Case report

SUPERIOR MESENTERIC ARTERY SYNDROME IN LATE MIDDLE-AGED MAN; CLINICO-RADIOLOGICAL FINDINGS.

ABSTRACT

Superior mesenteric artery syndrome (SMAS) is a rare cause of duodenal obstruction due to compression of the third part of duodenum between Superior mesenteric artery (SMA) and aorta. Clinical suspicion and radiological findings are playing important role in the diagnosis of SMAS. Most of the patients diagnosed with SMAS are managed conservatively, with surgery reserved for refractory cases not responding to medical treatment. Here in we report a 55-year-old male presenting with acute recurrent abdominal pain and vomiting, diagnosed to have SMAS who underwent surgical correction and describe the clinical and radiological findings.

Keywords

Aortomesenteric angle, Aortomesenteric distance, Superior mesenteric artery syndrome,

INTRODUCTION

Superior mesenteric artery syndrome (SMAS), also known as Wilkie syndrome, cast syndrome or Aorto-mesenteric duodenal compression syndrome, is an acquired vascular compression disorder in which acute angulation of SMA and aorta, results in compression of the third part of the duodenum leading to signs of obstruction or intermittent post-prandial pain [1]. The most common symptom in SMAS is intermittent abdominal (epigastric) pain and vomiting. Nausea and vomiting may affect in anorexia and weight loss. Reduced fat between the SMA and aorta either due to poor nutritional intake or weight loss results in reduced aortomesenteric angle (AMA) and aortomesenteric distance (AMD) [2]. Prevalence of the SMA syndrome varies from 0.2% to 0.78% in the barium series of the upper

gastrointestinal tract [3-5]. Manoeuvres similar as lying prone or espousing the left lateral decubitus position are described as reducing the pain [6].

SMAS is frequently an over-looked entity, and diagnosis can be delayed or even missed if not clinically suspected or known by the attending physician. High index of clinical suspicion and radiological findings plays important role in the diagnosis of this syndrome.

CASE REPORT

A 55-year-old Indian male presented to our hospital with prolonged history of intermittent abdominal pain and vomiting. There was history of characteristic relief of symptoms in prone position. Significant weight loss was present in past 1-year. He was previously hospitalized multiple times for similar complaints.

On general examination his vitals were stable. Abdominal examination revealed epigastric fullness. Cardiac, respiratory, and neurological examination was normal. Routine blood and urine investigation, liver function tests were within normal limits. Plain erect chest radiograph was normal. Plain and oral contrast erect abdomen (**Figure.1A, B**) showed double bubble sign suggestive of duodenal obstruction.

Upper gastrointestinal barium meal study revealed dilated stomach, first (D1) and second (D2) part of duodenum with a significant luminal narrowing at the third part of duodenum without any intrinsic mucosal abnormality **(Figure.2 A, B)**. Distal bowel loops were opacified with barium on prone spot films **(Fig.3A, B).**

Then contrast-enhanced computed tomography (CECT) was advised for confirmation. CECT revealed reduced AMA (20°) and short AMD (6.7 mm). Compression was seen at D3 level with dilatation of stomach and proximal duodenum (D1 and D2) **(Figures.4A, B)**.

