

Superior Mesenteric Artery Syndrome in Adolescents

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INTRODUCTION

Superior mesenteric artery (SMA) syndrome, also known as Wilkie's syndrome or Benign duodenal stasis is a rare cause of proximal small bowel obstruction and is linked to notable morbidity and mortality when the diagnosis is delayed. In the patients with SMA syndrome, the third portion (transverse part) of the duodenum is compressed externally between the SMA and abdominal aorta leading to duodenal stasis and gastrointestinal obstruction.(1)(2)Clinical symptoms include post-prandial pain, nausea, vomiting and weight loss.(3)While superior mesenteric artery syndrome is rare, the morbidity and mortality associated with its complications makes it a crucial differential to consider when concerned for bowel obstruction, especially in the setting of recent weight loss. Conservative management for SMA syndrome often fails, and laparoscopic duodeno-jejunostomy proves to be safe and effective as optimal definitive treatment.(5)

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The disease was first described in 1842 by Carl Von Rokitsky, and in 1927, Wilkie further detailed the pathophysiology and diagnostic findings of the disease.

CASE DESCRIPTION

A 15 year old female presented in OPD of Era's Lucknow Medical College (ELMCH) with complaints of pain in abdomen since 8 months associated with belching and vomiting. The pain was mainly localised in epigastric region, used to get relieved on taking medication and was associated with belching and vomiting containing food particles. As a result, her food intake dropped significantly and she had significant reduction of weight in past 3 months (BMI was 14.8 kg/m² at presentation). At the onset of symptoms, an endoscopy was done suggestive of diffuse ulceration in fundus and body of stomach, biopsy of which stated H.pylori associated gastritis. She was treated with clarithromycin, metronidazole and pantoprazole as per protocol but there was no relief in symptoms. She was further investigated for Subacute Intestinal Obstruction and barium meal follow through was done showing partial gastric outlet obstruction at 2nd and 3rd part of duodenum with dilated stomach. Her CT enterography demonstrated distension of stomach, 1st and 2nd part of duodenum with fluid and air level, narrowing of the aorto-mesenteric angle 18° and distance was 5 mm. Bowel loops proximal to this junction were dilated while the distal loops were collapsed- suggestive of SMA syndrome (Fig-1).

She was then transferred to pediatric surgery unit of ELMCH and surgical intervention planned. Diagnostic laparotomy followed by Strong procedure was done under

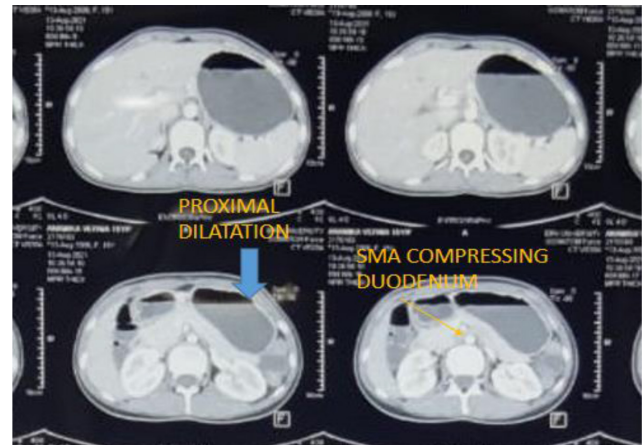


Figure 1: CT Scan : Showing SMA compressing Duodenum

general anesthesia. Post operative stay was uneventful and child improved clinically thereafter.

DISCUSSION

The diagnosis and treatment of SMA syndrome is particularly challenging due to its insidious and often vague presentation.(6)Its prevalence is 0.013-0.3% based on imaging studies. Overall, females are more commonly affected and two-thirds of patients are in the age group between 10 and 39 years.(3)(4)

Usually, SMA syndrome can present with an acute occurrence, such as a duodenal obstruction, or more insidiously,

such as our patient who presented with long standing vague abdominal pain, early satiety, anorexia and vomiting.(7)This condition mainly affects female patients, older children, adolescents and even underweight individuals with a history of rapid weight loss.(4)(8)

Few cases of SMA syndrome can be recognized by upper endoscopic examination which shows a pulsatile extrinsic compression but this is highly operator dependent finding and was not helpful in our patient.CECT criteria for diagnosis of SMA syndrome include an aortomesenteric angle of less than 22° and an aortomesenteric distance of less than 8-10 mm.(9)

As far as treatment of SMA syndrome is considered, although many patients require surgery but one series has demonstrated that conservative management such as nasogastric decompression, hyperalimentation followed by oral feedings, frequent small meals with prokinetics and proton pump inhibitors is able to avert surgical intervention.(4) However, surgery was required in our patient.

In conclusion, SMA syndrome is still adiagnostic and therapeutic challenge with endoscopy being less helpful and diagnosis is mostly confirmed by CECT scan. It is seen mostly in females, with long standing and chronic onset. However, further prospective studies, with a larger number of patients, are needed to frame better diagnostic and management guidelines of SMA syndrome.

FURTHER READING:

1. Zaraket V, Deeb L. Wilkie's Syndrome or Superior Mesenteric Artery Syndrome: Fact or Fantasy. *Case Rep Gastroenterol* 2015;9:194-199.
2. Shi Y, Shi G, Li Z, Chen Y, Tang S, Huang W. Superior mesenteric artery syndrome coexists with Nutcracker syndrome in a female: a case report. *BMC Gastroenterol*. 2019 Jan 23;19(1):15.
3. Welsch T, Büchler MW, Kiele P: Recalling superior mesenteric artery syndrome. *Dig Surg*2007;24:149-156.
4. Sinagra E, Raimondo D, Albano D, et al. Superior mesenteric Artery Syndrome: Clinical, Endoscopic, and Radiological Findings. *Gastroenterol Res Pract*. 2018;2018:1937416. Published 2018 Aug 27.
5. Ehlers TO, Tsamalaidze L, Pereira L, Stauffer J. Laparoscopic Duodenojejunostomy for the SMA Syndrome. *ZentralblChir*. 2018 Oct;143(5):461-463.
6. E. Sinagra, L. M. Montalbano, C. Linea et al., "Delayed-onset superior mesenteric artery syndrome presenting as oesophageal peptic stricture," *Case Reports in Gastroenterology*, vol. 6, no.1, pp.94-102, 2012.
7. C. Foster and A. Choudhary, "Severe malnutrition causing superior mesenteric artery syndrome in an adolescent with Triple A syndrome," *Journal of Pediatric Endocrinology and metabolism*, vol.29, no. 10, pp. 1221-1224, 2016.
8. P. Vulliamy, V. Hariharan, J. Gutmann, and D. Mukherjee, "Superior mesenteric artery syndrome and the "nutcracker phenomenon"," *BMJ Case Reports*, vol.2013, 2013.
9. M.E. Rabie, O. Ogunbiyi, A.S. Al Qahtani, S.B.M. Taha, A. El Hadad, and I. El Hakeem, "Superior mesenteric artery syndrome: clinical and radiological considerations," *Surgery Research and Practice*, vol.2015, Article ID 628705, 5 pages, 2015.