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Superior Mesenteric Artery Syndrome in Late Middle-Aged Man: Clinico-Radiological Findings

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

ABSTRACT

Superior mesenteric artery syndrome (SMAS) rarely causes duodenal obstruction by compression of the third part of duodenum between aorta & superior mesenteric artery (SMA). Clinical diagnosis and radiological features play crucial role in the diagnosis of SMAS. Majority of the patients diagnosed with SMAS are managed conservatively and surgery done for cases not responding to medical treatment. Here in we report a male aged 55 years, presenting with acute recurrent abdominal pain associated with vomiting, diagnosed to have SMAS who underwent surgical correction and describe the clinical and radiological findings.

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1. INTRODUCTION

"SMAS, also known as Wilkie syndrome, Cast or Aorto-mesenteric syndrome duodenal compression syndrome, is an acquired vascular compression syndrome in which acute angulation of aorta and SMA, results in compression of the third part of the duodenum leading to intermittent post-prandial pain and signs of duodenal obstruction" [1,2]. "The commonest symptom in SMAS is intermittent epigastric pain and vomiting. Nausea and vomiting may result in anorexia and reduction in body weight. Reduced fat between the aorta SMA, either due to poor nutritional intake or weight reduction results in (AMA) decreased aorto-mesenteric angle and aorto-mesenteric distance (AMD)" [3]. "Prevalence of the SMAS varies from 0.2% to 0.78% in the barium meal studies of the upper gastrointestinal tract" [4-6]. "Manoeuvres similar as lying prone or espousing the left lateral decubitus position may relieve the pain" [7].

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

2. CASE PRESENTATION

An Indian male aged 55 years presented to our hospital with long history of intermittent epigastric pain and vomiting. There was characteristic history of relief of symptoms in prone position. He was previously hospitalized many times for

similar complaints resulting in significant weight loss for the past one year.

On general examination his pulse, blood pressure and respiratory rate were stable. Abdominal examination revealed epigastric tenderness. Cardiac system, respiratory system and neurological examination was normal. Liver function tests and routine blood and urine investigation, were within normal limits. Plain erect chest radiograph was normal. Plain and oral contrast erect abdomen (Fig. 1A, B) showed double bubble sign suggestive of duodenal obstruction.

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

Contrast enhanced computed tomography (CECT) was advised for further evaluation and confirmation. CECT revealed reduced AMA (20°) and short AMD (6.7 mm). Compression was seen at third part of duodenum with dilatation of stomach and proximal duodenum. (Fig. 4A, B).

CECT 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.

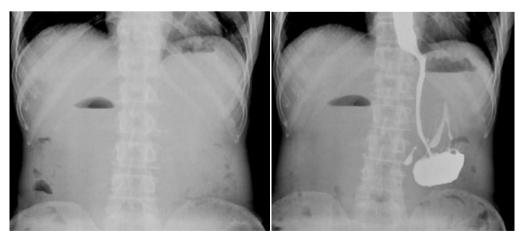


Fig. 1 (A, B). Abdominal plain (A) and contrast (B) erect radiograph demonstrates air fluid levels in left & right hypochondrium(Double bubble sign) suggestive of duodenal obstruction

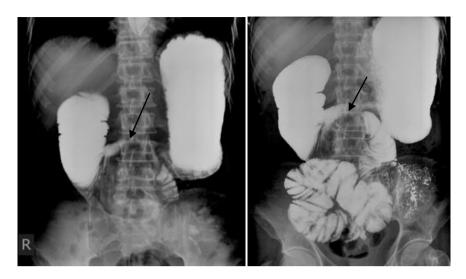


Fig. 2 (A, B). Barium meal study. Supine spot images of study demonstrating dilated stomach & duodenum with curvilinear extrinsic lucency (arrow) corresponding to SMA impression upon the third part of duodenum

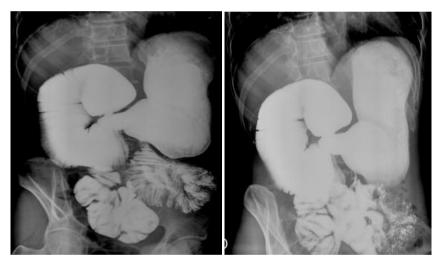


Fig. 3 (A, B). Preoperative Prone barium meal spot images shows dilated stomach, duodenum emptying into the jejunum

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

Diagnosis of SMAS was suggested on the basis of clinical features and radiological findings. Initial conservative approach was tried with nasogastric drainage, fluid electrolyte therapy, and small frequent meals, which did not improve the patient condition. Hence the patient was managed by duodeno-jejunostomy. After surgery our patient is symptom free.