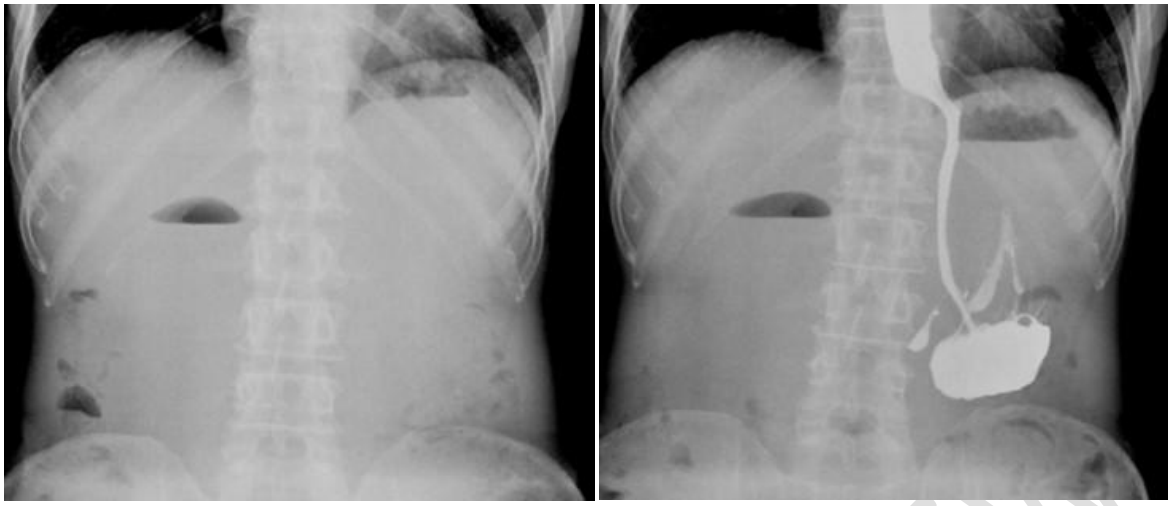

Fig.1(A, B). Abdominal plain (A) and contrast (B) erect radiograph demonstrates air fluid levels in left & right hypochondrium (Double bubble sign) suggestive of duodenal obstruction.

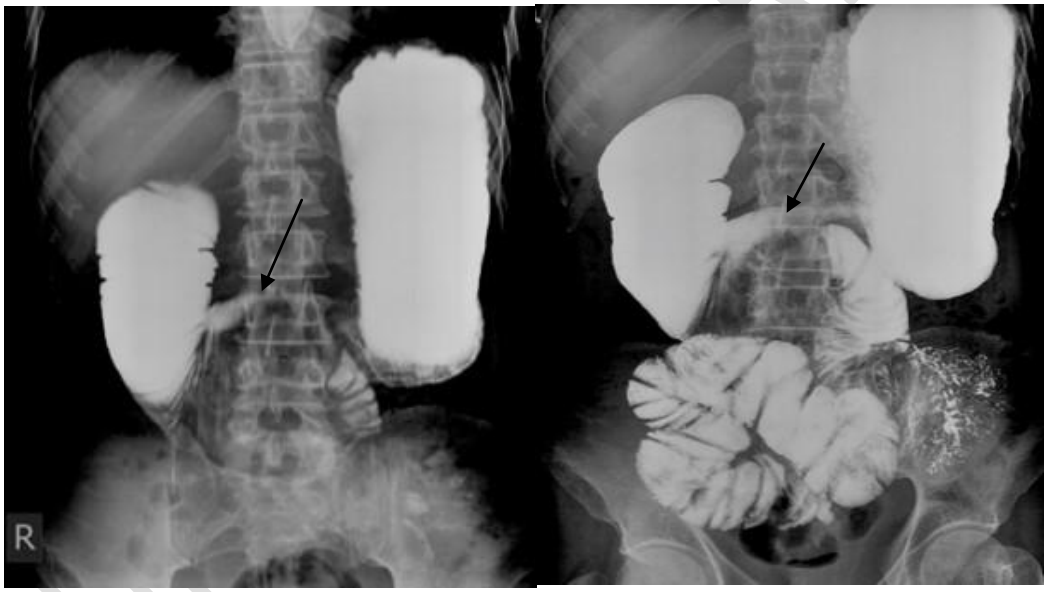

Fig.2(A, B). Barium meal studies. Preoperative supine spot images of study demonstrating dilated stomach & duodenum with curvilinear extrinsic lucency (arrow) corresponding to superior mesenteric artery impression upon the third part of duodenum.

