



## Superior Mesenteric Artery Syndrome with Para Duodenal Hernia: A Rare and Underdiagnosed Cause of Intestinal Obstruction

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### Authors' contributions

This work was carried out in collaboration among all authors. Author RKA designed the manuscript, performed the surgery, wrote the protocol and wrote the first draft of the manuscript. Author PA assisted in surgery and wrote the manuscript. Author PM gave the intensive and medical care to the patient. Author NG carried out investigation and reported CT scan of the patient. All authors managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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### ABSTRACT

**Introduction:** Superior Mesenteric Artery (SMA) syndrome also known as chronic duodenal ileus, Wilkie syndrome, arterio-mesenteric duodenal compression syndrome and cast syndrome is a rare cause of proximal bowel obstruction with an incidence that range between 0.013 and 0.3%.

**Case Report:** Our patient aged 35 years female presented in emergency department with complaints of severe abdominal pain, excessive vomiting, inability to pass flatus and motion for 2 days. CT scan of abdomen with contrast showed that stomach and duodenum were grossly distended up to third part of duodenum which was compressed by SMA with reduced aorto-mesenteric angle of 18 degree and distance of 7.3 mm. Nasogastric tube was passed and

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parenteral nutrition was maintained. But patient did not respond to conservative management, hence decision for laparotomy was taken and Duodenojejunosomy was done.

**Discussion:** SMA syndrome is a diagnostic and therapeutic challenge because of its rarity. Various surgical procedures have been considered for surgical management including gastrojejunosomy, Strong procedure (a division of the ligament of Treitz) and duodenojejunosomy.

Most surgeons prefer duodenojejunosomy due to reported success rates of 80% to 100% with reduced postoperative pain, shortened hospital stay.

**Conclusion:** SMA syndrome is an underdiagnosed condition. CT scan is a highly useful modality to diagnose SMA syndrome.

*Keywords: Superior mesenteric artery; intestinal obstruction; duodenojejunosomy.*

## 1. INTRODUCTION

Superior Mesenteric Artery (SMA) syndrome also known as chronic duodenal ileus, Wilkie syndrome, arterio-mesenteric duodenal compression syndrome and cast syndrome is a rare cause of proximal bowel obstruction with an incidence that range between 0.013 and .3% [1,2]. The disease was first reported by Carl Von Rokitansky in 1842 and in 1927, Wilkie further described the disease.

In SMA syndrome, the third portion (transverse portion) of the duodenum is compressed externally between the SMA and abdominal aorta (AA) leading to duodenal stasis and gastro intestinal obstruction. It usually occurs in adolescents and young adults with age range of 10-39 years, more commonly in females over males with a ratio of 3:2. An abnormal low insertion of the SMA or a high insertion of the angle of Treitz that dislocates the duodenum to a cranial position may support this condition. Duodenal compression may occur due to loss of mesenteric fat pad between the AA and SMA. The aorta –SMA angle ranges from 38° to 65° and the distance between the two ranges from 10 to 28 mm. In SMA syndrome there is narrowing of the angle to < 25° and the aorto-mesenteric distance decreases to < 10 mm causing compression of third part of duodenum. Symptoms vary from postprandial nausea, bilious vomiting, abdominal pain and weight loss which may be acute or chronic. Other symptoms include abdominal distension, early satiety and post prandial epigastric pain which worsens in the supine position.

Diagnosis of SMA syndrome is difficult due to vague symptoms of bowel obstruction. Various diagnostic modalities that can be used include plain film x- ray, barium x-ray, endoscopy, computed tomography (CT), Doppler ultrasound and Magnetic resonance imaging. A delay in diagnosis can potentially lead to many

complications such as electrolyte imbalance, catabolic wasting, peritonitis and gastric perforation.

Conservative therapy mainly consists of weight gain achieved orally or parenteral, increasing the mesenteric fat pad and there by aortomesenteric angle. If this approach fails then various surgical procedures like duodenojejunosomy, transabdominal or laparoscopic can be done [3,4].

Here we present a case of SMA syndrome where patient presented with intestinal obstruction and on CT scan this rare entity of SMA syndrome with para duodenal hernia was reported.

