

Unraveling the Silent Crisis: A Rare Case of Acute Intestinal Obstruction Caused by Superior Mesenteric Artery Syndrome

ABSTRACT

Superior Mesenteric Artery (SMA) Syndrome, also known as Wilkie's Syndrome, is a rare and potentially life-threatening gastrovascular disorder characterised by the compression of the third portion of the duodenum between the abdominal aorta and the superior mesenteric artery. This case report presents a 22-year-old male with acute intestinal obstruction secondary to SMA syndrome. The patient, who had kyphoscoliosis and syndromic features, presented with sudden-onset epigastric pain, abdominal distension, and constipation. Imaging studies, including ultrasonography and contrast-enhanced CT, confirmed SMA syndrome with associated Nutcracker syndrome. Given the severity of obstruction, the patient underwent emergency exploratory laparotomy with Roux-en-Y duodeno-jejunostomy and jejunio-jejunostomy. Postoperatively, he showed a favorable recovery and was discharged on postoperative day 10. This case highlights the importance of early recognition and prompt surgical intervention in SMA syndrome, particularly in patients with anatomical predispositions. Delayed diagnosis can lead to severe complications such as malnutrition, electrolyte imbalance, and gastrointestinal perforation. A multidisciplinary approach is crucial for optimal patient management and long-term outcomes.

Introduction

Superior mesenteric artery (SMA) syndrome, also named as Wilkie's syndrome, arterio-mesenteric duodenal compression syndrome and cast syndrome, is a rare, potentially life-threatening gastrovascular disorder and an uncommon cause of proximal bowel obstruction characterised by a compression of the third and final portion of the duodenum by the abdominal aorta and the overlying superior mesenteric artery

Presentation of case

A 22 year old male patient presented with complaints of Abdominal pain from 1 day of sudden onset in the epigastric region which gradually progressed in intensity with no aggravating and relieving factors, patient also complaints of abdominal distension, constipation, obstipation from 1 day.

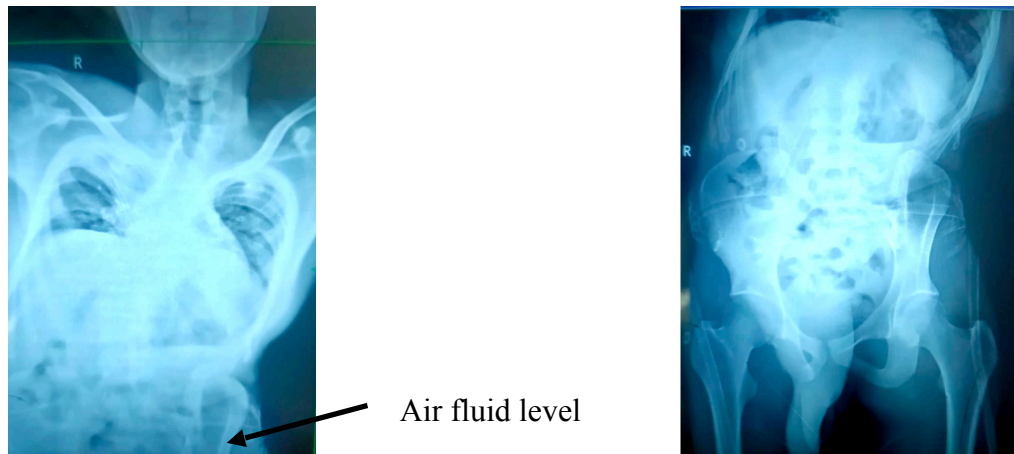
No history of vomiting, trauma to abdomen, weight loss

Patient had no known Co-morbidities and denies substance abuse

Patient has Kyphoscoliosis from birth with high arched palate having syndromic features.

On examination patient was vitally stable , per abdominal examination revealed distended abdomen with umbilicus displaced inferiorly ,guarding present in the epigastrium, left hypochondrium and umbilical region,Tenderness present in the epigastric region , left hypochondriac and umbilical region with no rigidity , Bowel sounds sluggish .Abdominal X-ray and chest rays were done and showed air fluid level

Fig 1: Abdominal X-ray and chest rays indicating air-fluid level



All Routine investigation was sent and was within normal limits

Nasogastric tube of 16 Fr was inserted and was continued to decompress the bowel

Foleys urinary catheter was inserted so as to monitor the intake and output

Patient was kept nil by mouth and was started on IV antibiotics, analgesics and Intravenous fluids

- Ultrasonography showed dilated bowel loops with to and fro peristalsis- likely suggestive of acute intestinal obstruction