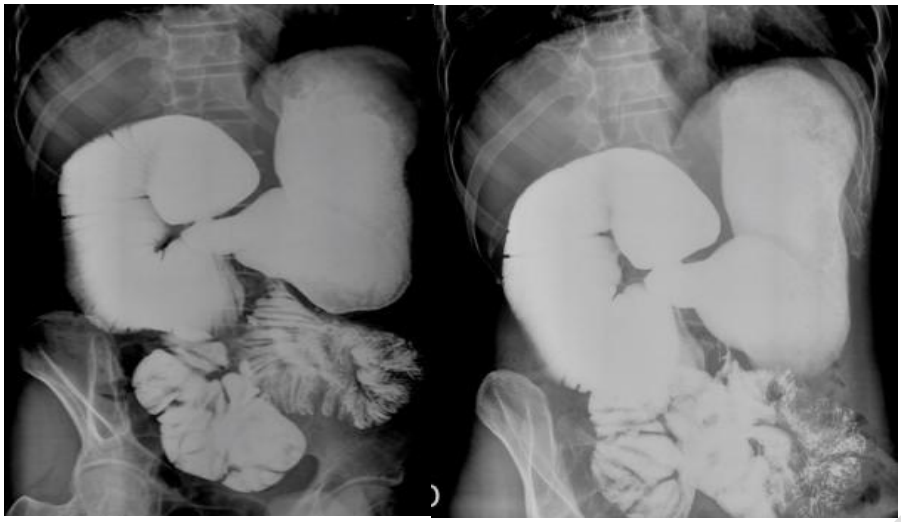

Fig.3(A, B). Preoperative Barium meal Prone spot images of study shows dilated stomach, duodenum emptying into the jejunum.

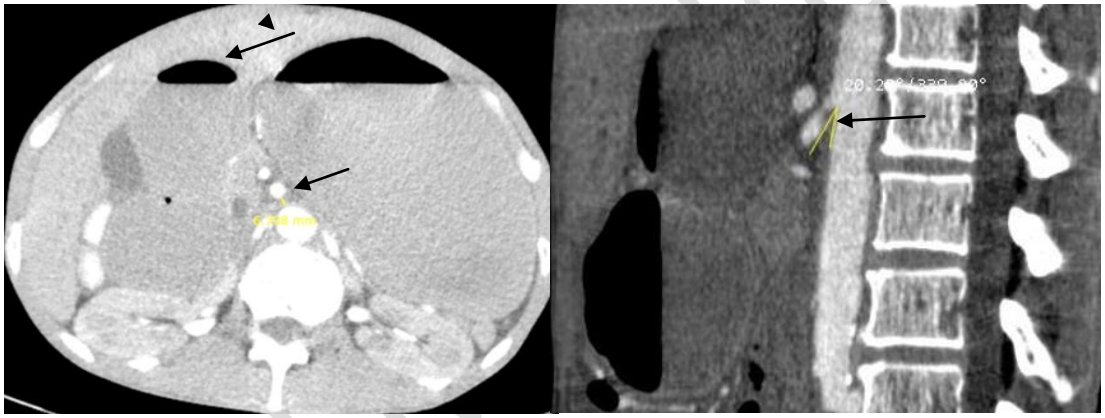

Fig.4(A, B). CT criteria suggestive of SMAS. Selected contrast enhanced A) mid abdominal axial and B) mid sagittal images demonstrating reduced AMD (small vertical line in region of interest in image A), dilated proximal second part of duodenum (arrow in image A), and reduced AMA (arrow in image B).

Computed tomography (CT) images **(Fig.4A, B**) were reviewed for AMA (measured on mid sagittal image between abdominal aorta and SMA, AMD (measured on axial image at level of horizontal third part of duodenum between abdominal aorta and SMA), and proximal duodenal dilatation. An AMA below 25° and AMD below 8 mm were considered positive for SMAS.

The abdominal ultrasound (US) showed dilated stomach & duodenum.

On the basis of clinical and radiological findings, a diagnosis of SMAS was suggested. Initial conservative approach was tried with nasogastric drainage, fluid electrolyte therapy, and small frequent meals, but because of its failure the patient was managed by duodeno-jejunostomy. At present our patient is symptom-free.

DISCUSSION

Foremost description of the SMAS was given by Carl Freiherr Von Rokitansky in 1861 and laterly by Wilkie in 1927, who handed a more detailed anatomical, clinical, and pathophysiological aspect of this syndrome [5]. The frequence of SMAS grounded on fluoroscopy studies is estimated to be 0.013–0.3% [7], whereas other sources quote a frequence of 0.0965% in a chronic hospital setting versus 0.00108–0.0052% in an acute general hospital setting [8]. There is a slight womanish preponderance (64–66%), with 75% of cases being in individualities who range in age from 10 to 39 years [9, 10].

