**Title page**

**An uncommon cause of sub-acute intestinal obstruction in young adult: Wilkie's syndrome**

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**Key Clinical Message**

Superior mesenteric artery (SMA) syndrome, also known as Wilkie's syndrome, is a rare disease presenting as an acute abdomen with clinical features similar to intestinal obstruction. This is a case of an uncommon definitive pathology due to decreased acuity of aortomesenteric angle diagnosed as Wilkie's Syndrome.

**Keywords:** Superior mesenteric artery syndrome, SMA, Wilkie Syndrome, Acute abdomen

1. **Introduction**

Superior mesenteric artery (SMA) syndrome, also known as Wilkie syndrome, is a rare disease presenting as an acute abdomen with clinical features similar to intestinal obstruction. Thin lean body habitus and reduced weight leading to loss of fat pad between the SMA and aorta, resulting in decreased acuity of aortomesenteric angle, is the pathophysiology in this syndrome. Imaging modalities such as barium meal and CT scan facilitate its diagnosis. The mainstay of management is conservative treatment with decompression using a nasogastric tube, bypassing duodenum with a nasojejunal tube, correction of nutritional deficiencies and electrolyte abnormalities, and total parenteral nutrition or oral nutrition as tolerated. Surgery remains the treatment of choice in refractory cases. This case report is in line with CARE guidelines.(1)

1. **Case report**

A 31 year old serving soldier with no known comorbidities presented to emergency with complaint of pain abdomen and progressive vomiting for three months. Pain abdomen was on and off over the left lumbar and periumbilical region, which was pricking in nature, increasing in severity, with no radiation. It was associated with vomiting which was immediately after food consumption, containing undigested food particles, non- foul smelling, non-bile stained and on-blood stained. It peculiarly aggravated on eating and relieved to a slighter extent on fasting. He gave history of six to eight episodes of vomiting per day. He also had anorexia and hadn’t passed stool for 1 week but had passed flatus. During these period he gave history of significant weight loss of more than 20 kg in the span of last three months. There was no however no history of fever, loose stool, burning micturition. Bladder function was normal. He had no history of any surgical intervention in the past. Vomiting, in particular, worsened for past three months. He had multiple episodes of forceful, projectile, watery, non-mucoid, and bilious vomiting with associated nausea and shortness of breath.

On examination, his general condition was ill looking, frail and wasted. He had scaphoid abdomen which was soft with no tenderness, no organomegaly and bowel sounds were present. Other systemic examinations were unremarkable.

* 1. **Diagnostic assessments**

Blood investigations showed complete blood count and random blood sugar levels to be within normal limits. However, blood amylase and lipase were 102U/L and 122.3u/l respectively. Blood urea and creatinine levels were raised with values 87.7mg/dl and 1.42mg/dl respectively. Renal Function tests showed all within normal limits with features of hyponatremia

Ultra Sonography showed dilated loops of 5.4 cm with to and fro motion of intraluminal contents in right upper and lower quadrant suggestive of sub-acute bowel obstruction.

CECT (A+P) showed grossly distended stomach with abrupt tapering at the junction of 2nd and 3rd part of duodenum with mass effects The narrowed aorto-mesenteric angle (AO) was 19.5 degrees (**Figure 1)** The decreased aorto-mesenteric distance was measuring 4.41mm (**Figure 2**), all findings concomitant with Wilkie’s syndrome.

* 1. **Treatment**

The patient was initially managed in Emergency Room, he was kept Nil Per Oral (NPO) and fluid along with hyponatremia was managed by Normal Saline. Vomiting was managed by Ondansetron, along with Pantoprazole. Then the patient was shifted to the ward for further investigation until the confirmation of diagnosis.

After the reports of Focused USG and CECT, the diagnosis was made Superior mesenteric artery (SMA) syndrome for which exploratory laparotomy with retrocolic duodenojejunostomy was done under general anesthesia.,

Intraoperative findings showed chronic midgut volvulus with 180 degrees counter rotation of SMA and tortuous vessels in small bowel mesentery. He was then kept under observation in the post-operative ward and kept NPO for 8 hours following the surgery and then shifted to the ward after 12 hours. He started on a liquid diet after 6 hours and tolerated it well.

* 1. **Follow up.**

He was discharged with Antibiotics (Cefixime and Metronidazole), Paracetamol, and Pantoprazole after 7 days only after the pain resolved and he made full recovery. The patient was advised for alternate day dressing with suture out on the 10th postoperative day. No post operative complications were countered during his follow-ups.

1. **Discussion**

Superior mesenteric artery (SMA) syndrome also known as Wilkie syndrome, chronic duodenal ileus, arterio-mesenteric duodenal compression syndrome and case syndrome is a rare disease defined as compression of the third portion of the duodenum between abdominal aorta and the superior mesenteric artery. Its incidence is estimated to be 0.1% to 0.3% and preferentially occurs in adolescents and young adults of age 10 to 39 years. It remains as a diagnosis of exclusion and presents with vague symptoms such as post prandial recurrent abdominal pain to more severe abdominal pain, nausea, vomiting, and electrolyte imbalances.(2) Similar presentation was found in our case with acute abdominal pain and post prandial vomiting. Loss of fat pad between the SMA and aorta due to thin lean body habitus and reduced weight is considered to be the main pathophysiology in SMA syndrome.(3) The presented case also has a history of weight loss of 20kg in 3 months and has a thin lean body habitus.

SMA syndrome has been associated various factors which decrease the acuity of the angle between the aorta and SMA such as malignancy, trauma, substance abuse, burns, spinal or bariatric surgery and anorexia nervosa.(4,5)

Diagnostic workup requires imaging modalities such as abdominal radiographs, barium studies and CT findings. Plain abdominal radiographs reveal findings suggestive of small bowel obstruction whereas barium studies show dilation of the first and second part of the duodenum and a relatively collapsed small bowel distal to point where the SMA crosses the duodenum. In normal CT findings, the aorto-mesenteric angle and aorto-mesenteric distance measure 28-650 and 10-34mm; in SMA syndrome, both parameters are reduced with values to 60 to 220 and 2 to 8mm.(3) In this case, the aorto-mesenteric angle was 19.50, and the aorto-mesenteric distance was 4.41mm.

Conservative management with decompression of stomach using nasogastric tube, bypassing duodenum by nasojejunal tube, correction of nutritional and electrolyte deficiencies by feeding tube or total parenteral nutrition remains the mainstay of management. The goal is to improve the nutritional status which in turn builds up the fat cushion between SMA and aorta. Prone position, left lateral decubitus, or knee-chest position has been found to relieve symptoms by removing tension from the mesentery and the SMA and opening the space between SMA and aorta.(6) However, surgical management with derotation of duodenum has been proven to be effective in cases refractory to conservative approach.(3)

SMA syndrome is thus a rare disease resulting in life threatening outcomes. A high index of suspicion is a must in cases of acute abdomen especially when it is associated with post prandial pain and weight loss.

1. **Conclusion**

The case of Wilkie Syndrome is rare and their clinical presentation can sometimes do not be so apparent. Enhanced CT remains the gold standard diagnostic modality for Wilkie syndrome. However, when complicated, they can result in life threatening outcomes, for which exploratory laparotomy with retrocolic duodenojejunostomy is preferred.

**CONFLICT OF INTEREST STATEMENT**

No conflict of interest.

**FUNDING INFORMATION**

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**DATA AVAILABILITY STATEMENT**

All the findings are present within the manuscript.

**CONSENT**

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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