Contrast enhanced CT scan of Abdomen and Pelvis was done which showed Gross dilatation of stomach , 1st and 2nd part of duodenum with abrupt cut off between 2nd and 3rd part of duodenum due to compression of 3rd part of duodenum with aorta and superior mesenteric artery at the level of vertebra L3

- Severe narrowing of aorta-mesenteric angle and aorto-mesenteric distance-Wilkie's syndrome
- Common origin of celiac and superior mesenteric artery
- Compression of left renal vein with abdominal aorta and superior mesenteric artery suggestive of Nutcracker syndrome

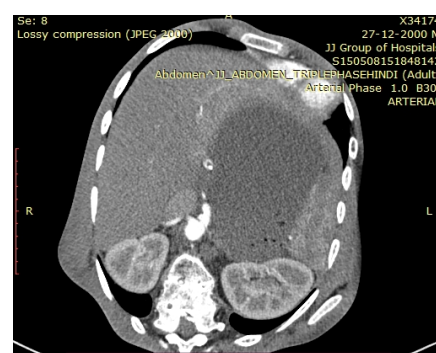
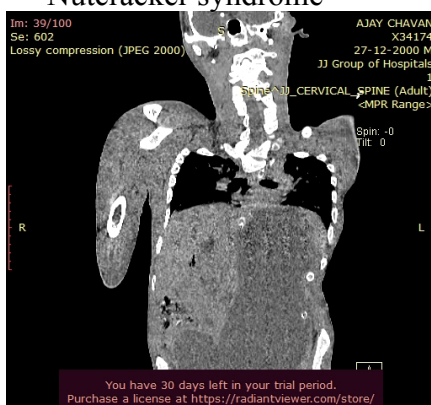


Fig 2: Contrast CT scan of the abdomen and pelvis showing gross dilatation of stomach

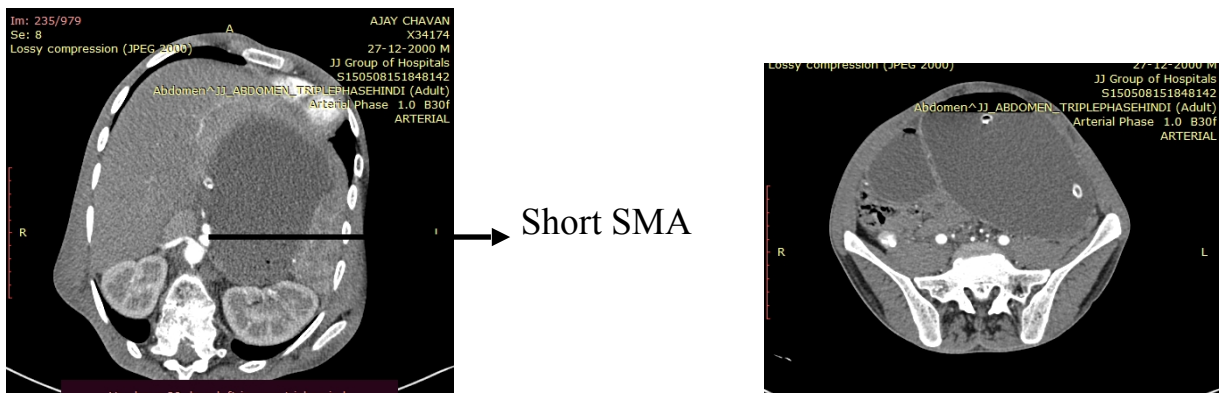


Fig 3: CT scan of the abdomen indicating superior mesenteric artery

- Oesophagoduodenoscopy was done which showed dilated stomach and 1st and 2nd part of duodenum

Patient and his relatives were explained about the complexities and the risk associated with the Disease and the urgent requirement of surgery with its risk and complications associated with it following their consent. Patient was taken up for Emergency Exploratory Laparotomy where in the stomach along with 1st and 2nd part of duodenum was grossly distended with collapsed 3rd part showing no gangrenous changes, 50 cm from the Duodeno-Jejunal flexure the jejunum was cut using blue linear cutting stapler and the second part of Duodenum Kocherised and side to side Roux-en Y duodeno-jejunostomy with side to side jejuno-jejunostomy and Abdominal drains were placed, near the site of anastomosis and the other at the pelvis

