

Wilkie's syndrome: Laparoscopic management – A case report with review of the literature

Shreya Shetty¹, Abhijit Joshi²

From ¹Clinical Associate, ²Consultant, Department of General and Laparoscopic Surgery, Dr. L H Hiranandani Hospital, Mumbai, Maharashtra, India

ABSTRACT

Superior mesenteric artery (SMA) syndrome also known as Wilkie's Syndrome is a rare condition characterized by symptoms of small bowel obstruction. It is caused when the angle between the SMA and aorta, also known as the aorto-mesenteric angle, reduces to $<25^\circ$ which, in turn, causes the compression of the third part of the duodenum which traverses between the SMA and the aorta. We, herein, present the case of a 31-year-old female who was diagnosed to have Wilkie's syndrome. Failing a trial of conservative management, she was ultimately definitively managed by surgery.

Key words: Aorta, Aorto-mesenteric angle, Duodenum, Superior mesenteric artery

Superior mesenteric artery (SMA) syndrome was first described in 1842 by the Austrian physician Professor Carl von Rokitansky. In 1927, Sir D.P.D Wilkie of the University of Edinburgh published the first series of 75 patients; hence, SMA syndrome is also called Wilkie's syndrome [1]. Only about 500 cases have been reported, worldwide, so far [2]. SMA syndrome has a prevalence of approximately 0.3% [3,4]. It occurs more commonly in females than males with a ratio of 3:2. No ethnic predisposition has been reported [5]. A higher peak incidence is noted between 10 and 18 years of age [3,4]. The SMA syndrome can be managed conservatively in most cases. However, some patients do require surgical intervention.

We present the case of a 31-year-old female who was diagnosed to have Wilkie's syndrome. The rationale for reporting this case is to make our own small contribution to the existing limited literature on this rare condition.


CASE REPORT

A 31-year-old female was referred to us with complaints of vomiting 30–40 min after intake of food. The vomiting was spontaneous, non-projectile, and consisted of undigested food particles along with bile. The patient had 5–6 such episodes daily for the past month. She also experienced sudden-onset post-prandial epigastric pain about 15–20 min after consuming food. The pain was continuous and colicky in nature and lasted until

the food was vomited. She had mild relief when she assumed the knee-elbow position. She had consulted a general practitioner and was prescribed anti-emetics and antacids based on an empiric diagnosis of acid peptic disease. However, they did not alleviate her symptoms; and hence, she was referred to us for further evaluation and management. She, further, revealed that she had an unintentional weight loss of 12 kg over 3 months.

On examination, her vitals were normal. Her body mass index (BMI) was 15. A per abdomen examination revealed a tympanic distended upper abdomen with tenderness present in the epigastric region. She did not have any organomegaly or free fluid and her bowel sounds were sluggish.

Her routine blood investigations and thyroid function tests were within normal limits. Her chest X-ray was normal. X-ray erect abdomen revealed a dilated stomach with a prominent air-fluid level in the upper abdomen. An ultrasonography (USG) scan of the abdomen showed dilated stomach with gastric contents. USG Doppler revealed an aorto-mesenteric (A-M) angle of 12 degrees. A contrast-enhanced computed tomography scan of the abdomen was, then, performed and it revealed a grossly dilated stomach and duodenum with abrupt narrowing in the third part of the duodenum. An oesophago-gastro-duodenoscopy also showed narrowing in the third part of the duodenum due to extrinsic compression. Barium meal stomach-duodenum was done which revealed dilated stomach, dilated first, second, and proximal third part of the duodenum, with to and fro peristalsis. Extrinsic pressure effect was noted at the junction of the third and fourth parts of the duodenum. A computed tomography aortography was then done which revealed an A-M angle of 11

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Correspondence to: Dr. Shreya Shetty, A/501 Dipti Greens, Opp. Bombay Cambridge School, J. B. Nagar, Andheri East, Mumbai - 400099, Maharashtra, India. E-mail: shreya_8592@yahoo.com

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degrees (Fig. 1a, c, d). Thus, a diagnosis of SMA syndrome was reached.

