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A case of Wilkie's syndrome

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Abstract
Superior Mesenteric Artery (SMA) Syndrome, also known as Wilkies syndrome, is a rare cause of duodenal obstruction resulting from compression of the third part of the duodenum between the superior mesenteric artery and the abdominal aorta. Despite its rarity, with a worldwide incidence of 0.1%, it represents an important differential diagnosis in young patients presenting with postprandial vomiting and abdominal pain.

Keywords: Superior mesenteric artery syndrome, Wilkies syndrome, duodenal obstruction, gastrojejunostomy, aorto-mesenteric angle

Introduction

Superior Mesenteric Artery (SMA) Syndrome, first described by Carl Von Rokitansky in 1842 and later studied in detail by Wilkie in 1927 [2], represents an uncommon yet well-recognized cause of chronic, intermittent, partial or complete duodenal obstruction [1,2]. The syndrome results from compression of the third part of the duodenum between the superior mesenteric artery and the abdominal aorta [3]. The condition has a worldwide incidence of approximately 0.1%, with a female predominance (female to male ratio of 3:2) and typically
affects individuals between 10 to 30 years of age [4,5]. The normal aorto-mesenteric angle ranges from 38 to 65 degrees, whereas in SMA syndrome, this angle is characteristically reduced to 9-22 degrees [6].
Multiple etiological factors have been identified, including constitutional thinness, anorexia nervosa, rapid weight loss due to catabolic states (burns, trauma, malignancies), malabsorption syndromes, severe injuries requiring prolonged bed rest, spinal deformities, familial predisposition, and idiopathic causes [7,8,9,10].

Case Summary

A 20-year-old female presented to our institution with a 6-month history of abdominal pain that was characteristically aggravated by food intake and relieved when lying in the prone position. She also reported postprandial vomiting occurring 1-2 episodes per day, containing undigested food particles.
On physical examination revealed a poorly built and poorly nourished patient. Anthropometric measurements showed: - Weight: 36 kg, Height: 1.4 m, Body Mass Index (BMI): 18.3 kg/m².
Vital signs were within normal limits. Abdominal examination revealed a soft abdomen with tenderness over the epigastric and umbilical regions. Bowel sounds were present and normal.
Initial laboratory investigations were within normal limits. Ultrasound abdomen and pelvis showed normal findings. Upper gastrointestinal endoscopy: Demonstrated gastric dilatation. Contrast-enhanced computed tomography (CECT) of abdomen and pelvis revealed a reduced angle between the superior mesenteric artery and aorta, measuring 14-19 degrees, resulting in compression of the third part of the duodenum. The proximal duodenum (D2) showed significant dilatation. Additionally, the superior mesenteric artery was observed to be indenting the left renal vein.

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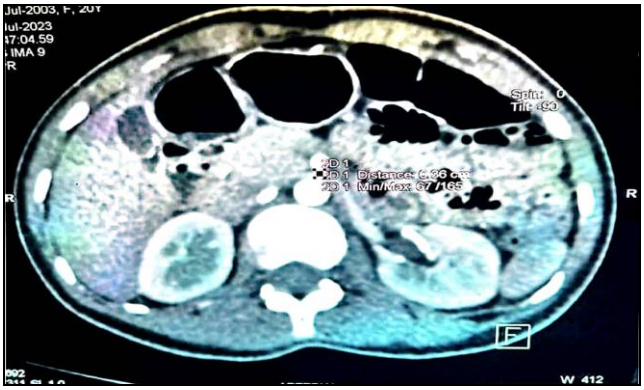