In humans, SMA leaves the aorta at an acute downward angle. This results in the conformation of vascular angle between aorta and SMA through which the third part of duodenum passes [5]. The duodenum is typically girdled by mesenteric adipose tissue as it traverses the aortomesenteric plane. This tissue functions as a natural adipose cushion and prevents extrinsic compression [9]. Normal AMD and AMA is 10–28 mm and 25–60° respectively. In SMA syndrome AMA narrowed to 7–22° and AMD reduced to 2–8 mm [1,5] Cut-off values were reported to be 22° for AMA (42% sensitivity and 100% specificity) and 8 mm for AMD (100% sensitivity and specificity) [11]. In our case, CT dimension of AMA and AMD is lower than cut off value.

The clinical presentation may be acute or chronic. The typical symptoms of SMAS are postprandial or intermittent abdominal pain and features of bowel obstruction such as vomiting, nausea, and anorexia [10]. The symptoms are relieved in left lateral decubitus, prone or knee to chest position [12]. Analogous common complaints and relieving factor was present in our case.

There are multiple causes for SMAS like severe cachexia or catabolic state due to neoplasm, anorexia nervosa, severe head injury or burn, prolonged bed rest, bariatric surgery, Nissan fundoplication, spinal scoliosis surgery, spinal deformity, high insertion of ligament of Treitz, intestinal malrotation, peritoneal adhesions, low origin of SMA, and elderly patient with

heavily calcified or tortuous aorta [12]. Patients are often underweight, with a reported median BMI at diagnosis of 17.4[9].

A pubertal growth spurt is a threat factor for SMAS, and weight loss is not necessary for the development of the syndrome in the paediatric population [13]. Acute and inordinate ingestion of water and food can spark a manifestation of SMAS in adolescents whose visceral adipose tissue has formerly been depleted. It is important to consider SMAS in the differential diagnosis of adolescents with intermittent abdominal symptoms [13]. The association of SMAS with acute pancreatitis is infrequently reported [14,15]. Secondary occlusive post papillary syndrome due to SMAS may produce a retrograde reflux of bile into the pancreatic duct, which activates the inflammatory phenomena responsible for pancreatitis [15].

Symptoms resulting from duodenal obstruction include abdominal pain, vomiting, and weight loss. All other conditions of duodenal obstruction, including natural causes such as benign or malignant tumours, and extrinsic causes similar as pancreatitis, mesenteric mass, or aortic aneurysm must be ruled out with customized imaging modalities before a opinion of SMAS is established [6]. Elderly cachexic patient with reduced retroperitoneal fat and abdominal aortic tortuosity was the cause of SMAS in our case.

Upper gastrointestinal fluoroscopy can demonstrate dilatation of the first and second part of the duodenum **(Fig.1A, B),** extrinsic compression of the third part of the duodenum, and a fairly collapsed small bowel distal to point where the SMA crosses the duodenum [16].

Conventional Barium meal and Hypotonic duodenography are important for the diagnosis of SMAS. Still, the radiological appearances of these studies are non-specific, and have been reported as various conditions similar as scleroderma, diabetes, pancreatitis, peptic ulcer, lupus erythematosus, or idiopathic intestinal pseudo-obstruction [3,17]. Barium examination shows perpendicular band of external compression at the third part of the duodenum with proximal gastric and duodenal dilatation **(Figure 2A, B).** Other findings on barium include reverse barium flow or anti-peristaltic flow of barium proximal to the site of obstruction and relief of obstruction when the patient is in a prone **(Fig.3A, B)** or left lateral decubitus [7].

In the history, Angiographic dimension of the AMA was noted as being a more accurate investigation than routine barium meal examination [18]. Still, the procedure is invasive, and some patients are unable to tolerate it.

Lately, Konen et al have noted the advantage of 3-dimensional (3D) CT angiographic reconstruction, which can help to exclude the incorrect diagnoses that appear from the angulations of the SMA [19]. The AMD is the shortest distance between the walls of the

SMA and the aorta at the position where the third part of duodenum passes between them **(Fig.4A).** The AMA is the angle between the SMA and the aorta, which is measured on sagittal MPR images **(Fig.4B).** In SMAS, both parameters are reduced, with values of 6-22 degrees and 2-8 mm, respectively [19]. Imaging features of SMAS on CT include a dilated stomach and proximal duodenum, with an abrupt transition point at the third part of the duodenum, where the SMA crosses the duodenum. CT has been set up to be largely harmonious for diagnosing gastric and duodenal dilatation **(Fig.4A).**, which may be not well demonstrated by barium studies [20]. Additional value of CT is to show the exact anatomic position of the duodenum and excluding other causes of obstruction [21]. The features described above were present in this case.