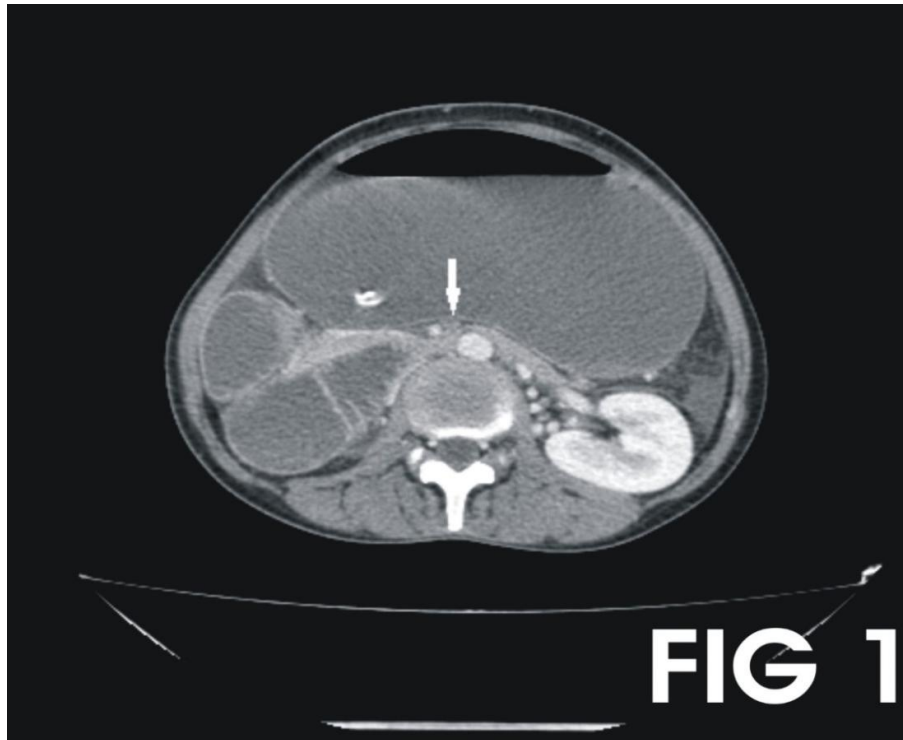
## 2. CASE REPORT

Our patient aged 35 years female presented in emergency department with complaints of severe abdominal pain, excessive vomiting, inability to pass flatus and motion for 2 days. She had recurrent episodes of vomiting with vague abdominal pain, loss of appetite and weight loss of about 10 kilograms in last 3 months. Patient had two full term vaginal deliveries with no significant surgical history. There was no history suggestive of trauma, anxiety or depression, medical problems like abdominal tuberculosis, pelvic inflammatory disease or inflammatory bowel disease. Patient belonged to lower socioeconomic status, illiterate and belonged to remote rural area. Her body weight was 43 kilograms after last child birth which was 5 years back. She had early satiety and pain in abdomen after eating meals for last two years. These were 3-4 episodes when pain got aggravated but she was taken to local practitioners where antacids and analgesics were given and she got relief. Sometimes she had vomiting episodes also but then she stopped eating meals and use to get some relief. She had scanty menses for last one year with associated abdominal pain. She

corelated this abdominal pain with her menstrual problems. On admission her weight was 33 kilograms (loss of about 10 kgs in last three months) and height was 150 cms. with body mass index of 14.66, cachexic with tachycardia and dehydration. She was febrile 101°f, BP-90/60 mm/hg. On per abdomen examination, abdomen was markedly distended and tender. Her relevant blood investigation were haemoglobin-12.5 gm/dl, WBC count- $15.7 \times 10^3$  /cu.mm, random blood sugar 201.0 mg/dl, glycosylated haemoglobin -5.2%, serum sodium -138.0 MEq/ L, serum potassium -3.1 MEq/ L, serum chloride-100.0 MEq/ L. Ultrasonography revealed gross collection with multiple floating echogenic foci, in peritoneal and pelvic cavity suggestive of perforation peritoneal. X-ray abdomen suggested intestinal obstruction. Upper gastrointestinal endoscopy revealed dilated stomach and duodenum. CT scan of abdomen with contrast showed that stomach and duodenum were grossly distended up to third part of duodenum which was compressed by SMA with reduced aorto-mesenteric angle of 18 degree and distance of 7.3 mm. The part of duodenum beyond ligament of Treitz was compressed (not dilated). Abnormal cluster of the jejunal loops was seen in the left anterior

pararenal space posterior to the inferior mesenteric vein suggestive of left para duodenal hernia (Figs. 1, 2, 3, 4). Jejunal loops were collapsed and with normal appearance (Fig. 5). Also mild ascites and left sided pleural effusion was seen.

