Superior Mesenteric Artery Syndrome: A Rare Cause of Intestinal Obstruction

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Abstract: Superior mesenteric artery (SMA) syndrome is a rare cause of gastrointestinal tract obstruction. It is accompanied with epigastric pain and vomiting. Normally, the angle created by aorta and SMA is between 45-60 degrees. Any factor that narrows the aorto-mesenteric border (less than 10 degrees) can cause entrapment and compression of the third part of the duodenum, leading to SMA syndrome. We recently cared for a patient who presented with typical clinical symptoms and was eventually diagnosed with SMA syndrome. Radiological investigations form the mainstay for diagnosing the condition. X-ray abdomen standing of our patient showed "double bubble sign". Computed tomography of abdomen and pelvis demonstrated duodenal compression between abdominal aorta and superior mesenteric artery. SMA angiography would be the gold standard investigation in these cases. These patients can complicate with electrolyte disturbances, gastric perforation, gastric pneumatosis and portal venous gas, obstructing duodenal bezoar. Many patients can be managed medically, by resting the bowel after decompressing it and correcting electrolyte disturbances. However, patients with long standing history should be treated surgically. Looking at the rarity of the condition, we hope this case report will help educate the readers about the condition and would highlight the importance of increased vigilance required on the part of clinicians to diagnose this condition.

Keywords: abdominal pain, case report, India, superior mesenteric artery.

I. Introduction

Superior mesenteric artery (SMA) syndrome is a rare cause of gastrointestinal tract obstruction. It poses a great diagnostic challenge to the surgeon and clinician and the patient may be misdiagnosed leading to life threatening complications. We recently cared for a patient who came in with features of intestinal obstruction who was eventually diagnosed with SMA syndrome. This report highlights the importance of hightened vigilance required by clinicians in order to diagnose SMA syndrome in a timely fashion.

II. Case Report

A 21 year old married female presented with complaints of epigastric pain for 4-6 days, associated with greenish vomiting and abdominal cramps. Her cramps were aggravated after meals or in supine position and were relieved by changing to prone position. She had no history of fever, loose stools, constipation, decreased urination, eating disorder or medication intake. Her past medical and menstrual histories were insignificant and she had no history of any abdominal surgery.

Physical examination revealed a thinly built afebrile patient. Her heart rate was 92/min, blood pressure was 110/70 mm Hg, respiratory rate was 18/min and general examination showed pallor. Abdominal examinaton revealed tenderness in epigastrium and right hypochondrium. Rest of the systemic examination was within normal limits. With acute gastritis, pancreatitis, cholecystitis and subacute intestinal obstruction in mind the patient was investigated further. Haematological investigations showed mild anemia (hemoglobin 10.4 mg/dL) and low platelet count (40,000).

The patient had normal electrolytes, random blood sugar, serum bilirubin, serum amylase, blood urea nitrogen, serum alkaline phosphatase and negative serologies for dengue and leptospirosis. Electrocardiogram and chest x-ray were normal but abdominal ultrasound showed distended abdomen and proximal duodenum. X-ray abdomen erect exhibited a double bubble sign. Computed tomography (CT) abdomen and pelvis demonstrated duodenal compression between abdominal aorta and superior mesenteric artery (Figure 1 and 2). In light of these investigations, a final diagnosis of superior mesenteric artery (SMA) syndrome was made. Patient was kept nil by mouth and her gastrointestinal system was decompressed with a nasogastric tube. Patient was hydrated and treated with broad antibiotic coverage, antacids and antiemetics. Patients' symptoms were relieved and she was followed up with normalization of her nutritional and haematological parameters.

III. Discussion

SMA syndrome is also known as "Cast Syndrome", "Wilkie Syndrome", "Arteriomesenteric Duodenal obstruction" and Chronic Duodenal Ileus". It was first described by von Rokitanski in 1861. [1] Later Wilkie described the anatomical, clinical and patho-physiologic basis of the disease and published a series of 75 cases in 1927.[2] SMA syndrome is a rare but well recognised clinical entity, with incidence varing from 0.013-0.3% in barium series of upper GI tract.[2,3]

Third portion of the duodenum passes between the aorta and SMA around third lumbar vertebra. It is suspended in position by the ligament of Treitz. Normally, the angle created by these two blood vessels is between 45-60 degrees. The mesenteric fat pad also helps in maintaining this angle. Any factor that narrows the aorto-mesenteric border (less than 10 degrees) can cause entrapment and compression of the third part of the duodenum, leading to SMA syndrome (Figure 3).