Non-invasive, low-cost power Doppler ultrasound can detect reduced AMA and may lead to suspected diagnosis of SMA syndrome indeed in cases where gastroscopy and X-ray with barium do not detect duodenal compression. Ultrasound examination performed in the standing position in addition to the standard supine procedure assessed the AMA indeed in the symptomatic-free period when plain hypotonic duodenography and gastroscopy cannot reveal diagnostic findings [22].

Differential diagnosis includes irritable bowel syndrome, peptic ulcer, pancreatitis, duodenitis, cholelithiasis, neoplastic obstructions, adhesions, visceral neuropathy, porphyria and aneurysm of the abdominal vessels [23].

Treatment is generally conservative, and includes fluid and electrolyte resuscitation, naso-jejunal feeding and mobilisation of the patient into the prone or left lateral decubitus position. Following this, conservative management focuses on nutritional support aimed at restoration of retroperitoneal fat and weight gain [7,21]. If conservative treatment fails, surgical procedures aiming to bypass the obstructed segment may be indicated. Options include duodenojejunostomy, gastrojejunostomy or lysis of the ligament of Treitz with de-rotation of the bowel (Strong's procedure) [7]. Duodenojejunostomy is the surgical option to relieve the obstruction, with good results being reported in 79-100% of the cases [7].

Due to severe malnutrition, our case was initially treated concurrently with both enteral and total parenteral nutrition with the aim of expediting recovery and optimising his nutritional state should he require surgery. An open duodenojejunostomy was performed when symptoms failed to ameliorate after 2 weeks. The patient subsequently made an uneventful recovery.

CONCLUSION

SMA syndrome is a rare condition with non-specific symptoms, and frequently confused with other anatomical or motility-related causes of duodenal obstruction. A history of epigastric pain, post prandial vomiting and rapid weight loss, with pain being relieved in the prone/left lateral decubitus position should raise the clinical suspicion of SMAS. Hence SMAS is clinically a diagnosis of exclusion. Still, radiological examinations, particularly multidetector CT scan can conclusively diagnose this condition grounded on the objective parameters of AMD and AMA. Conservative management with nutritional support is supported for cases who have a short clinical history, and cases with habitual symptoms are more likely to require surgical management if symptoms don't ameliorate with nutritive support. Surgical options for the treatment of SMA syndrome include Strong's procedure, gastrojejunostomy and duodenojejunostomy.

REFERENCES;

1. Madhu B, Govardhan B, Krishna B. Cast syndrome. Oxf Med Case Reports. 2019 Apr 29;2019(4): omz025. doi: 10.1093/omcr/omz025. PMID: 31049210; PMCID: PMC6487991.
2. Warncke ES, Gursahaney DL, Mascolo M, Dee E. Superior mesenteric artery syndrome: a radiographic review. Abdom Radiol (NY). 2019 Sep;44(9):3188-3194. doi: 10.1007/s00261-019-02066-4. PMID: 31127323.
3. Anderson JR, Earnshaw PM, Fraser GM. Extrinsic compression of the third part of the duodenum. Clin Radiol. 1982 Jan;33(1):75-81. doi: 10.1016/s0009-9260(82)80358-9. PMID: 7067340.
4. Rosa-Jiménez F, Rodríguez González FJ, Puente Gutiérrez JJ, Muñoz Sánchez R, Adarraga Cansino MD, Zambrana García JL. Duodenal compression caused by superior mesenteric artery: study of 10 patients. Rev Esp Enferm Dig. 2003 Jul;95(7):485-9, 480-4. English, Spanish. PMID: 12952509.
5. Ahmed AR, Taylor I. Superior mesenteric artery syndrome. Postgrad Med J. 1997 Dec;73(866):776-8. doi: 10.1136/pgmj.73.866.776. PMID: 9497945; PMCID: PMC2431524.