Initially, the patient was put on a trial of conservative management for a period of 4 weeks. A nasogastric tube was inserted to decompress the stomach and intravenous hydration was initiated. She was told to follow a diet comprising of small portions of frequent semi-solid meals to improve her nutritional status and help in weight gain. She was recommended to assume the knee-elbow position after consumption of these meals. However, these measures did not relieve her symptoms, significantly.

Failing the trial of conservative management, she was then planned for surgery. A laparoscopic duodenojejunostomy (D-J) was performed. The obstruction was bypassed by doing a side-to-side anastomosis of the duodenum (proximal to the D3 narrowing) and a loop of the jejunum. A harmonic scalpel was used as the energy source and the D-J was performed using an Endo-GIA stapler loaded with a blue cartridge and intracorporeal suturing with 2-0 Vicryl (Figs. 2 and 3).

Her post-operative recovery was uneventful. She was started with a liquid diet on the post-operative day (POD) 3 and a soft diet on POD 5 which were tolerated well. She passed stools on POD 6 and was subsequently discharged from the hospital. On her POD 10 outpatient department follow-up visit, all her wounds had healed well. On her 3 monthly follow-up visit, she had gained 6 kg and was asymptomatic.

DISCUSSION

At the level of the L1 vertebral body, the SMA emerges from the anterior portion of the abdominal aorta and extends at an acute angle

into the mesentery. The normal angle of this entry is around 38–56° (Fig. 1b) and the normal A-M distance is 10–20 mm [2]. The third part of the duodenum (D3) traverses through the space between the aorta and the SMA. When there is a reduction of this angle to <25°, it causes compression of the third segment of the duodenum which, then, causes duodenal obstruction [2]. The narrowed A-M angle can be due to anomalous anatomy such as high insertion of the duodenum at the ligament of Treitz, low origin of the SMA, or due to loss of retroperitoneal fat causing a reduction in the angle [6]. Loss of retroperitoneal fat due to weight loss in the pathogenesis of SMA syndrome in patients with malabsorption, anorexia nervosa, hyperthyroidism, during chemotherapy, profound gastroenteritis, extensive burns, major surgical procedures especially orthopedic and neurosurgical operations, supine immobilization, operative correction of scoliosis [7,8], severe traumatic brain injury, spine injuries and placement of full-body cast, and hip-spica cast application (hence, SMA syndrome is also known as Cast

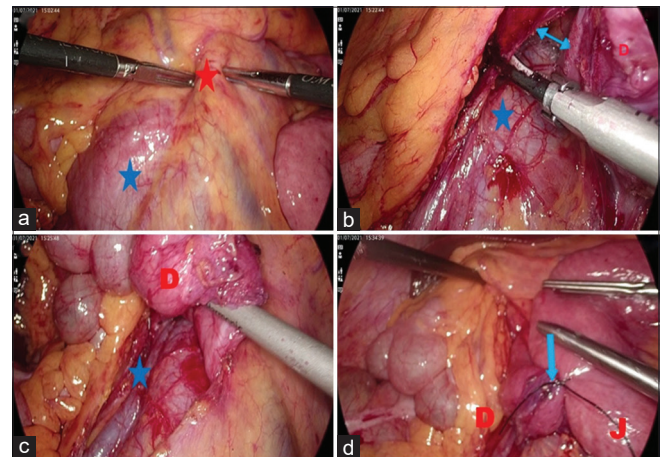


Figure 2: Operative pics (a) superior mesenteric artery (red asterisk) seen running over and compressing D3 (blue asterisk) and (b) visualization of the inferior vena cava (blue asterisk), while Kocherization of duodenum (red D) being done, (c) Kocherization complete, and (d) fixation stitch taken between duodenum and jejunum

