International Surgery Journal
Kushwaha SS et al. Int Surg J. 2019 Jul;6(7):2591-2594
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20192999

Case Report

Facts and fantasies about superior mesenteric artery syndrome: an unusual cause of intestinal obstruction

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Received: 30 April 2019
Accepted: 14 June 2019

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ABSTRACT

Superior mesenteric artery (SMA) syndrome (also known as Wilkie's syndrome) is an unusual cause of proximal intestinal obstruction, attributable to vascular compression of the third part of duodenum between the superior mesenteric artery and the abdominal aorta due to acute angulation of SMA. It is a life threatening disease as it poses a diagnostic dilemma and often diagnosed by exclusion of other causes. It is an acquired disorder and is commonly due to loss of fatty tissue as a result of a variety of debilitating conditions. We report a case of SMA syndrome in a 23 year young asthenic female patient, with a long history of recurrent abdominal pain, epigastric fullness, voluminous vomiting, and weight loss. Symptoms persisted for 1 year and the patient underwent extensive investigations, but to no avail. Thereafter she developed proximal intestinal obstruction, which unravelled her diagnosis. Abdominal examination revealed epigastric fullness, tenderness and hyper peristaltic bowel sounds. We performed small bowel enteroclysis, upper gastrointestinal series, abdominal computer-tomography (CT) and ultrasonography to establish the diagnosis. Conservative treatment was tried for one month but failed. There was no relief of symptoms in the left lateral decubitus or prone position. Finally, the patient successfully underwent Roux-en-Y duodenojejunal anastomosis with a postoperative favourable outcome. This case emphasizes the challenges in the diagnosis of SMA syndrome and the need for increased awareness of this entity. This will improve early recognition in order to reduce irrelevant tests and unnecessary treatments.

Keywords: Acute angulation, Duodenojejunal anastomosis, SMA syndrome

INTRODUCTION

Superior mesenteric artery (SMA) syndrome is an uncommon condition caused by intermittent functional obstruction of the third part of the duodenum between the superior mesenteric artery and the abdominal aorta. Clinical symptoms include epigastric pain, a sensation of fullness, nausea and bilious vomiting.[1]The syndrome mostly develops after a rapid and dramatic weight loss. Weight loss continues after developing symptoms because patients often regurgitate their food or become afraid to eat, which in turn exacerbate the condition. Symptoms can be relieved by postural changes in the prone or knee–chest position, suggesting vascular compression. However, since relief with postural change occurs inconsistently, its absence does not rule out vascular compressive syndromes.

Many causes for SMAS have been suggested. These include a high insertion of the duodenum at the ligament of Treitz, a congenitally low origin of the superior mesenteric artery and compression of the duodenum caused by peritoneal adhesions, which are a result of duodenal malrotation.[2,3] However, the most common cause is thought to be narrowing of the aortomesenteric angle and the distance between the aorta and the superior mesenteric artery at the level of the duodenum, with consequent extrinsic compression of the duodenum.

Symptoms of SMA Syndrome are non-specific, and the syndrome can be difficult to differentiate from peptic ulcer disease, cholelithiasis, and pancreatitis. As the diagnosis is generally problematic so its imperative to convincingly exclude other causes which can cause obstruction of the duodenum. This syndrome can occur as an acute illness but patient generally has a longer history. The first proposed surgery for SMA Syndrome was described as duodenojejunostomy.

CASE REPORT

A 23 year young asthenic female patient presented to our surgical emergency room with 3 days history of upper central abdominal pain and fullness associated with repeated attacks of vomiting. Pain was colicky in nature, mainly localized in epigastric region, which gets precipitated by eating and relieved after bouts of vomiting (undigested food). She had previous similar attacks of recurrent abdominal pain, epigastric fullness, voluminous vomiting and weight loss since 1 year for which she was taking treatment from a private practitioner. Patient was managed conservatively for intestinal obstruction twice in the past. No significant previous surgical or medical problem. Patient reported that she lost weight over the months preceding her symptoms. Examination revealed very thin dehydrated patient with normal vitals. She looked anxious and her physical examination was unremarkable except for a diffuse tenderness with fullness over epigastric area.

Laboratory evaluation at admission was normal except for elevated bicarbonate (31 mmol/l) consistent with emesis and contraction alkalosis. White blood cell count was 5,000/mm^3, hemoglobin 12 g/dl, sodium level 140 mEq/l, potassium 4 mEq/l, blood urea level 18 mg/dl and creatinine 0.75 mg/dl. The liver profile was normal with AST 26 IU/l, ALT 21 IU/l, total bilirubin 0.4 mg/dl and alkaline phosphatase 26 IU/l.

