

Intractable Emesis in Children: A Pediatrician's Unusual Dilemma. A Case Report and Review of The Literature

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Abstract

Wilkie's syndrome, commonly referred to as Superior mesenteric artery syndrome, is an unusual encounter in the medical field. It could mimic a simple gastritis in an acute setting to a more serious clinical suspicion of bowel obstruction or neoplasia on a chronic onset. While this syndrome most commonly happens as a sporadic case, it can be favored by genetic predisposition of a congenital short ligament of Trietz or when the superior mesenteric artery is more distal than normal on the abdominal aorta. Inciting medical factors can contribute to its etiology or simply, it could happen as a result of surgical complication. Recent literature reveals that the superior mesenteric artery may co-occur with other phenomena, such as Nutcracker syndrome, as both share similar anatomic levels. In our report, we describe a case of a 10-year-old boy of Middle Eastern descent who was brought to the emergency department with repeated vomiting that had started one month ago, accompanied by abdominal pain and change in bowel habits that was initially attributed to gastroenteritis. After multiple and failed conservative attempts from different pediatricians in the team, a contrast-enhanced abdominal CT confirmed the presence of superior mesenteric artery syndrome that happened as a result of rapid weight loss due to his gastroenteritis. Early diagnosis of superior mesenteric artery syndrome is a crucial step in its management to prevent serious health-related repercussions.

Keywords: Superior Mesenteric Artery Syndrome (SMAS), Wilkie's Syndrome, Nutcracker Syndrome, Small Bowel Obstruction

1. Introduction

The superior mesenteric artery syndrome (SMAS) was first coined by Rokitsky when he described an autopsy finding and published about it in 1842[1,2]. Later, Wilkie published a large case series involving 75 patients with this syndrome; hence, both names were attributed to the syndrome [3]. The main pathophysiology involved in this condition is the compression of the third part of the duodenum between the superior mesenteric artery and the aorta due to loss of the mesenteric fat pad between both structures, which is caused by weight loss. In normal patients, the angle between the superior mesenteric artery and aorta (aorto-mesenteric angle) ranges between 38° and 65°, depending on the person's body mass index, while the aorto-mesenteric distance should not exceed 10 mm. The most sensitive radiologic measure of diagnosis is decreased aorto-mesenteric angle to less than 25° (below 20°, patients develop symptoms of bowel obstruction in adults; therefore, it is safe to presume that the pediatric age group has a lower cut-off angle before symptoms develop) and a decline in the aorto-mesenteric distance to less than 8 mm along

with marked dilation of the proximal duodenum compared to the compressed segment. SMAS shows a female predominance and is more common in thin pediatric patients and adolescents. Clinical complaints present with a constellation of three main symptoms of postprandial pain in almost 88.9% of patients, vomiting in 55.6% of patients, and early satiety in 51.9% [4]. Many modalities are used to diagnose superior mesenteric artery syndrome, including barium series, abdominal CT scan, abdominal angiography, and magnetic resonance arteriography [1]. Radiologic imaging, such as CT scan with contrast, is paramount to rule out other differential diagnoses and assess the severity of the case by measuring the degree of duodenal compression, the quantity of adipose and lymphatic tissue, and other coexisting conditions such as nutcracker syndrome and abdominal aortic aneurysm. Endoscopy is sometimes performed to rule out SMAS mimics such as peptic ulcer disease or intestinal obstruction. Imaging modalities can give clinicians insight if the diagnosis of SMAS is 'suggestive' (reduced aorto-mesenteric angle and distance with proximal duodenal dilatation), 'possible' (reduced angle and distance

without proximal duodenal dilatation) and ‘probable’ (reduction of either angle or distance) [5].

2. Case Presentation

A 10-year-old boy previously healthy with recent travel history to Morocco one month ago and no past surgical history was brought to the emergency department complaining of repeated non-bilious, non-bloody and non-projectile vomiting, 3–4 times per day that started three days ago accompanied by diffuse abdominal pain without radiation, decreased bowel habit along with passage of semi-formed stools and weight loss of 6 kg over one-month duration. The patient was afebrile, tachycardiac, emaciated, moderately dehydrated, and complained of pain. Abdominal examination revealed a soft, mildly tender abdomen. He had three previous similar encounters in the pediatric unit within the last month with less severe symptoms, for which he was diagnosed as a case of gastroenteritis that was managed conservatively and improved. However, because of gastroenteritis that lasted for

almost three weeks, he lost a total of 6 kg within this period, when comparing his third visit with his first visit. Surprisingly, all his blood labs, stool tests and ultrasound findings were unremarkable. He was then booked for an abdominal CT scan. CT scans of the abdomen and pelvis with oral and IV contrast were performed. Scan findings revealed the aorto-mesenteric angle was reduced by 19° (Figure B and C) and the aorto-mesenteric distance decreased to 2 mm (Figure A). Consequent significant compression of the third part of the duodenum is noted with subsequent sluggish and delayed passage of the contrast column into the jejunum associated with significant and persistent dilation of the stomach and the first and second parts of the duodenum (Figure D, E and F). His presentation and imaging are relatively consistent with superior mesenteric artery syndrome that was predisposed by rapid weight loss due to infectious diarrhea he had in the earlier month; thus, his symptoms markedly improved with nutritional support and after putting on an additional weight of 3kg, the patient did not experience similar episodes during his 7-month follow-up.