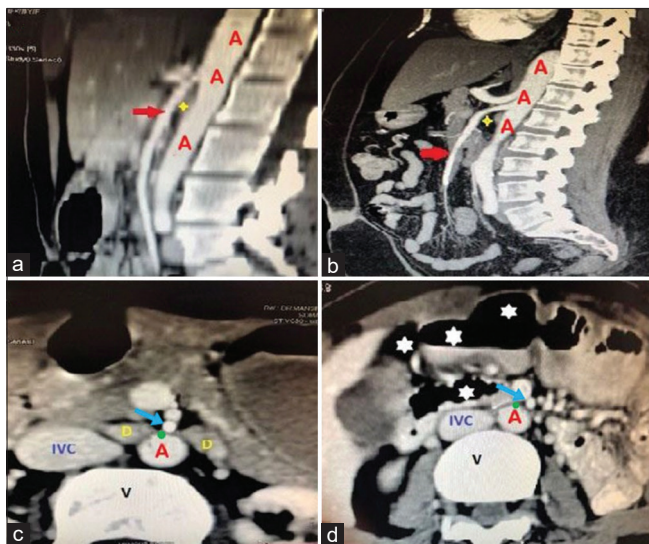


Figure 1: Radiology evaluation (a) abdominal computed tomography (CT) angiogram of the patient showing the aorta (the red As) and superior mesenteric artery (red arrow) with the narrowed A-M angle (yellow asterisk), (b) CT angio of a normal person for comparison, showing normal A-M angle, (c) CT abdomen axial cut of the pt. showing the vertebral body (V), aorta (A), inferior vena cava, duodenum (yellow D), the compressed part of D3 (green asterisk) and SMA (blue arrow), and (d) another axial cut showing dilated duodenum (white asterisks) proximal to level of obstruction (green asterisk)

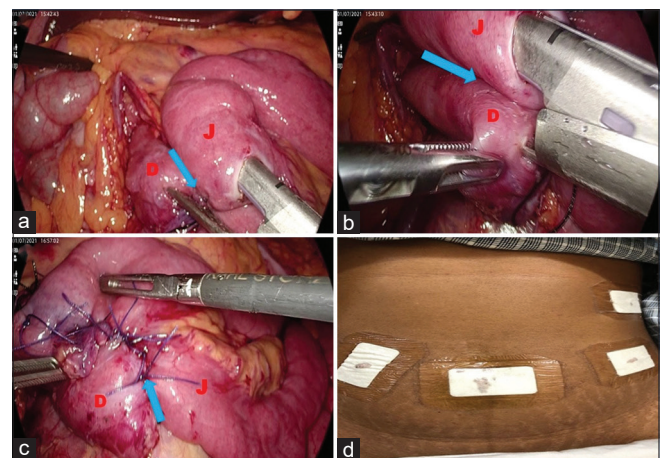


Figure 3: Operative pics (a) EndoGIA™ being inserted (blue arrow) into the duodenum and jejunum after making a duodenotomy and jejunotomy, respectively, (b) EndoGIA inserted completely (blue arrow) and ready to close and fire, (c) Endresult – bypassing stapled cum sutured D-J (blue arrow), and (d) trocar sites

syndrome) [3]. Weight loss usually occurs before the onset of symptoms and also contributes to the syndrome, as happened in our case. A-M duodenal compression, A-M artery compression, and duodenal vascular compression are some of the other terms used to describe SMA syndrome. Rare instances of SMA syndrome concurrently existing with Nutcracker syndrome have been reported. Nutcracker syndrome is the compression of the left renal vein between the SMA and the aorta causing symptoms related to renal congestion [9].