X-ray abdomen with both erect and supine view showed virtually gasless abdomen with an air bubble in the region of stomach. Ultrasound abdomen revealed severe gastric dilatation reaching down to below the umbilicus raising the suspicion of gastric outlet obstruction (GOO). Patient was admitted and started on IV hydration and gastric decompression was done through nasogastric tube. Upper gastrointestinal series showed dilated stomach with dilated second part of the duodenum and cut off at the third part of duodenum with no intrinsic mucosal abnormalities. Thereafter patient underwent contrast enhanced CT scan abdomen that revealed severely dilated stomach and significant dilatation of the duodenum up to the level of the distal third part, abrupt narrowing (transition zone) seen just anterior to the abdominal aorta and posterior to the superior mesenteric artery as well as significant reduction of the aortomesenteric angle (measuring 18 degrees) and aorto-mesenteric distance measuring about 5 mm.

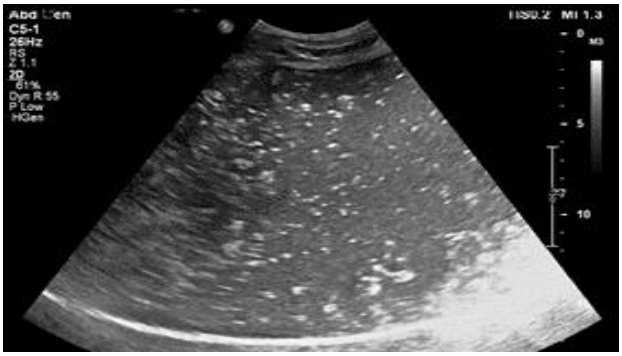

Figure 1: Abdominal USG showing grossly dilated stomach.

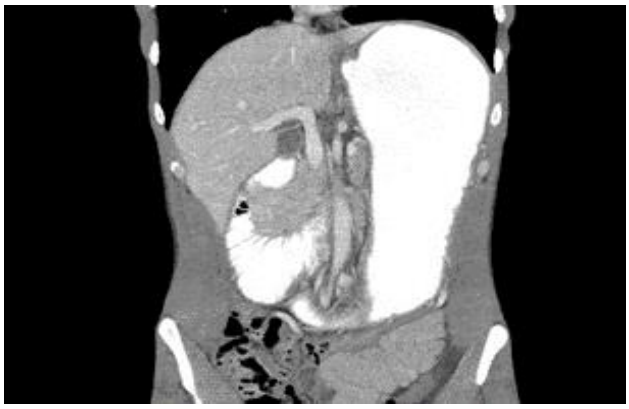

Figure 2: Abdominal CECT showing grossly dilated stomach.

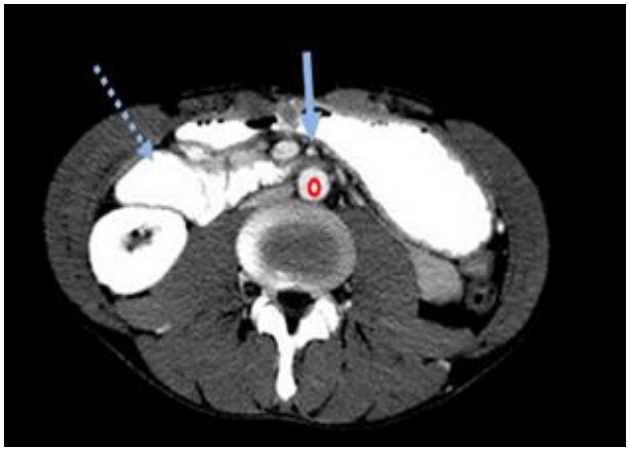

Figure 3: Abdominal CECT showing superior mesenteric artery (Arrow), abdominal Aorta (red circle), dilated duodenum (Dash arrow).

Conservative management was planned and tried for 1 month. The patient was told to take frequent small meals of nutritious liquid, advised to lie on left side/prone following meals. Metaclopramide was also advised but no relief of symptoms found, so surgery was planned. Exploratory laparotomy through a midline incision was done. Intra-operative findings confirmed the extrinsic obstruction of third part of duodenum with distension of

2nd part. Small insignificant mesenteric lymph nodes were also present. Mesenteric lymph nodal biopsy was taken. A Roux-en-Y duodenojejunostomy was done, proximal end of the 40 cm long Roux limb was anastomosed end-to-side to the second part of the duodenum. In post-operative follow up, patient was symptom free and started taking normal diet and added some weight.

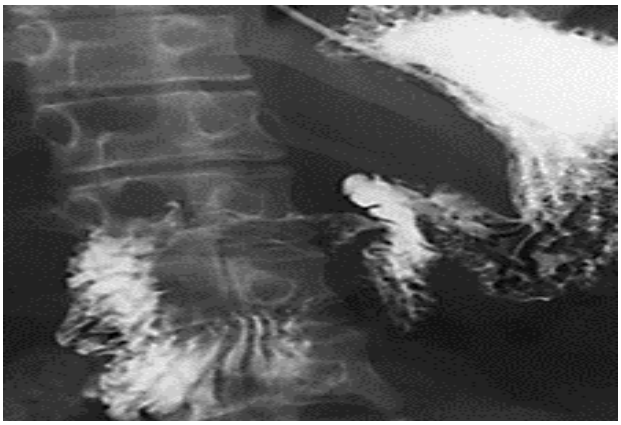

Figure 4: Barium study showing dilated stomach and proximal duodenum.