Figure A: Axial CT of the abdomen with intravenous and oral contrast shows reduced aorto-mesenteric distance of 2 mm (normal > 8mm)

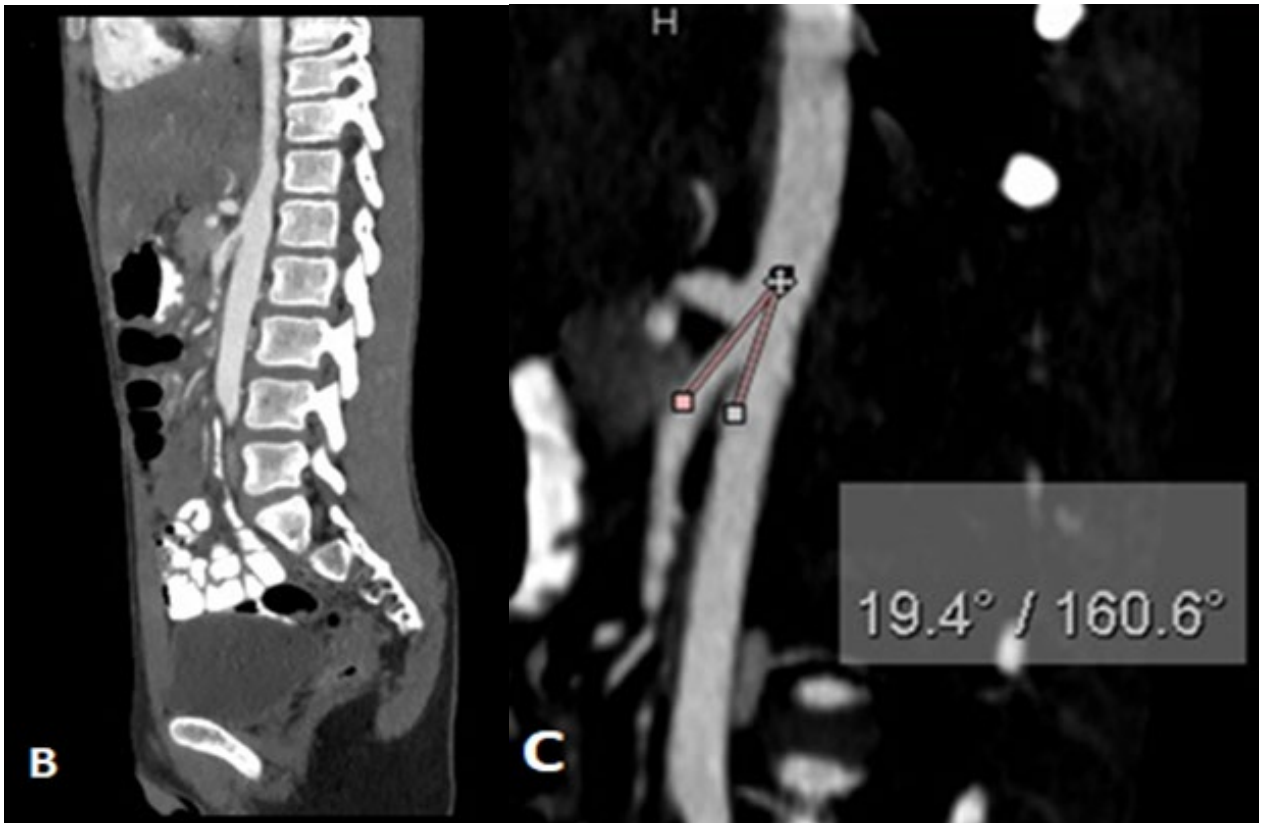


Figure B & C: Sagittal CT of the abdomen with intravenous and oral contrast shows reduced aorto-mesenteric angle measures 19.4° (normal >38°)

