

Superior mesenteric artery syndrome in an 18-year old female: Case reportKarishma Lal,¹ Akash Khetpal,² Muhammad Saad Ali Mallick,³ Ansab Godil,⁴ Arsalan Majeed Adam,⁵ Zahid Ali Memon⁶**Abstract**

We report an unusual case of an 18-year old woman, who presented to Civil Hospital Karachi in May 2016 with complaints of vomiting, abdominal pain, dysphagia, altered bowel habits, loss of appetite and chronic weight loss. On examination, abdomen was found to be soft and non-tender with discomfort on breathing. CT angiogram revealed reduction of aortomesenteric angle and aortomesenteric distance which were both consistent with superior mesenteric artery syndrome.

Reduction in angle was thought to be because of weight loss and adipose tissue depletion so patient was started on enteral and parenteral nutritional supplements. Upon seeing little to no improvement, duodenojejunostomy was performed and patient was kept under observation. Nutritional supplements were continued. The after procedure course was uneventful.

Keywords: Superior mesenteric artery syndrome, Aortomesenteric angle, Aortomesenteric distance, Ligament of Treitz.

Introduction

Superior mesenteric artery (SMA) syndrome, also known as Wilkie's syndrome, was first described by Rokitsansky in 1861. It is characterized by extensive compression of 3rd or transverse part of duodenum between aorta and superior mesenteric artery. This results in chronic, intermittent or acute obstruction of the small bowel.¹

It is a fairly uncommon disease, incidence being 0.1-0.3%, which results from loss of fat and lymphatic tissue surrounding the transverse part of duodenum which causes reduction in distance between aorta and superior mesenteric artery. This may be caused by extensive weight loss associated with conditions such as anorexia nervosa, malabsorption, hypercatabolic state (burns, major surgeries or malignancy) or severe congestive

heart failure leading to cachexia.² Clinically, it presents with very vague GI symptoms such as nausea, vomiting, epigastric pain, irregular bowel movements and epigastric fullness.³ The diagnosis can be confirmed by radiological findings which gives a clear evidence of bowel obstruction.

Herein, we present a case of an 18 year old female patient suffering from this rare disease, where history and clinical examination proved to be inadequate. Hence, the diagnosis was made on radiological grounds and then appropriate treatment was employed.

Case Report

An 18-year old female patient presented to Civil

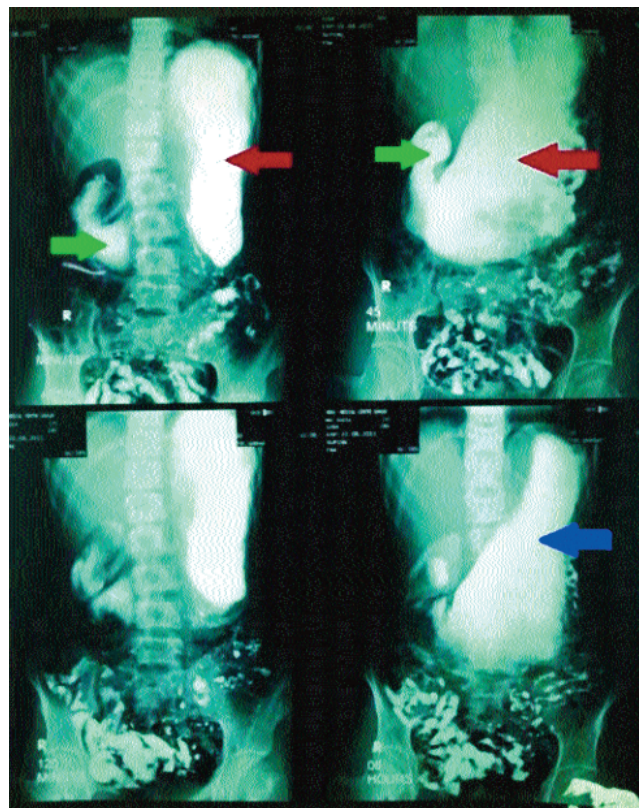


Figure-1: Shows dilatation of stomach (red arrow) as well as 1st, 2nd and 3rd part of duodenum (green arrow). Rest of the bowel loops seem to be undilated. It also shows presence of contrast in stomach at 6 hours (blue arrow) indicating delayed transit.