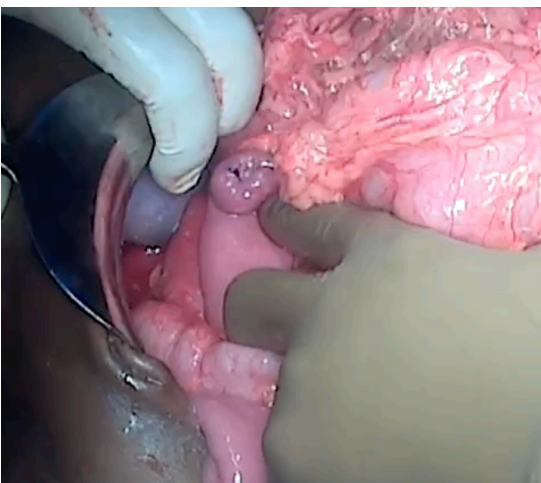


fig 4 : **Duodenojejunostomy**

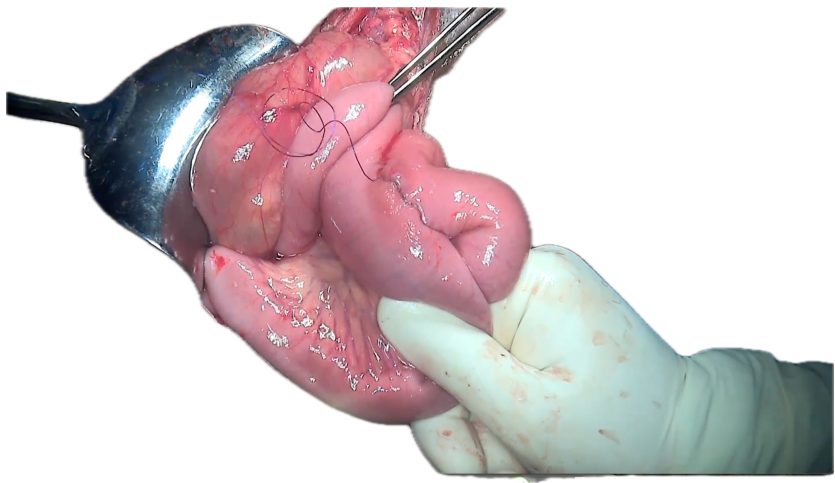
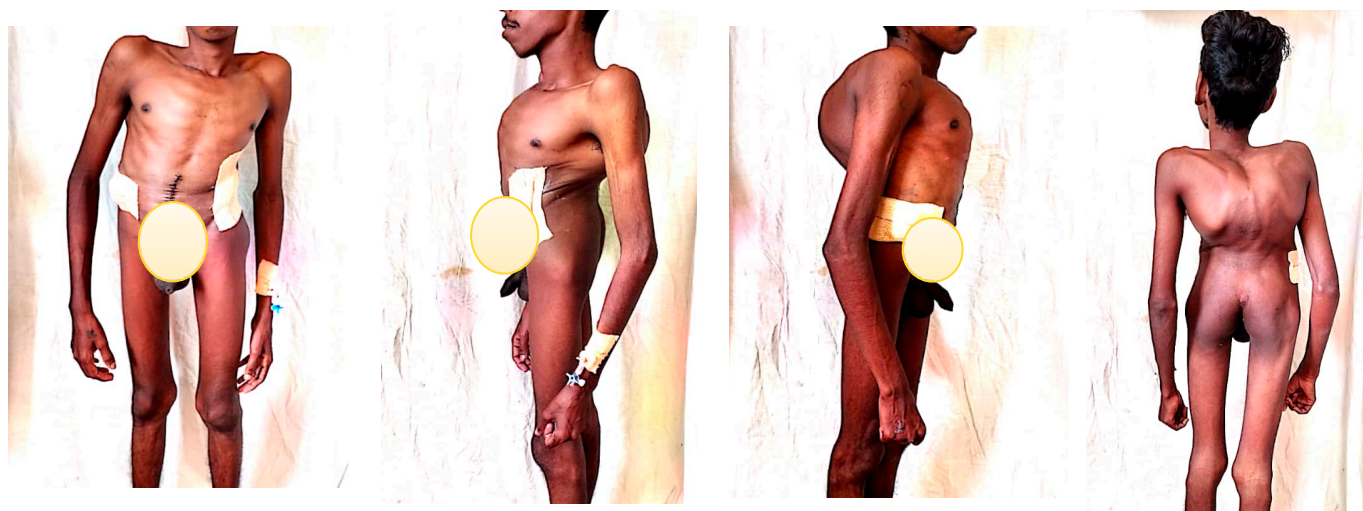


fig 5 : **Jejunojejunostomy**

Patient was then shifted to Critical care unit and was extubated gradually .Patient was started on oral sips on Post operative day(POD) 2 and was started on liquid diet on Post operative day-2 and was mobilised. Ryles tube and foley’s were removed and started on liquid diet on Post Operative Day 3 and was gradually started on full diet.