Nasogastric tube was passed and parenteral nutrition was maintained. But patient did not respond to conservative management, hence decision for laparotomy was taken. Laparotomy was done from midline vertical incision. Gross distention of stomach and duodenum proximal to ligament of Treitz was noted. Distal duodenum (third part of duodenum) was collapsed. Duodenojejunostomy with standard 4 layers approximation and release of ligament of Treitz was done. Para duodenal hernia was seen, jejunal loops taken out from the defect and fascia over superior mesenteric vein forming pouch stitched with periaortic fascia. Postoperative period uneventful. Patient was kept nil orally for 3 days, only sips were given on 4<sup>th</sup> & 5<sup>th</sup> postoperative day, thereafter soft diet and then small frequent meals were advised. Intraperitoneal drain was removed on 7<sup>th</sup> postoperative day. Patient was discharged on 10<sup>th</sup> postoperative day.



**Fig. 1. Decreased Aorto-mesenteric space (white arrow) causing compression of the 3rd part of the duodenum with resultant dilatation of the proximal duodenum and stomach**

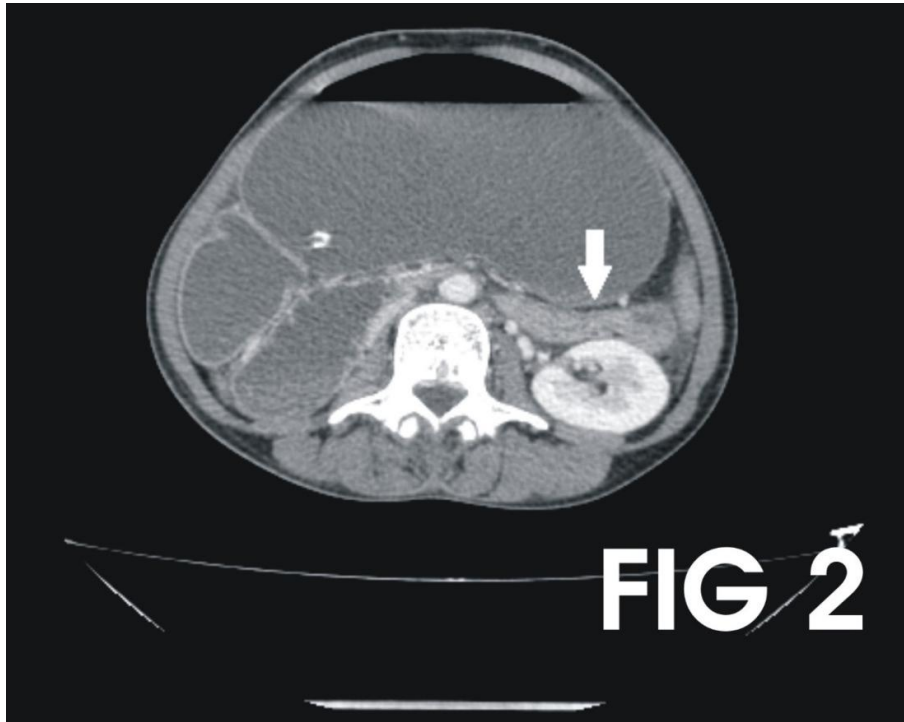


Fig. 2. Abnormal cluster of the small bowel loops in the left anterior para-renal space (white arrow)