SMA syndrome may also occur in healthy children and is accompanied by growth spurts during puberty. In the early puberty, especially boys have a significant increase in lean body mass and a concomitant loss of adipose tissues which may lead to a narrowing of the A-M angle. Acute consumption of food and water can cause a transient obstruction in the already-narrowed space, which can manifest SMA syndrome. Thus, weight loss is not necessarily a criterion for the development of SMA syndrome in the pediatric population, at puberty [10]. SMA syndrome has also been reported in the adolescent population after posterior spinal fusion from T2 to L3 for idiopathic adolescent scoliosis. These patients were treated conservatively and responded well to the treatment [8]. The surgical correction of scoliosis causes realignment and longitudinal traction of the aorta increasing the height of the patient and subsequently causing narrowing of the angle between the aorta and SMA causing obstruction of the third portion of the duodenum, especially in patients with a BMI <18 or a weight percentile for the height of 5% that suggests low or no periphery duodenal visceral fat which is a well-known risk factor [11-13].

Usually, the coeliac trunk and the SMA have distinct origins from the abdominal aorta. The celiacomesenteric trunk (CMT) is the least frequently reported anatomic variation of all abdominal vascular anomalies. CMT denotes a common trunk of origin of the coeliac and superior mesenteric arteries. Only one case of a 59-year-old man presenting with duodenal obstruction due to SMA syndrome with CMT is reported in the literature [14].

Diagnosis is established by a barium follow-through series or a hypotonic duodenography (SMA syndrome shows an abrupt or near-total cessation of the flow of barium from the duodenum to jejunum) [15]. USG Doppler or MR angiography can confirm the A-M angle. CT angiography is currently favored in the literature for diagnosis as it can not only show the narrowed A-M angle and A-M distance, but also the extent of duodenal obstruction [16].

Usually, a trial of conservative management is given initially which includes nasogastric decompression, correction of dehydration, prevention/correction of electrolyte imbalance, and prokinetic drugs (cisapride, and metoclopramide). In severe cases, nasojejunal feeding or total parenteral nutrition is effective to prevent further weight loss. Psychiatric assessment is necessary for those with eating disorders. Dietary modifications are an important part of conservative management. A frequent small portions of meals are recommended instead of heavy meals. Post meals, patients are advised to assume the knee-elbow position. Patients with acute SMA syndrome, particularly pediatric patients often respond well to this conservative treatment.

Endoscopic ultrasound-guided deployment of lumen apposing gastrojejunostomy stent has also been reported, where conservative treatment had failed and the patient was not deemed fit to undergo a surgical procedure due to old age and comorbidities [17]. Patients with chronic SMA syndrome require surgical intervention. Surgery is contemplated when the medical management fails or when there is a long-standing disease with progressive weight loss and duodenal dilatation with stasis or when there is a complicated peptic ulcer disease secondary to biliary stasis and reflux. The surgical options for SMA syndrome are Strong's Procedure, Gastrojejunostomy, and Duodenojejunostomy. Strong's procedure mobilizes the duodenum by dividing the ligament of Treitz. Once the ligament of Treitz is divided, it allows the 3rd part of the duodenum to drop lower from the narrow aorto-mesentric angle thereby relieving the obstruction. The advantage of this procedure is that there is no bowel anastomosis, and hence, there is early post-operative recovery. However, the failure rate of Strong's procedure is 25%. The other 2 procedures are surgical bypasses. At present, laparoscopic duodenojejunostomy is the surgery of choice. It was first performed by Gersin and Heniford in 1998 and has a long-term success rate above 90%. Alternatively, gastrojejunostomy can also be done when gastric distension is present which has caused gastroparesis and delayed emptying of the stomach. Vascular reimplantation of the SMA is used as a last option due to the morbidity associated with the procedure.

CONCLUSION

SMA syndrome is an extremely rare clinical entity and very few cases have been reported to date. There are various causes of SMA syndrome and it has been noted that it occurs after significant weight loss in a large number of cases. It is usually managed conservatively using drugs, changes in dietary habits, and sleeping postures. Surgical management is undertaken if the initial medical management fails. As seen from this report, laparoscopic duodenojejunostomy gives excellent results and is the surgery of choice.

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