Although seen mainly in adolescents or young adults, vascular compression of the duodenum can present at any age. [4] The exact etiology is not known but various risk factors for developing SMA syndrome have been identified, including significant weight loss (due to malignancy, malabsorption, anorexia, trauma, wasting diseases), surgical correction of scoliosis, congenital short ligament of Treitz, gravid uterus reducing abdominal cavity volume, retroperitoneal hematoma following aneurysmal repair and prolonged bed rest. Patient typically present with symptoms consistent with acute or chronic small bowel obstruction, like early satiety, post prandial epigastric pain, nausea and vomiting, bilious vomiting and abdominal distension with high pitched bowel sounds. Characteristically symptoms reside when the patient lies prone or on left side, as was the case with our patient, however it is not a uninversal finding. These patients can complicate with electrolyte disturbances (hypokalemia, metabolic alkalosis), gastric perforation, gastric pneumatosis and portal venous gas, obstructing duodenal bezoar. Moreover, peptic ulcer disease coexists with SMA syndrome in up to 25% of cases. [5,6,7]

As a rare cause of small bowel obstruction, patients generally require many investigative work up studies to arrive at the diagnosis, as was the case in our patient. A formal diagnosis requires extrinsic compression on duodenum between SMA and aorta, a distended duodenum and an aorto-mesenteric angle less than 20 degrees. Intestinal peristalsis should still be present. Clinical diagnosis has to be confirmed by radiological studies. Akin et al demonstrated that a diagnosis of vascular compression of duodenum was made radiologically in 95% of the cases they reported. [8] X-ray abdomen standing generally shows dilatation of stomach and proximal duodenum. Barium study would show gastric and duodenal dilatation with contrast retention and marked delay in passage of contrast from duodenum to distal small bowel. [9] Unfortunately, these findings are non specific for SMA syndrome and may be seen in other conditions like diabetes, pancreatitis, scleroderma, malignant lymph nodes or tumors in mesentery root.[10] CT/Magnetic resonance imaging, similar to barium study, would show intra abdominal fat concentration and other intra abdominal pathology. SMA arteriography would demonstrate and confirm the narrowing of the angle between SMA and aorta.

Medical management is usually successful in patients with a short history. Conservative management includes resting the bowel after decompression with a nasogastric tube, correcting fluid and electrolyte imbalance and providing nasojejunal feeds or total parenteral nutrition. Surgery is indicated for chronic and long standing cases or after failure of medical management. Duodenal mobilization and repositioning is an option, enabling the duodenum to free itself from the sharp aorto-mesenteric angle. [11] Duodenojejunostomy has been described to be effective in 90% of the patients. [12] Patients may require further care for refractory gastroparesis caused as a result of corrective surgery.

IV. Conclusion

SMA syndrome is a rare cause of small bowel obstruction and our report provides an overview of steps involved in diagnosing and managing the condition. High degree of clinical suspicion and radiological investigations are required to diagnose the condition. Depending on the duration of the diasease and other factors, such patients can be managed medically or surgically.

References

- [1]. Rokitanski CV. Vienna. 3rd edn. Vol. 3. Braumulller and Siedel: 1861. Lehrbuch der Pathologische Anatomie; p. 187.
- [2]. Wilkie D. Chronic Duodenal ileus. Am J Med Sci. 1927;173:643–649.
- [3]. Superior mesenteric artery syndrome: diagnosis and treatment from the gastroenterologist's view. Lippl F, Hannig C, Weiss W, Allescher HD, Classen M, Kurjak MJ Gastroenterol. 2002; 37:640-3.
- [4]. Ylinen P, Kinnunen J, Hockerstedt K. Superior mesenteric artery syndrome a follow-up study of 16 operated patients. J Clin Gastroenterol 1989;11:386-91.
- [5]. Lundell L, Thulin A: Wilkie's syndrome— a rarity? Br J Surg 1980;67:604-6.
- [6]. Hutchinson DT, Bassett GS. Superior mesenteric artery syndrome in pediatric orthopedic patients. Clin Orthop 1990;250:250-7
- [7]. Anderson WC, Vivit R, Kirsh IE. Arteriomesenteric duodenal compression syndrome. Its association with peptic ulcer. Am J Surg 1973;125:681-9.

- [8]. Akin JT, Gray SW, Skandalakis JE. Vascular compression of the duodenum: presentation of 10 cases and review of the literature. Surgery 1976;79:515-22.
- [9]. Selami S, Hayrunisa K, Arzu A, Gulsen E, Haluk GB. Incomplete duodenal obstruction in a newborn. Indian J Paediatr 2006;73:364-6.
- [10]. Konen E, Amitai , Apter S et al. CT Angiography of Superior Mesenteric Artery Syndrome. Amer J Roentgenol 1998;171:1279 -81.
- [11]. Laffont I, Bensmail D, Rech C, Prigent G, Loubert G, Dizien O. Late superior mesenteric artery syndrome in paraplegia: case report and review. Spinal Cord 2002;40: 88-91.
- [12]. Palanivelu C, Rangarajan M, Senthilkumar R, Parthasarathi R, Jani K. Laparoscopic duodenojejunostomy for superior mesenteric artery syndrome. J Soc laparoendoscopic Surgeons 2006;10:531-4.

Figures

Figure 1. Axial Computed Tomography section demonstrating duodenal compression between the abdominal aorta and superior mesenteric artery



Figure 2. Axial Computed Tomography demonstrating duodenal compression



Figure 3. Pictorial representation of the angle between superior mesenteric artery and aorta. Courtesy of Sherry Scovell, MD.

