A Rare Case Presentation: Superior Mesenteric Artery Syndrome.

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Abstract:
Superior mesenteric artery (SMA) syndrome is a rare disorder that is caused by a reduction in the aortomesenteric angle causing a duodenal obstruction, most frequently occurring in patients who have had rapid weight loss. Identification of this syndrome can be a diagnostic dilemma and is frequently made by exclusion. The symptoms are caused by compression of the third portion of the duodenum against the posterior structures by a narrow angled superior mesenteric artery. When nonsurgical management is not possible or the problem is refractory, surgical intervention is necessary.

We report a case of SMA syndrome in a 22 years old female patient, its diagnostic challenges and treatment options.

Keywords: Superior mesenteric artery syndrome, Duodenal obstruction, Weight loss, Duodenoojejunostomy.

Introduction:
Superior mesenteric artery syndrome is an interesting phenomenon that occurs when the Superior mesenteric artery (SMA) compresses the third part of the duodenum. SMA syndrome was first described by von Rokitanski in 1861. It is eponymous with Wilkie, who published the first case series of 75 patients in 1927 and is also known as Wilkie's syndrome and Cast syndrome [1].

The SMA branches off the aorta at the level of L1 and passes anteriorly and inferiorly traversing over the duodenum, left renal vein and the uncinate process of the pancreas. The presence of the fat pad normally creates an angle of more than 35° between the SMA and the aorta however this is decreased to less than 25° in SMA syndrome and occurs due to loss of the fat pad that is normally between the duodenum and SMA, with resultant vascular compression of the third part of the duodenum.

Radiological confirmation of SMA syndrome classically demonstrates the decreased aortomesenteric angle with dilatation of the second part of the duodenum and compression of the third part. The aortomesenteric distance is less than 8 mm [2,3].

Case Report:
A 22 years old female presented to our emergency room with complaints of dull epigastric abdominal pain and vomiting of 2 days duration. Radiological investigations like X-ray erect abdomen and USG abdomen were done in the ER which were found to be unremarkable. It was thought to be a case of gastritis and symptomatic treatment was given which relieved the patient of her complaints. She was advised to attend Surgical OPD later on for upper GI endoscopy.

After 1 week, patient comes with similar complaints and reveals that she has been experiencing these episodes for up to 1 month associated with abdominal pain, bloating and gastric distension followed by vomiting. Pain seems to be occurring 1-2 hours after taking food.

Upper GI endoscopy revealed multiple erosions in stomach and duodenum. Patient was treated conservatively with PPI’s.
All the routine blood investigations were within normal limits. 2 weeks later she was kept on H.Pylori treatment regimen when symptoms did not subside. Patient was still having the same complaints and had lost weight during this 2 months. We decided to further investigate the patient and ordered a Barium meal follow through (BMFT) which revealed Dilated 2nd part duodenum; Vertical linear compression on 3rd part duodenum; partial obstruction to flow in 3rd part duodenum [Figure 1]. So a diagnosis of SMA syndrome was arrived at and patient was informed about the management aspects, she opted for surgery. We did Duodenojunostomy with release of Treitz ligament [Figure 2,3]. Post operative period was uneventful. Patient is on follow-up for 6 months now with no complaints and has gained weight.

Discussion:
SMA syndrome is a rare pathology with an incidence that ranges between 0.013 and 0.3% [4]. 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 humans, the aorta-SMA angle ranges from 38 to 65°, due to the erect posture. The main anatomic feature of SMA syndrome is a narrowing of the aorta-SMA angle to <25°, and as a result, the aortomesenteric distance decreases to <10 mm, from normally 10 to 28 mm [5,6].

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 [7] 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 [8]. Severe weight loss, leading to a depletion of the fatty cushion around the SMA is a major cause of SMA syndrome. Catabolic states like burns [9], eating disorders such as anorexia nervosa [10], or wasting conditions such as neoplastic diseases and malabsorptive states are the most commonly reported reasons of drastic weight loss.

Patients may present insidiously with a longstanding history of intermittent nausea, vomiting and epigastric pain, which classically is relieved by lying in the left lateral decubitus position. Alternatively, acute onset may occur in association with rapid weight loss, particularly in surgical patients characterized by signs and symptoms of duodenal obstruction [5].

The diagnosis of the SMA syndrome is challenging and often delayed due to its insidious presentation. High clinical suspicion is warranted and diagnosis is based on clinical evidence supported by radiological findings.
Barium radiography demonstrates dilatation of the first and second part of the duodenum with or without gastric dilatation, anti-peristaltic flow of barium proximal to the obstruction and a delay of 4-6 hours in gastroduodenjejunal transit time, with relief of obstruction when the patient is placed in the prone, knee-chest or left lateral position [4,11]. Contrast-enhanced CT or magnetic resonance angiography enable visualization of the vascular compression of the duodenum and precise measurement of the aortomesenteric angle and distance. Endoscopic examination may visualize a pulsatile extrinsic compression suggestive of this condition [12].

Conservative management is attempted initially through high calorie intake via parenteral feeding, decompression of the small bowel with a nasogastric tube, fluid resuscitation and correction of electrolyte abnormalities. Enteral feeding may be introduced with a nasojejunal tube placed distal to the obstruction [4]. Posturing maneuvers during meals and motility agents may be helpful in some patients. No time limit has yet been defined for the medical treatment. Surgery may be considered if conservative treatment fails.

Duodenojunostomy is the operation of choice to relieve the obstruction, with a success rate up to 90%. Another less invasive surgical option, known as Strong's procedure, involves lysis of the ligament of Treitz with mobilization of the duodenum. 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 [5,11]. These procedures have more recently been performed laparoscopically and the anastomoses formed using an endoscopic gastrointestinal anastomotic (GIA) stapler. They have been reported as safe and feasible techniques. Experience is limited due to the sporadic presentation of the disease and the infrequent number of operations associated with it however outcomes have been favourable with limited complications in these small studies [13,14].

Conclusion:
We feel that SMA syndrome is under diagnosed. High clinical suspicion is of utmost importance, especially in patients with severe weight loss and symptoms of epigastric dull postprandial pain, vomitings and gastric distension. We would like to point out the difficulty of achieving an accurate diagnosis of SMA syndrome in an atypical clinical setting similar to what happened in our case. Heightened awareness is advised for early recognition to avoid unnecessary suffering for the patient.

References: