Increasing Trend of Surgical Management in Superior Mesenteric Artery Syndrome

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Abstract

Superior Mesenteric Artery Syndrome (SMA syndrome), a diagnosis of exclusion, since first described in 1861 by Von Rokintansky up till date remains a diagnostic dilemma to physicians. It is characterized by a reduced aorto-mesenteric angle and aorto-mesenteric distance, leading to compression of the third part of duodenum between the two vessels. Patients who are usually young, thin females present with postprandial abdominal pain and vomiting. We report a case of 17-years female who presented to us with postprandial epigastric pain and vomiting for the last 4 years, underwent multiple unfruitful invasive and non-invasive investigations. Later when diagnosed on magnetic resonant enterography, underwent laparoscopic duodenojejunostomy. Found to be a successful procedure, evident by the gain in weight and improved quality of life for the patient.

Keywords: Superior Mesenteric Artery Syndrome; Duodenojejunostomy; Laparoscopic Surgery

Introduction

Superior Mesenteric Artery (SMA) syndrome also known as Wilkie’s syndrome or Cast syndrome, arterio-mesenteric duodenal obstruction, and chronic duodenal ileus is a rare disorder characterized by compression of the third portion of the duodenum due to narrowing of the space between the SMA and aorta and is primarily attributed to loss of the intervening mesenteric fat pad. Most frequently occurring after weight loss, associated with severe debilitating illnesses, such as malignancy, malabsorption syndromes, AIDS, trauma and burns. It has also been described in a variety of other disorders associated with extreme weight loss including bariatric surgery, spinal cord injury, paraplegia, drug abuse, prolonged bed rest and anorexia nervosa. Surgeries for spinal deformities as well as high insertion of the ligament of Treitz are other potential causes for the occurrence of SMA syndrome [1,2]. One report documented a patient with celiac axis compression, which was believed to be the cause of superior mesenteric artery syndrome. Rare cause includes congenitally short ligament of treitz. Its incidence varies from 0.1 - 0.3%. Symptoms may vary from mild obstruction to acute life-threatening emergencies. Physical examination does not always help in establishing the diagnosis, sometimes mild distension is all what you can see. Radiologic imaging is key in diagnosis specifically CT/MR arteriography [3,4]. Conservative management has its role, but usually requires surgery. Duodenojejunostomy is the preferred procedure for this syndrome [5,6].

Case Presentation

17-years old female presented to gastroenterologist with 4 years history of postprandial abdominal pain and vomiting. Pain was more marked in epigastrium. Nothing abnormal on physical examination. Past history was also not significant. She has been to multiple doctors, done many investigations and tried multiple medicines but nothing helped her. She was labeled as a psychopath by few doctors, and as if she is faking her symptoms. She was in a terrible state of mind. Missing her school and getting confined to the bed. Upon arrival to us,
we did a gastroscopy and found mild gastritis. Hence proton pump inhibitor (PPI) were prescribed. This doesn’t seem to work. Planned to do MR enterography, as the next suspicion for small bowel pathology. MRE showed extrinsic compression at the level of distal duodenum causing poor emptying (Figure 1). Patient was explained about the diagnosis and symptomatic treatment advised, over a year. She visited us again after that with failure to respond to conservative management. At that time case was discussed in our multidisciplinary team meeting and with the decision of all planned for surgery. CT Angiography was done before taking her for surgery, which confirms the diagnosis and showed an aorto-mesenteric angle of 5 degrees and aorto-mesenteric distance of only 7 mm. There was loss of fat plane as well (Figure 2). Laparoscopic duodenojejunostomy was done, using three ports and staplers were used for creating side to side anastomosis. Patient recovered gradually well, diet was started gradually. She was discharged on 4th post-operative day. After a couple of weeks she came back with vomiting and abdominal pain, conservative measures made her improve. CT scan was also done that showed mild edema at the anastomosis site. She continued to have mild symptoms after that and we repeated an endoscopy that was inconclusive. Her symptoms have gradually settled almost completely after about 4 weeks and she is tolerating diet and enjoying her life (Figure 3).

\textit{Figure 1: MRI showing extrinsic compression of distal duodenum.}

\textit{Figure 2: Showing aorto-mesenteric angle of 5 degrees, aorto-mesenteric distance of 7 mm and loss of fat plane causing duodenal compression between two vessels (blue lines showing the angle and distance).}

Discussion

SMA syndrome is an uncommon cause of duodenal obstruction. The SMA usually forms an angle between 38 - 56 degrees with the aorta. SMA syndrome presumed to be present when this angle is less than 25 degrees. The aorto-mesenteric distance which normally is 10 - 20 mm can be decreased to 2 - 8 mm in these cases [3,4]. Symptomatology is consistent with proximal small bowel obstruction, may present acutely or insidiously. Some patients may have only postprandial pain in epigastrium and early satiety, while some can present with advanced obstruction, with bilious vomiting and weight loss. Symptoms may relieve by lying prone, left decubitus or knee chest position [2], relieving tension from the mesentery and SMA, opening the space between SMA and aorta.

Physical examination can include abdominal distension, succession splash and high-pitched bowel sounds. Laboratory examination can be normal, only if patient have severe vomiting electrolyte imbalance is evident. Patients with clinical suspicion should undergo further investigations, starting from plain films to CT and MR arteriography all can be done in a stepwise pattern. Of which CT and MR arteriography are considered gold standard for the diagnosis. Once diagnosed the next step in management is to give the patient a trial of conservative measures, including gastrointestinal decompression, correction of electrolytes and nutritional support. If the conservative management fails, patient should be considered for surgical treatment. Surgery can be done laparoscopic or open, depending on the expertise. Strong’s procedure was the earliest procedure described for this syndrome, it involves division of the ligament of treitz. Some surgeons have also performed gastrojejunostomy. Both of these have inferior results to duodenojejunostomy [5,6] according to multiple studies, in term of resolution of symptoms, post op complications and length of hospital stay.

Conclusion

Still after ages SMA syndrome remain an uncommon entity. Most of the cases are misdiagnosed or diagnosed very late. A high index of suspicion is required especially in young females. CT/MR arteriography should be the imaging modality and diagnosis should base on the aorto-mesenteric angle and distance. Increasingly surgical treatment is becoming more successful, and majority of patients are symptom free after that.

Bibliography


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