathophysiology of these syndromes, in this case, was multifactorial. He had rapid weight loss, which is a well-established cause of these syndromes. Changes in the velocity of intestinal blood may have played a role, as shown by Giovagnorio et al, (18) who demonstrated postprandial resistive changes in the SMA through Doppler sonography in patients with active CD (11,18). Extensive inflammation of the bowel wall resulting in reduced mesenteric and retroperitoneal fat is documented in patients with a severe wasting disease with a hypermetabolic state from chronic illness (19), such as CD.

The diagnosis of SMAS can be confirmed by upper gastrointestinal series, enhanced CT, MRE, or laparotomy (2). The diagnosis of CD was rendered in this case based on the abnormal findings in the MRE and biopsies obtained. Wong et al (14) described the use of esophagogastroduodenoscopy as part of the evaluation in a pediatric patient with an uncertain etiology of SMA until a final diagnosis of CD was made.

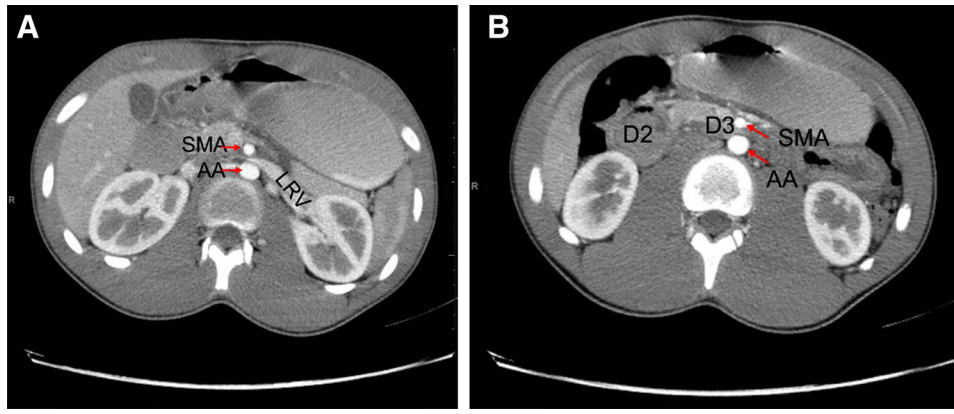

FIGURE 3. Axial contrast-enhanced CT images demonstrating the compression of the left renal vein (LRV) between the superior mesenteric artery (SMA) and abdominal aorta (AA) resulting in the "beak sign" (A), and compression of the duodenum (D3) between the SMA and AA. Thickening of the descending portion of the duodenum (D2) is also noted (B).

Test	Initial	Follow up	Reference range
albumin (g/dL)	3.7		3.7- 5.6 g/dL
pre-albumin (mg/dL)	14.0 (L)		18.0-35.7 mg/dL
lipase (U/L)	54		10-95 U/L
free T4 (ng/dL)	1.600		0.93-1.600 ng/dL
TSH (mclU/mL)	1.69		0.53-3.59 mclU/mL
serum IgA (mg/dL)	223		61-348 mg/dL
tissue transglutaminase IgA (U/mL)	0.4		< 7 U/L
c-reactive protein (mg/dL)	0.60	<0.50	<1.00 mg/dL
erythrocyte sedimentation rate (mm/hr)	15.0 (H)	25.0 (H)	0.0-10.0 mm/hr
creatinine kinase (U/L)	68		38-174 U/L
creatinine serum (mg/dL)	1.16 (H)	0.83 (H)	0.20-0.70 mg/dL
urinalysis, protein (mg/dL)	10.0		negative
cystatin C (mg/L)	0.73		0.61-0.95 mg/L
H. pylori antigen, stool	Negative		negative
fecal calprotectin (mcg/g)	881 (H)		< 50.0 mcg/g
fecal occult blood test	negative		negative

FIGURE 4. Diagnostic laboratory evaluation for SMAS, NCS, and CD. CD = Crohn's disease; NCS = nutcracker syndrome; SMAS = superior mesenteric artery syndrome.

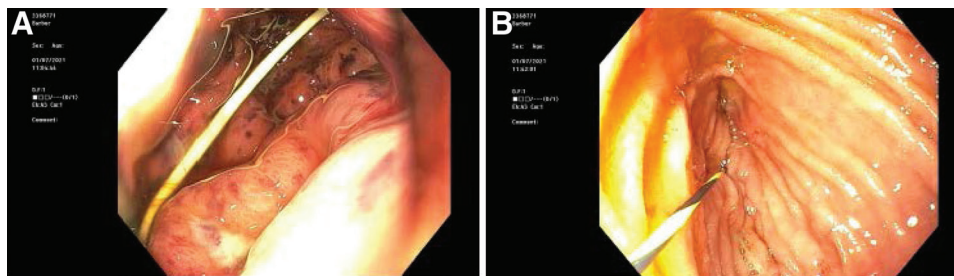

FIGURE 5. Esophagogastroduodenoscopy images showing severe gastritis with stigmata of bleeding and an enteric tube in place (A), and duodenitis of the third portion of the duodenum and a wire traversing the narrowed lumen (B).