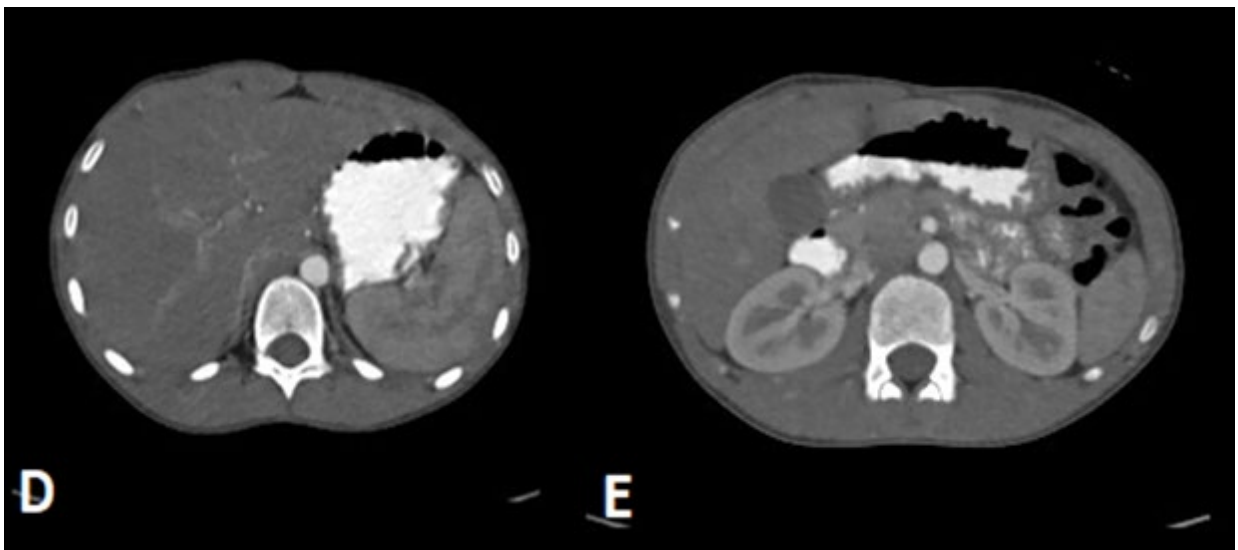


Figure D & E: Axial CT of the abdomen shows distended stomach and proximal portion of the duodenum



Figure F: Coronal CT of the abdomen shows distended 2nd part of the duodenum (large arrow) and collapsed 4th part of the duodenum (small arrow)

3. Discussion

Superior mesenteric artery syndrome is uncommon, with a prevalence ranging from 0.013% to 0.3% in the general population [6]. However, it usually accompanies changes in bowel habits, mostly constipation, which is a common presentation in the pediatric age group, and many studies have focused on clinical history rather than radiographic findings to identify the cause of constipation to prevent misdiagnosis [7]. Prolonged constipation and rapid weight loss are especially important for superior mesenteric artery syndrome. In patients with significant stomach discomfort, prolonged emesis, or bilious emesis, other diagnoses including obstructive conditions should be investigated. SMAS occurs when the superior mesenteric artery leaves the abdominal aorta at an excessively acute angle, often 22°–25°, and runs closer to the aorta than normal, often 2–8 mm, depending on the patient's BMI [8]. There are no absolute diagnostic numbers and imaging must be interpreted based on the patient's clinical history [8]. The early identification of predisposing and precipitating factors is crucial for diagnosis. Predisposing factors include an abnormal acute aorto-mesenteric angle, high fixation of the duodo-jejunal flexure relative to the ligament of Trietz, low origin of the superior mesenteric artery, lumbar lordosis, and supine position. In contrast, precipitating factors lead to marked and rapid weight loss, such as cancers, burn injuries, dietary disorders such as anorexia,

malabsorption, injuries to the head and spine, and postoperative state or deformities [9]. SMAS is managed conservatively by a multidisciplinary team of pediatricians, gastroenterologists, dietitians, radiologists, and surgeons. It involves NGT insertion for gastric decompression, feeding through total parenteral nutrition (TPN) or nasogastric tube, orexigenic medications, correction of any electrolyte abnormalities, anti-emetics or prokinetic drugs, lastly posturing maneuvers like the right decubitus and prone position which helps to relieve symptoms especially if the patient is tolerating oral feeds [8,10]. The success rate of conservative measures is 85%, and patients need to resort to surgical interventions only when these measures fail to improve symptoms. Laparoscopic duodeno-jejunostomy has proven to be an effective surgical procedure, with a success rate of up to 100%, along with the added advantage of decreased blood loss and early recovery time compared to other procedures [10].

4. Conclusion

Superior mesenteric artery syndrome is an unusual encounter in the medical field presenting with a range of symptoms that closely mimic other disorders, from collagen vascular diseases to small intestinal obstruction, posing a diagnostic dilemma for physicians. Prompt recognition of the syndrome is only attainable if physicians focus on clinical history, identify the predisposing

and precipitating factors, and perform necessary imaging. Management is done by a multidisciplinary team whose aim is to stabilize the patient and improve his/her nutritional status. Failure of a conservative approach to alleviate symptoms means surgical intervention should be exercised as a last resort.

Acknowledgement

Statement of Ethics

Ethics approval is not required. Written informed consent was obtained from participant's parents for publication of the details of their medical case and accompanying images.

Conflict of Interest

The authors have no conflicts of interest to declare.

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Authors' Contribution

Author 1 and author 2 both contributed in preparing the manuscript. Author 2 added the radiological images. All authors approved the final manuscript.

Data Availability Statement

The data that support the findings of this study are available in the figures shared for this study

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