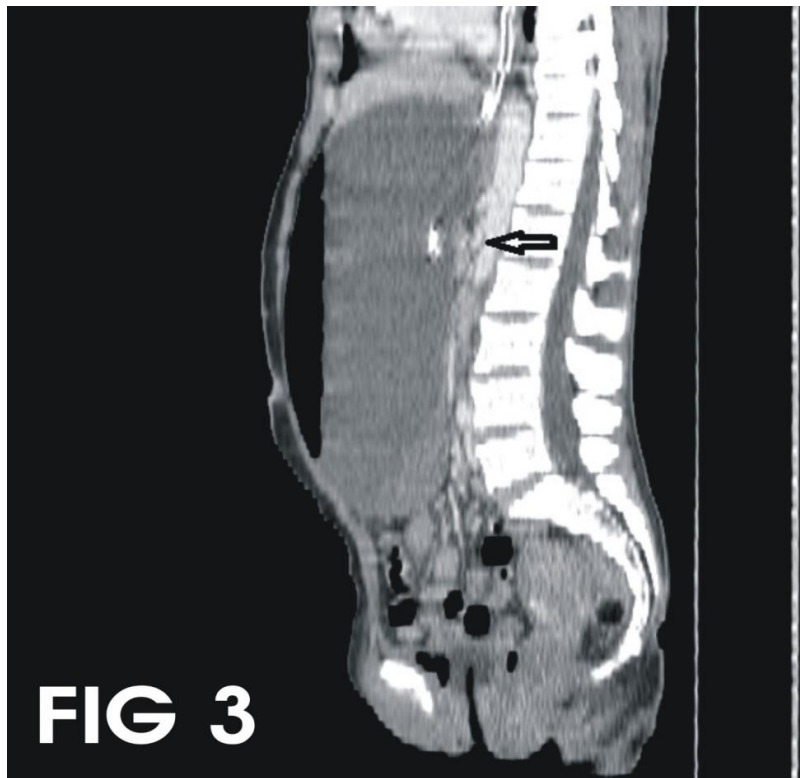


Fig. 3. Decreased Aorto-mesenteric angle (arrow)