Conservative treatment with bowel decompression, hydration, and nutritional support are the mainstays of management for patients with upper gastrointestinal obstruction caused by SMAS and NCS (2,5–9). Feeding management focuses on safe weight gain while monitoring for refeeding syndrome. Changes in feeding position are used to bypass the obstruction, including knee-chest position and left lateral decubitus position (1). Proteinuria, observed in NCS, often requires treatment with an ACE inhibitor (9). Most pediatric patients with SMAS and NCS have complete resolution of symptoms with medical management alone (2,5–8,11,16). If medical management fails, surgery may then be considered (2,9). Complications of SMAS include electrolyte imbalance, dehydration, hypovolemic shock, gastric perforation, pneumomediastinum, and even death (2,20–22). NCS can lead to severe anemia, irreversible renal damage, gonadal vein engorgement, pelvic congestion syndrome, and renal vein thrombosis (11,12).

To our knowledge, this is the first reported pediatric case of coexistent SMAS and NCS in the setting of newly diagnosed CD. In patients with CD presenting with abdominal pain, bilious vomiting, and significant weight loss, there should be high suspicion for SMAS. CT scan or MRE should be entertained in diagnosing SMAS, as both could discover a coexistent NCS.

ACKNOWLEDGMENTS

Informed parent consent and patient assent were obtained for the publication of case details.

REFERENCES

1. Burrington JD. Superior mesenteric artery syndrome in children. *Am J Dis Child*. 1976;130:1367–1370.
2. Biank V, Werlin S. Superior mesenteric artery syndrome in children: a 20-year experience. *J Pediatr Gastroenterol Nutr*. 2006;42:522–525.
3. Goin LS, Wilk SP. Intermittent arteriomesenteric occlusion of the duodenum. *Radiology*. 1956;67:729–737.
4. Anderson JR, Earnshaw PM, Fraser GM. Extrinsic compression of the third part of the duodenum. *Clin Radiol*. 1982;33:75–81.
5. Min KW, Lee OK, Kim MK. Nutcracker syndrome combined with superior mesenteric artery syndrome in a pediatric patient: a case report. *Child Kidney Dis*. 2018;22:75–80.
6. Diab S, Hayek F. 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:e 922619.
7. Lin TH, Lin CC, Tsai JD. Superior mesenteric artery syndrome and nutcracker syndrome. *Pediatr Neonatol*. 2020;61:351–352.
8. Wood D, Fagbemi A, Jago L, et al. Superior mesenteric artery and nutcracker syndromes in a healthy 14-year-old girl requiring surgical intervention after failed conservative management. *Gastro Open J*. 2020;5:1–3.
9. Meyer J, Rother U, Stehr M, et al. Nutcracker syndrome in children: appearance, diagnostics, and treatment - a systematic review. *J Pediatr Surg*. 2022;57:716–722.
10. Wang RF, Zhou CZ, Fu YQ, et al. Nutcracker syndrome accompanied by hypertension: a case report and literature review. *J Int Med Res*. 2021;49:300060520985733.
11. Kolber MK, Cui Z, Chen CK, et al. Nutcracker syndrome: diagnosis and therapy. *Cardiovasc Diagn Ther*. 2021;11:1140–1149.
12. Ananthan K, Onida S, Davies AH. Nutcracker syndrome: an update on current diagnostic criteria and management guidelines. *Eur J Vasc Endovasc Surg*. 2017;53:886–894.
13. Granata A, Distefano G, Sturiale A, et al. From nutcracker phenomenon to nutcracker syndrome: a pictorial review. *Diagnostics (Basel)*. 2021;11:101.
14. Wong JY, Vo N, Zabrowski D, et al. Superior mesenteric artery syndrome as a presenting sign of inflammatory bowel disease. *J Pediatr Gastroenterol Nutr*. 2020;70:e51–e52.
15. Cicero G, D'Angelo T, Bottari A, et al. Superior mesenteric artery syndrome in patients with Crohn's disease: a description of 2 cases studied with a novel magnetic resonance enterography (MRE) Procedure. *Am J Case Rep*. 2018;19:431–437.
16. Wang C, Wang F, Zhao B, et al. Coexisting nutcracker phenomenon and superior mesenteric artery syndrome in a patient with IgA nephropathy: a case report. *Medicine (Baltimore)*. 2021;100:e26611.
17. Arthurs OJ, Mehta U, Set PA. Nutcracker and SMA syndromes: what is the normal SMA angle in children? *Eur J Radiol*. 2012;81:e854–e861.
18. Giovagnorio F, Diacinti D, Vernia P. Doppler sonography of the superior mesenteric artery in Crohn's disease. *AJR Am J Roentgenol*. 1998;170:123–126.
19. Pivawer G, Haller JO, Rabinowitz SS, et al. Superior mesenteric artery syndrome and its ramifications. *CMIG Extra: Cases*. 2004;28:8–10.
20. Ooi GC, Crundwell M, Ferrando J. Pneumomediastinum: an acute presentation of the superior mesenteric artery syndrome. *Australas Radiol*. 1996;40:447–449.
21. Ko KH, Tsai SH, Yu CY, et al. Unusual complication of superior mesenteric artery syndrome: spontaneous upper gastrointestinal bleeding with hypovolemic shock. *J Chin Med Assoc*. 2009;72:45–47.
22. Demir MK, Cevher T. A rare cause and complication of acute gastric dilatation: superior mesenteric artery syndrome and perforation. *Eurasian J Med*. 2018;50:60–61.