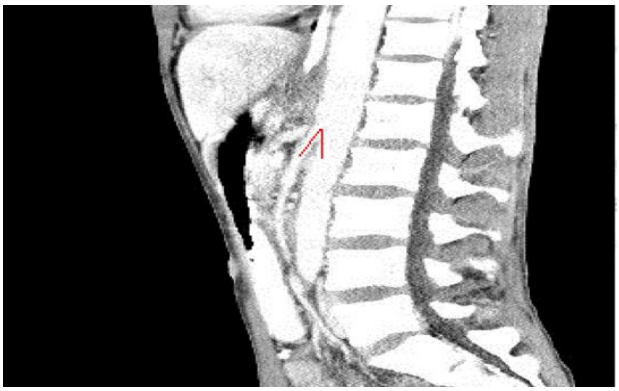

Figure 5: Abdominal CECT showing decreased aortomesenteric angle.

DISCUSSION

Since the entity was first described at autopsy by von Rokitansky in 1861, about 400 cases have been reported in the medical literature. The condition is often referred to as Wilkie's syndrome or cast syndrome. In 1927, Wilkie published the largest and most complete study of this disease, based on 75 cases.[4] Skepticism about the existence of the condition continued until the 1960s, when new radiologic techniques provided evidence to support the existence of SMA Syndrome.

Currently, the reported incidence is on the rise, possibly due to increased physician awareness. SMA syndrome is a rare pathology with an incidence that ranges between 0.013 and 0.3% reported in literature; however the true incidence remains unknown.[5]

The defining feature of this entity is upper gastrointestinal obstruction caused by compression of the third part of the duodenum between the SMA anteriorly and the aorta posteriorly. In normal anatomy the aortomesenteric angle and aortomesenteric distance is 25 degree - 60 degree and 10–28 mm, respectively. The main anatomic feature of SMA syndrome is the narrowing of the aorta-SMA angle to <25°, and the aortomesenteric distance decreases to <10 mm.[6,7]

Etiological factors can be either a congenital or an acquired anatomic abnormality or, more commonly, a debilitating condition causing severe weight loss. Congenital etiologies include abnormally low insertion of the SMA or high insertion of the angle of Treitz dislocating the duodenum to a cranial position. Acquired anatomic abnormalities can occur following corrective spinal surgery such as scoliosis surgery by a relative lengthening of the spine, 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. Severe weight loss, leading to a depletion of the fatty cushion around the SMA is a major cause of SMA syndrome. The disorders that predispose to SMA Syndrome have been divided into five categories: severe wasting diseases such as burns, cancer and endocrine diseases; severe injuries such as head trauma; spinal trauma, deformity, including the application of a body cast; dietary disorders such as anorexia nervosa or malabsorptive syndromes; and the postoperative state.

Females aged between 10 to 40 years are more commonly affected. Patient might presents with acute symptoms of intestinal obstruction as in our case or more commonly with chronic symptoms as recurrent abdominal pain with cramps, early satiety and postprandial fullness. Sometimes pain will be aggravated with lying supine and get relieved in knee chest position; a maneuver that increases the aortomesenteric angle with subsequent relief of bowel obstruction.

SMA syndrome diagnosis is challenging and often delayed due to its insidious onset. The diagnosis should be suspected based on clinical presentation and supported by radiological tests. Barium studies might show duodenal dilatation and sometimes gastric dilatation with slow gastroduodenojejunal transit. Contrast-enhanced CT or magnetic resonance angiography enables visualization of the vascular compression of the duodenum and precise measurement of the aortomesenteric angle and distance.

Treatment is usually conservative which includes gastric decompression, fluid electrolytes imbalance correction and nutritional support either through total parenteral nutrition or post pyloric tube feeding (nasojejunal tube). Conservative treatment focuses on nutritional support aimed at restoration of retroperitoneal fat and weight gain. Posturing maneuvers during meals and motility agents may be helpful in some patients. If conservative

treatment fails to relieve obstruction surgical procedures to bypass the obstruction should be considered. Duodenojejunostomy is the procedure of choice with success rate reach up to 90%. Another simpler operation called Strong's procedure which involves division of the ligament of Treitz with mobilization of the duodenum. Gastrojejunostomy has been reported in literature in treating such condition but has increased postoperative complication like blind loop syndrome and recurrence of symptoms. These procedures have more recently been performed laparoscopically and the anastomses formed using an endoscopic gastrointestinal anastomotic (GIA) stapler.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Kushwaha SS, Goja S. Facts and fantasies about superior mesenteric artery syndrome: an unusual cause of intestinal obstruction. Int Surg J 2019;6:2591-4.