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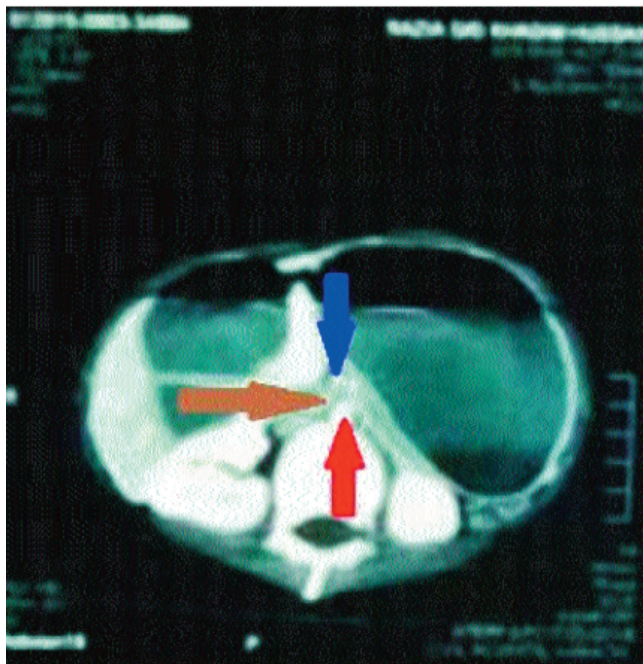


Figure-2: Abdominal aorta (red arrow) and superior mesenteric artery (blue arrow) causing extrinsic compression effect over 3rd part of duodenum (brown arrow) resulting in moderate dilation of stomach and duodenum proximal to compression. Bowel loops distal to compression seem collapsed.

Hospital Karachi in May 2016, with bilious vomiting (post-prandial). She complained of abdominal pain and significant weight loss since a year. She also complained of palpitations, intermittent shortness of breath, burning micturition with increased frequency, alternating bowel habits and inability to walk since 8 months. It was also found that she had dysphagia and epigastric fullness since 5 years. These symptoms explained poor appetite (as reported by the patient's mother) which in turn explained the severe weight loss. Other probable causes of weight loss such as tuberculosis, trauma and abdominal surgery were excluded based on history. On examination, her weight was found to be 29 kilograms (47 kg a year before), BP-90/60, pulse rate -98, respiratory rate- 17/minute and she was afebrile. Abdomen was found to be soft and non-tender with discomfort on breathing. She also looked cachexic and anaemic but there were no signs of jaundice, cyanosis or oedema.

History and clinical examination were inconclusive. Esophagogastroduodenoscopy revealed erosive esophagitis, mild pangastritis and duodenitis with biliary reflux. Barium-meal follow through showed dilated stomach with slow gastroduodenojejunal transit (Figure-1). Contrast CT scan revealed narrowing of the third part of duodenum noted between SMA and

abdominal aorta with proximal massive dilatation of duodenum and stomach which were suggestive of SMA syndrome (Figure-2). CT angiography of abdomen revealed significant reduction of the angle between aorta and superior mesenteric artery to about 20 degrees (normal being 45-60 degrees) and distance between aorta and SMA was found to be 6mm (normal being 10mm). Lower GI malignancies were excluded based on colonoscopy. Lab results revealed electrolyte imbalance with sodium, chloride and bicarbonate ions being low and urea being high. Other laboratory results were unremarkable.

The patient was started on IV fluids as well as nutritional supplements, to manage the rapid weight loss. Prior to oral supplement, a maneuver was performed in which the patient was made to lie laterally which opened the angle subsequently relieving the vomiting. She was also prescribed Omeprazole, Metoclopramide and Aminovel injection (amino acid supplement). Her weight, caloric count and electrolytes were monitored daily to avoid complications.

After no improvement from nutritional supplements for 1 month, the surgeon, upon informed consent from the patient, opted for duodenojejunostomy to correct the obstruction. The procedure involved releasing of the compressed part of the duodenum and creation of an anastomosis between the dilated part of the duodenum and jejunum, anterior to the SMA. A proximal loop of jejunum about 10 inches from the duodenojejunal flexure was placed below the dilated duodenum and fixed with interrupted Vicryl sutures. The duodenal and jejunal lumen were opened by a diathermy and an EndoGIA stapler was inserted and fired. The openings in the two lumens were then closed with continuous full-thickness sutures. Finally, the anastomosis so formed was tested for a leak by instilling a methylene dye solution via a nasogastric tube. Supplements were continued and patient was kept under observation. The patient was followed up for 6 months and weight gain was progressive and was found to be 41 kilograms (roughly 40 percent increase compared to the initial presentation). She also denied any symptoms found earlier. Her post-operative course was uneventful, hence the surgery proved to be successful.