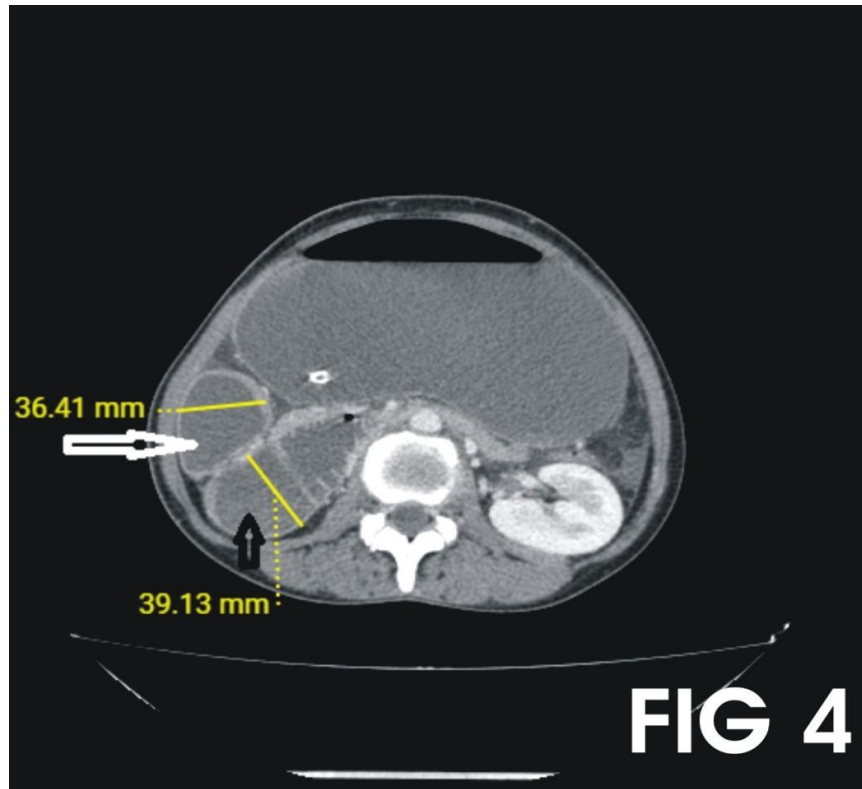


Fig. 4. Dilated 2nd part (white arrow) and 3rd part (black arrow) of duodenum

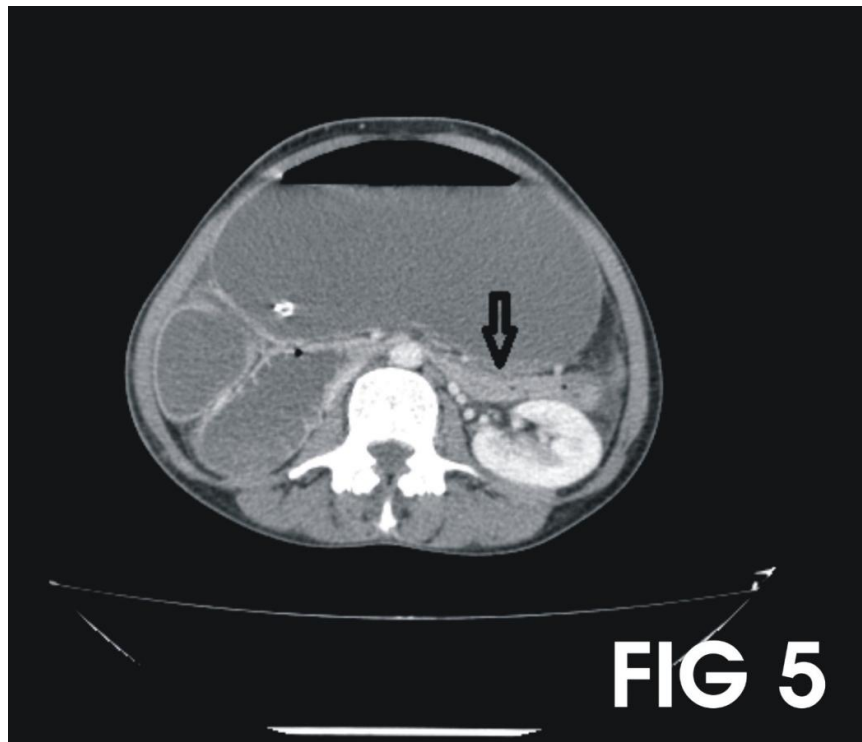


Fig. 5. Collapsed jejunal loops (black arrow) distal to the Aorto-mesenteric interval

### 3. DISCUSSION

SMA syndrome is a diagnostic and therapeutic challenge because of its rarity [5,6,7,8]. Our patient had severe acute weight loss over period of 3 months. Such significant weight loss can occur in conditions like anorexia nervosa, malabsorption, hypercatabolic states (burns, major surgery, malignancy), severe congestive heart failure causing cachexia. Other conditions which may precipitate this condition are corrective spinal trauma and after abdominal surgery such as total proctocolectomy and ileal J-pouch anal anastomosis due to tension and caudal pull of the small bowel mesentery. Despite of weight loss and recurrent vomiting our patient didn't take any treatment and was brought only when the abdominal pain become intolerable.

The diagnosis of SMA syndrome is challenging. Barium radiography demonstrates dilatation of the first and second part of the duodenum with or without gastric dilatation, antiperistaltic flow of barium proximal to the obstruction and a delay of 4-6 hours in gastro duodenojejunal transit time, with relief of obstruction when the patient is placed in the prone, knee, chest or left lateral position. In our case CT scan clearly mentioned the reduced aortomesenteric angle and distance contributing significantly to this rare diagnosis. CT scan also suggested para duodenal hernia which if not corrected intraoperatively could have further led to obstruction as jejunal loops have entered the sac.

Traditionally treatment starts with conservative measures like decompression of the stomach and duodenum with a nasogastric tube, correction of nutritional and electrolyte deficiencies, through TPN, or preferably, if possible enteral feeding with a naso jejunal tube passed beyond the point of compression, which fulfills nutritional requirements while avoiding the complications of TPN. When tolerated, oral feeding may be resumed [9,10,11]. Various posturing manoeuvre like prone, knee to the chest and left lateral decubitus positions after meals may be helpful. Prokinetic agents (such as mosapride) proton pump inhibitors or physiotherapy may help to improve the gastrointestinal motor function. Sinagra et al (9) did not require any surgical intervention in their series.

Various surgical procedures have been considered for surgical management including

gastrojejunostomy, Strong procedure (a division of the ligament of Treitz) and duodenojejunostomy. Traditional open bypass surgery was the standard care until 1998 when the first successful Laparoscopic duodenojejunostomy was performed and this has now replaced open bypass [12,13,14].

Most surgeons prefer duodenojejunostomy due to reported success rates of 80% to 100% with reduced postoperative pain, shortened hospital stay. Gastrojejunostomy, a previously reported surgical treatment, has been abandoned because of increased postoperative complications like blind loop syndrome and recurrence of symptoms due to non-decompression of the duodenum. After corrective surgery our patient has completed 4 weeks with total relief of abdominal pain, increased appetite and weight gain of 2 kilograms. She has been advised small frequent meals and prokinetic drugs.

### 4. CONCLUSION

SMA syndrome is an underdiagnosed condition. Persistent vomiting with history of weight loss and clinical findings suggestive of subacute, chronic or acute intestinal obstruction should raise the suspicion of this diagnosis. Contrast enhanced CT scan is a highly useful modality to diagnose SMA syndrome.

### CONSENT AND ETHICAL APPROVAL

As per university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

### COMPETING INTERESTS

Authors have declared that no competing interests exist.

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