Fig 1

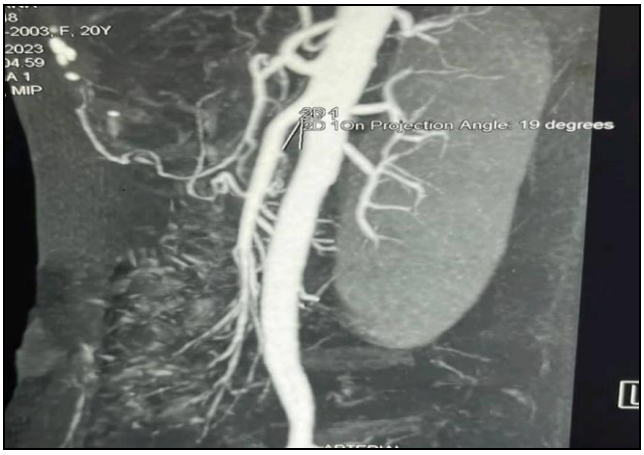

Fig 2

Surgical Intervention

Based on the clinical presentation and imaging findings, a diagnosis of SMA syndrome was established. Given the chronic nature of symptoms and failure of conservative management, surgical intervention was indicated.

The patient underwent posterior gastrojejunostomy.
Intraoperative Findings: Direct visualization confirmed compression of the third part of the duodenum by the superior mesenteric artery, corroborating the preoperative imaging findings.

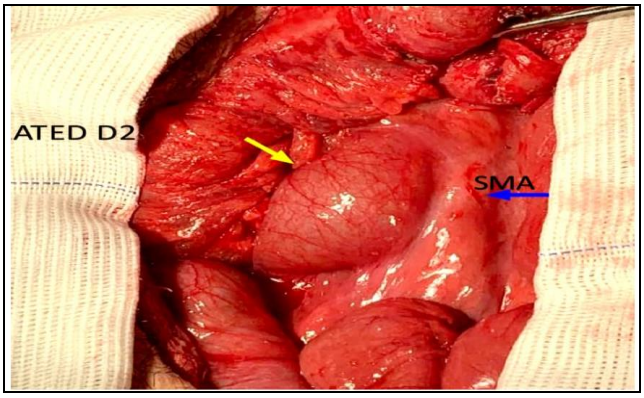

Fig 3

Postoperative Course
The postoperative period was uneventful. The patient was initiated on oral feeding on postoperative day 3, which was well tolerated. The surgical drain was removed on postoperative day 5. At 3-month follow-up, the patient remained completely asymptomatic with resolution of all presenting symptoms.

Discussion
SMA syndrome represents a challenging diagnostic entity that requires high clinical suspicion, particularly in young, thin individuals
presenting with characteristic symptoms [11]. The pathophysiology involves the loss of retroperitoneal fat and mesenteric fat pad, which normally provides cushioning between the superior mesenteric artery and the duodenum [12]. The classic clinical presentation includes abdominal pain and vomiting that are characteristically relieved by positioning in left lateral decubitus, prone, or knee-to-chest positions, and aggravated in the supine position [13,14]. This positional variation in symptoms is pathognomonic and should raise suspicion for the diagnosis. Initial management typically involves conservative treatment focusing on nutritional rehabilitation and correction of electrolyte abnormalities. However, in cases where conservative management fails or in patients with severe symptoms, surgical intervention becomes necessary [15].

Several surgical options are available for the management of refractory SMA syndrome
1. **Duodenojejunostomy:** Direct bypass of the obstruction
2. **Gastrojejunostomy:** As performed in our case
3. **Strong's procedure:** Mobilization of the duodenum

The choice of surgical procedure depends on individual patient factors and surgeon preference. In our case, posterior gastrojejunostomy was selected and resulted in excellent outcomes with complete symptom resolution.

Conclusion
SMA syndrome should be considered as a potential diagnosis in young patients presenting with chronic abdominal pain, postprandial vomiting, and weight loss, particularly in those with a thin body habitus. The diagnosis can be confirmed through appropriate imaging studies demonstrating a reduced aorto-mesenteric angle and duodenal compression. While conservative management may be attempted initially, surgical intervention often provides definitive treatment with excellent long-term outcomes, as demonstrated in our case.
Early recognition and appropriate management are crucial for preventing complications and improving patient quality of life. Healthcare
providers should maintain a high index of suspicion for this rare but treatable condition.

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