6. Merrett ND, Wilson RB, Cosman P, Biankin AV. Superior mesenteric artery syndrome: diagnosis and treatment strategies. J Gastrointest Surg. 2009 Feb;13(2):287-92. doi: 10.1007/s11605-008-0695-4. Epub 2008 Sep 23. PMID: 18810558.
7. Welsch T, Büchler MW, Kienle P. Recalling superior mesenteric artery syndrome. Dig Surg. 2007;24(3):149-56. doi: 10.1159/000102097. Epub 2007 Apr 27. PMID: 17476104.
8. Lee CS, Mangla JC. Superior mesenteric artery compression syndrome. Am J Gastroenterol. 1978 Aug;70(2):141-50. PMID: 717365.
9. Lee TH, Lee JS, Jo Y, Park KS, Cheon JH, Kim YS, Jang JY, Kang YW. Superior mesenteric artery syndrome: where do we stand today? J Gastrointest Surg. 2012 Dec;16(12):2203-11. doi: 10.1007/s11605-012-2049-5. Epub 2012 Oct 18. PMID: 23076975.
10. Biank V, Werlin S. Superior mesenteric artery syndrome in children: a 20-year experience. J Pediatr Gastroenterol Nutr. 2006 May;42(5):522-5. doi: 10.1097/01.mpg.0000221888. 36501. f2. PMID: 16707974.
11. Ushiki A, Koizumi T, Yamamoto H, Hanaoka M, Kubo K, Matsushita M. Superior mesenteric artery syndrome following initiation of cisplatin-containing chemotherapy: a case report. J Med Case Rep. 2012 Jan 16;6: 14. doi: 10.1186/1752-1947-6-14. PMID: 22248296; PMCID: PMC3275446.
12. Sophia R, Bashir WA. Superior mesenteric artery syndrome. In New Advances in the Basic and Clinical Gastroenterology, T. Brzozowski (ed.). InTech, Croatia, Europe 2012, pp. 415-418.
13. Okamoto T, Takumi Sato, Yukio Sasaki. Superior mesenteric artery syndrome in a healthy active adolescent BMJ Case Rep 2019;12: e228758. doi:10.1136/bcr-2018-228758.
14. Inoue M, Uchida K, Otake K, et al. Development of acute pancreatitis after Nissen fundoplication. Pediatr Int 2015;57: e48–9.
15. Sihuay-Diburga DJ, Accarino-Garaventa A, Vilaseca-Montplet J, et al. Acute pancreatitis and superior mesenteric artery syndrome. Rev Esp Enferm Dig 2013;105: 626–8

16. Van Horne N, Jackson JP. Superior Mesenteric Artery Syndrome. 2022 Jul 18. In: Stat Pearls [Internet]. Treasure Island (FL): Stat Pearls Publishing; 2023 Jan. PMID: 29489172.
17. Cohen LB, Field SP, Sachar DB. The superior mesenteric artery syndrome. The disease that isn't, or is it? Clin Gastroenterol. 1985;7: 113-6.
18. Gustafsson L, Falk A, Lukes PJ, Gamklou R. Diagnosis and treatment of superior mesenteric artery syndrome. Br J Surg. 1984;71: 499-501
19. Konen E, Amitai M, Apter S, et al. CT angiography of superior mesenteric artery syndrome. AJR Am J Roentgenol. 1998;171: 1279-81
20. Birsen Ü, Aykut A, Gökhan K, et al. Superior mesenteric artery syndrome: CT and ultrasonography findings. Diagn Interv Radiol. 2005;11: 90-5.
21. Agrawal GA, Johnson PT, Fishman EK. Multidetector row CT of superior mesenteric artery syndrome. J Clin Gastroenterol 2007;41: 62–5.
22. Neri S, Signorelli SS, Mondati E, Pulvirenti D, Campanile E, Di Pino L, Scuderi M, Giustolisi N, Di Prima P, Mauceri B, Abate G, Cilio D, Misseri M, Scuderi R. Ultrasound imaging in diagnosis of superior mesenteric artery syndrome. J Intern Med. 2005 Apr;257(4):346-51. doi: 10.1111/j.1365-2796.2005. 01456.x. PMID: 15788004.
23. Lippl F, Hannig C, Weiss W, Allescher HD, Classen M, Kurjak M. Superior mesenteric artery syndrome: diagnosis and treatment from the gastroenterologist's view. J Gastroenterol. 2002;37(8):640-3. doi: 10.1007/s005350200101. PMID: 12203080.