Discussion

Superior mesenteric artery originates from aorta at the level of first lumbar vertebra. It extends inferiorly at an acute angle to aorta which ranges from 38 to 60 degrees normally. At the same level, the third or transverse part of duodenum passes in between

superior mesenteric artery and aorta.^{1,4} Superior mesenteric artery syndrome, as first described by Rokitsky in 1861, is characterized by reduction in aortomesenteric angle to below 25 degrees, which causes compression of the transverse duodenum,^{1,2} which in our case was found to be 20 degrees.

It is an unusual clinical entity, incidence being 0.1-0.3%, with a mortality rate of 33%.^{1,3} It occurs more frequently in women.³ The aortomesenteric angle's width is directly proportional to the BMI of a person, which reduces as the weight of the person decreases, as happened with the patient in our case. The syndrome

can be congenital or acquired. Congenital being lower than normal insertion of SMA or higher than normal insertion of angle of Treitz or shorter ligament of Treitz dislocating duodenum to cranial position.² Acquired causes may include any conditions that cause weight loss or reduction in fat pads between aorta and superior mesenteric artery, which leads to decrease in aortomesenteric distance from 10-28 mm to less than 10 mm in SMA syndrome. These conditions may be anorexia nervosa, malabsorption, cancer, Diabetes Mellitus, AIDS, surgery, traumas, burns or psychiatric illnesses. Other conditions that contribute towards the disease include increased lumbar lordosis, application of body casts or aneurysms in abdominal aorta.⁵

Symptoms of superior mesenteric artery syndrome are non-specific. The patient may present with some or all of the upper GI obstruction symptoms, like bilious vomiting, nausea, epigastric pain, early fullness and post prandial discomfort.^{1,5} The symptoms are temporarily relieved by knee to chest positioning of the body, or with Hayes maneuver in which pressure is applied below navel upward and towards the back causing the superior mesenteric artery to be lifted up.⁵ Left lateral positioning of the body during feeding could also be done to allow mesentery along with the small bowel to move towards the left opening up the angle between aorta and superior mesenteric artery.³ Diagnosis can be made on the basis of CT scan, angiography, endoscopy, ultrasound or MRI. Barium radiography is also used for the diagnosis of superior mesenteric artery syndrome but it is non-specific since the findings may also be present in case of megaduodenum.⁶

The aim of the treatment is to reverse or correct the symptoms that have arisen because of obstruction of transverse part of duodenum. However, to date, there are no proper guidelines for the treatment of superior mesenteric artery syndrome. The longest treatment has

lasted for is 169 days.³ If it is diagnosed earlier, measures can be taken to reverse it by removing the causes responsible. For weight loss, nutritional supplements should be given. If it is a late diagnosis, duodenojejunostomy is performed which is successful in 90 % of the cases. This is the most sought-after intervention and it turned out to be beneficial in our patient as well. Strong operation, devised by Strong in 1958, has also proved successful. He suggested the mobilization of duodenum by sectioning the ligament of Treitz.⁷ This procedure maintains bowel integrity and thus is less invasive and quicker. However, this procedure can be aggravating and become impossible due to adhesion formation.⁶ It also has a failure rate of 25%.² Gastrojejunostomy, another procedure commonly employed provides adequate gastric decompression but can lead to incomplete release of duodenal obstruction leading to blind loop syndrome, gastric bile reflux and ulceration.⁶ Subtotal gastrectomy and Billroth II gastrojejunostomy, and repositioning of duodenum anteriorly are some of the other less commonly used options when going for a surgical intervention.⁵ The aforementioned interventions proved successful in a study of seven patients by M. Ezzedien Rabie.⁸ These patients, one male and six females, had mean age of 17.1 years. Four of these patients responded to medical treatment which consisted of decompression of stomach and duodenum by nasogastric tube followed by total parenteral nutrition (TPN) or small oral meals. Two of the patients were surgically treated with primary duodenojejunostomy and responded well while the third required laproscopic sectioning of ligament of Treitz.⁸

Conclusion

SMA, although rare, should be considered as one of the differentials in the presence of partial or complete gastrointestinal obstruction. Conservative management via nutritional supplementation is first-line therapy and aims to restore the fat content of the body in a bid to increase the distance between the aorta and the superior mesenteric artery which gave rise to the syndrome in the first place. Surgical treatment is adopted in refractory or chronic cases and with the advent of laparoscopic techniques, surgical procedures like laparoscopic duodenojejunostomy have become feasible solutions.

Consent: Informed consent was obtained from the patient to reproduce her case in this report.

Disclaimer: The material submitted to Journal of the

Pakistan Medical Association is new, original and has not been submitted to another publication for concurrent consideration.

Conflict of Interest: None to declare.

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