Drain outputs were monitored and anastomotic site drain was removed on POD 5 and pelvis drain was removed on POD 7. Patient also underwent 2D-echo and was found to be normal and chest medicine reference was done to rule out any lung pathology and was normal. Orthopaedic reference was done in view of Kyphoscoliosis and was advised to follow up in OPD after 3 months post surgery. Patient was tolerating feeds orally and was well mobilised and was discharged on POD 10

Fig 6: Post-operative stage of the patient who underwent the emergency exploratory laparotomy



Discussion

The disease was first reported in 1842 by Carl Von Rokitansky, and in 1927, Wilkie further detailed the pathophysiology and diagnostic findings of the disease, the morbidity and mortality associated with its complications make it a crucial differential to consider when concerned for bowel obstruction, especially in the setting of recent weight loss

Incidence 0.1-0.3 %

Male:Female-2:3

Age-10-39

Pathophysiology-

Duodenal compression is usually due to the loss of the intervening mesenteric fat pad between the aorta and SMA, which in turn, results in a narrower angle between the vessels. The fat pad cushion functions to hold the SMA off the spine and protect it from duodenal compression. A normal aortomesenteric angle is 38 to 65 degrees; however, decreasing the angle less than 25 degrees will decrease the distance to less than 10 mm and cause compression to the third part of the duodenum. Decreases in the aortomesenteric angle can be either congenital or acquired. SMA syndrome is associated with significant weight loss including situations of hypermetabolism (trauma and burns) dietary conditions (anorexia nervosa and malabsorptive diseases) and cachexia causing conditions (AIDS, cancer, paraplegia). Other risk factors include surgical correction of scoliosis, congenitally

short or hypertrophic ligament of Treitz, peritoneal adhesions, duodenal malrotation, Ladd's bands, abdominal aortic aneurysm, lumbar hyperlordosis, and mesenteric root neoplasm.

The management of this condition can vary, and it's essential to approach it in a multi-faceted manner, considering both conservative and surgical options.

Conservative Management

- **Nutritional Support:** Initially, patients may benefit from nutritional support via parenteral nutrition or a modified diet (e.g., small frequent meals that are low in fat) to alleviate symptoms.
- **Weight Gain:** In cases where malnutrition is a factor, an increase in body weight can help widen the angle between the aorta and the SMA, potentially relieving compression.
- **Physical Therapy:** Some patients may benefit from physical therapy to strengthen the abdominal muscles and improve posture, which can sometimes relieve symptoms
- **Pharmacological Management**
 - **Pain Management:** Analgesics may be prescribed to help control abdominal pain as needed.
 - **Prokinetics:** Medications that increase gastrointestinal motility can help alleviate symptoms of nausea and vomiting.
 - **Other Medications:** Depending on the patient's symptoms, antiemetics may also be considered.

Surgical Intervention

- **Decompression Surgery:** If conservative management fails, surgical options may be considered. Techniques include:
 - **Duodenojejunostomy:** Creation of an anastomosis between the duodenum and jejunum to bypass the obstructed region.
 - **SMA Release(Strong procedure):** Surgical division of the ligament of Trietz (the suspensory ligament of the duodenum) may be performed to relieve the pressure on the duodenum.⁷
 - **Angioplasty or SMA reconstruction:** In some cases, vascular intervention may be indicated to relieve the compression.
- **Laparoscopic Approaches:** Minimally invasive techniques may be employed where appropriate, offering quicker recovery and less postoperative pain.

Long-term Follow-up and Management

- **Ongoing Monitoring:** Regular follow-up is necessary to monitor for symptoms of recurrence and nutritional status.
- **Patient Education:** Educating patients about the condition, possible dietary modifications, and the importance of seeking medical help for recurring symptoms.

In our case the patient presented with features of acute obstruction clinically without any history of acute weight loss or any recent surgical procedure and X-ray and CT showed features of acute obstruction which was a rare presentation in a patient with kyphoscoliosis preoperatively and hence the patient was taken up for emergency exploratory laparotomy and a Roux-en Y duodeno-jejunosomy with side to side jejunojejunosomy was done

SMA syndrome is important to consider because the delay in diagnosis can result in significant morbidity and mortality from malnutrition, dehydration, electrolyte abnormalities, gastric pneumatosis and portal venous gas, gastrointestinal haemorrhage and gastric perforation.

Conclusion

The management of superior mesenteric artery syndrome requires a comprehensive approach tailored to the patient's specific needs. While conservative management may suffice in some cases, surgical intervention may be necessary for patients who do not respond to dietary changes and pharmacotherapy. A multidisciplinary team, including surgeons, dietitians, and primary care providers, is essential for effective management and improved patient outcomes.

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