3. DISCUSSION

Foremost depiction of the SMAS was by Carl Freiherr VonRokitansky in 1861 and laterly by

Wilkie in 1927, who handed a more detailed clinical, anatomical and patho-physiological aspect of this syndrome [6]. The frequency of SMAS grounded on fluoroscopic examination is estimated to be 0.013–0.3% [8], whereas other sources quote a frequency of 0.0965% in a chronic hospital setting versus 0.00108–0.0052% in an acute general hospital setting [9]. "There is a womanish preponderance (64–66%), with 75% of cases being in individualities in age range from 10 to 39 years" [10,11].

"SMA leaves the aorta at an acute downward angle in humans. This results in the conformation of vascular angle between SMA and aorta, through which the third part of duodenum traverses" [6]. "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" [10]. 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 [1,6] Cut-off values were reported to be 22° for AMA (42% sensitivity and 100% specificity) and 8 mm for AMD (100% sensitivity and specificity) [12]. In this case, CT dimension of AMA and AMD is lower than cut off value.

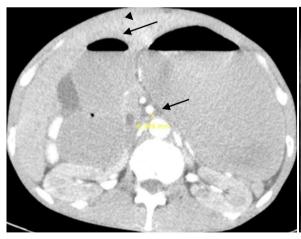
"The clinical presentation may be acute or chronic. The classical symptoms of SMAS are postprandial or intermittent abdominal pain and features of small bowel obstruction such as nausea, vomiting and anorexia" [11]. "The patient symptoms are relieved in left lateral decubitus, prone or knee to chest position" [13]. Analogous common symptoms and relieving factor was present in this case.

Causes of SMAS are multiple which include anorexia nervosa, severe cachexia or catabolic state due to neoplasm, severe head injury or burn, bariatric surgery, prolonged bed rest, high insertion of ligament of Treitz, low origin of SMA, Nissan fundoplication, spinal deformity, spinal scoliosis surgery, intestinal mal-rotation, peritoneal adhesions, and elderly patient with heavily calcified or tortuous aorta [13]. "Patients are often underweight, with a reported median BMI of 17.4 at diagnosis" [10].

Weight loss is not necessary for the development of the SMAS in the paediatric population but pubertal growth spurt is a threat factor for SMAS [14]. In adolescents whose visceral adipose tissue has formerly been depleted, acute and inordinate ingestion of water and food can spark a manifestation of SMAS. In adolescents with intermittent abdominal symptoms, it is important to consider SMAS in the differential diagnosis [14]. The association of acute pancreatitis with SMAS infrequently reported [15,16]. 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 pancreatitis [16].

"Symptoms caused from duodenal obstruction include abdominal pain, vomiting, and weight loss. All other causes of duodenal obstruction, such as benign or malignant tumours, and extrinsic causes similar as pancreatitis, mesenteric mass, or aortic aneurysm must be ruled out with routine imaging modalities before a opinion of SMAS is confirmed" [7]. Elderly cachexic patient with decreased retroperitoneal fat and tortuous abdominal aorta was the cause of SMAS in this case.

Fluoroscopic studies of upper gastrointestinal tract can demonstrate extrinsic compression of the third part of the duodenum, dilatation of the first and second part of the duodenum (Fig.1A,B) and a fairly collapsed small bowel distal to point where the SMA crosses the duodenum [17].



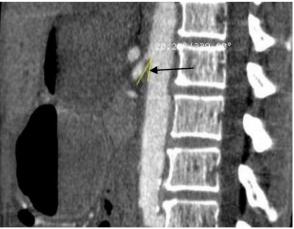


Fig. 4 (A, B). CT criteria suggestive of SMAS. Selected contrast enhanced mid abdominal axial (A) and mid sagittal (B) images demonstrating reduced AMD (small vertical line in region of interest in image A), dilated proximal second part of duodenum (arrow in image A), and reduced AMA (arrow in image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

Conventional Barium meal studies and Hypotonic duodenography are crucial 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 [4,18]. Barium meal examination shows proximal gastric duodenal dilatation with perpendicular band of external compression at the third part of the duodenum (Fig. 2A,B). Other findings on barium meal study include reverse barium flow or antiperistaltic flow of barium proximal to the site of obstruction and relief of obstruction when the patient is in a left lateral decubitus or prone position [8] (Fig. 3A,B).

In the history, Angiographic dimension of the AMA was recognised as being a more accurate investigation than routine barium meal studies [19]. Still, the procedure is invasive, and some patients are unable to tolerate it.

Lately, Konen et al have noted the advantage of 3D CT angiographic reconstruction, which helps to exclude the incorrect diagnoses that appear from the angulations of the SMA [20]. 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 AMA and AMD are reduced, with values of 6-22 degrees and 2-8 mm, respectively [20]. 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 [21]. CT is of additional value in demonstrating the exact anatomic position of the duodenum excluding other causes of duodenal obstruction [22]. The features described above were present in this case.

Low-cost, non-invasive, power Doppler ultrasound can detect reduced AMA and may lead to suspected diagnosis of SMAS, indeed in cases where gastroscopy and barium meal study do not detect duodenal compression. Ultrasound examination performed in the standard supine position in addition to the standing position assessed the AMA, indeed in the symptomatic-

free period, when gastroscopy and plain hypotonic duodenography cannot reveal diagnostic findings [23].

Differential diagnosis includes peptic ulcer, duodenitis, pancreatitis, cholelithiasis, neoplastic obstructions, adhesions, irritable bowel syndrome, visceral neuropathy, porphyria and aneurysm of the abdominal vessels [24].

Treatment is generally conservative, includes fluid and electrolyte correction, nasojejunal feeding and mobilisation of the patient into the prone or left lateral decubitus position. Conservative management focuses on nutritional support aimed at restoration of retroperitoneal fat and weight gain [8,22]. If conservative treatment fails, surgical procedures aiming to bypass the obstructed segment may be indicated. Options include duodeno-jejunostomy, gastroieiunostomy or lysis of the ligament of Treitz with de-rotation of the bowel (Strong's procedure) [8]. Duodeno-jejunostomy is the surgical method of choice to relieve the obstruction, with good results being reported in 79-100% of the cases [8].

Bloomberg et al. [25] reported SMAS in a 18 years old female who presented with abdominal pain, distension and vomiting due to anorexia nervosa with narrow AMA and compression of duodenum.

Berken Jonathan A et al. [26] diagnosed SMAS in a 17 year old adolescent due to precipitous cannibinoid weight loss resulting from hyperemesis syndrome. Alvina Karam et al. [27] reported SMAS in a 20 year old malnourished female with nausea, vomiting, weight loss and abdominal pain who improved with conservative treatment. Kimura A et al. [28] diagnosed SMAS in a 64 year old man with appetite loss and vomiting who underwent laparoscopic gastroieiunostomy and percutaneous endoscopic gastrotomy with no recurrence of symptoms on follow up.

The clinical features and radiological findings in our case were similar to the cases reported in the literature [25-28]. Due to severe malnutrition, our case was initially treated with both enteral and total parenteral nutrition with the aim of speeding recovery and optimising his nutritional state should he require surgery. When symptoms failed to ameliorate after 2 weeks, an open duodeno-jejunostomy was performed. The

patient subsequently made an uneventful recovery.

4. CONCLUSION

SMAS is a rare disease with non-specific symptoms, and frequently mistaken with other anatomical or motility-related causes of duodenal obstruction. A history of post prandial vomiting, epigastric pain, and rapid weight reduction with pain being relieved in the prone or left lateral decubitus position should raise the clinical suspicion of SMAS. Hence SMAS is clinically a exclusion. diagnosis of Still. radiological examinations, particularly multidetector CT scan conclusively diagnose this condition grounded on the objective parameters of AMD and AMA. For cases who have a short clinical conservative management nutritional support is recommended and cases with habitual symptoms are more likely to require management if symptoms ameliorate with nutritive support.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard guideline participant consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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