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

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 painassociated with vomiting, diagnosed to have SMAS who underwent surgical correction and describe the clinical and radiological findings.

Keywords

Aorto-mesenteric angle, Aorto-mesenteric distance,Abdominal vascular compression syndrome, Cast syndrome, Superior mesenteric artery syndrome,Wilkie syndrome.

INTRODUCTION

“SMAS, also known as Wilkie syndrome, Cast syndrome or Aorto-mesenteric duodenal compression syndrome, is an acquired vascular compression syndrome in which acute angulation of aortaandSMA, results in compression of the third part of the duodenum leading to intermittent post-prandial painand signs of duodenal obstruction”[1,28]. “The commonestsymptom 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 decreased aorto-mesenteric angle(AMA) and aorto-mesenteric distance(AMD)” [2]. “Prevalence of the SMAS varies from 0.2% to 0.78% in the barium meal studies of the upper gastrointestinal

tract" [3-5]. "Manoeuvres similar as lying prone or espousing the left lateral decubitus position may relieve the pain" [6].

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.

CASE PRESENTATION

An Indian male aged 55 years presented to our hospital with long history of intermittentepigastricpain and vomiting. There was characteristichistory 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 (**Figure.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**(Figure.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. **(Figure.4A,B)**.

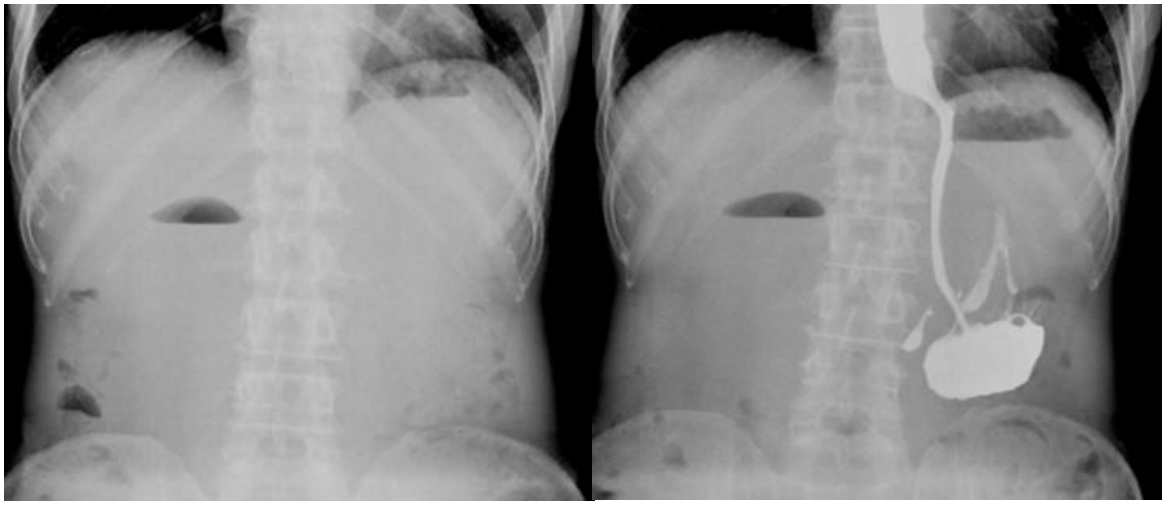

Fig.1(A, B). Abdominal plain (A) and contrast (B) erect radiograph demonstrates air fluid levels in left & right hypochondrium(Double bubble sign) suggestive of duodenal obstruction.

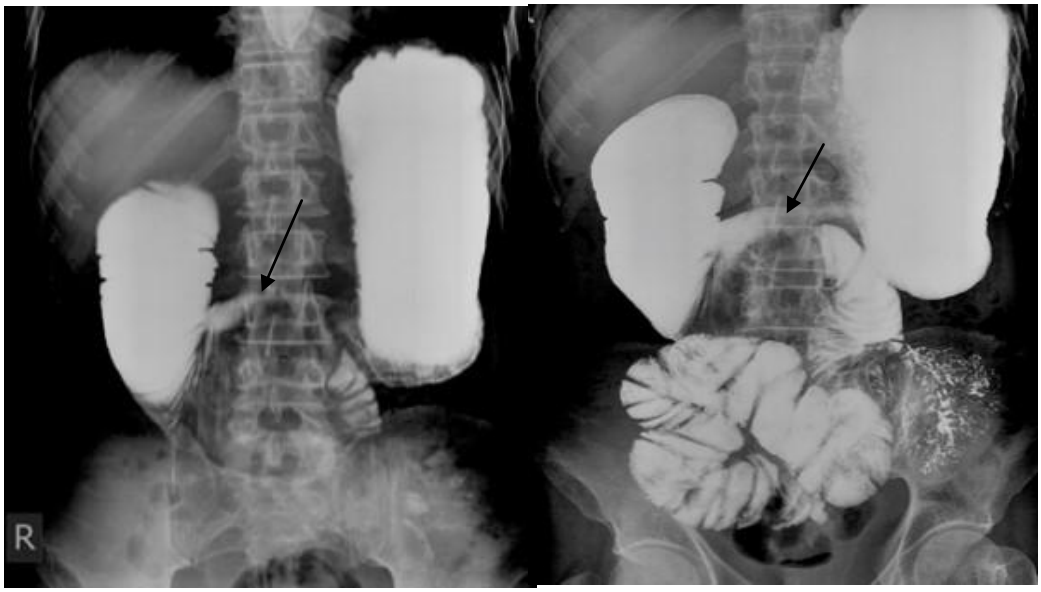

Fig.2(A,B).Barium meal study. Supine spot images of study demonstrating dilated stomach & duodenum with curvilinear extrinsic lucency(arrow) corresponding to SMA impression upon the third part of duodenum.

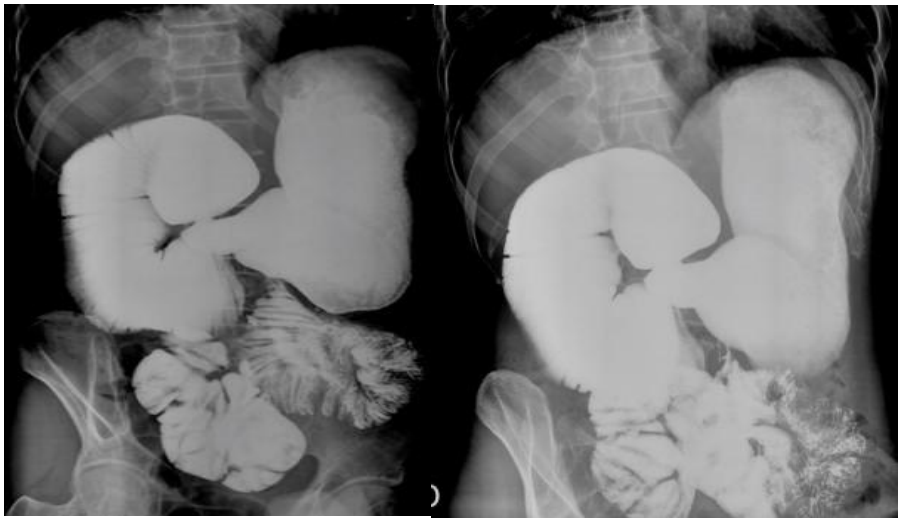

Fig.3(A,B).Preoperative Prone Barium meal spot imagesshows dilated stomach, duodenum emptying into the jejunum.

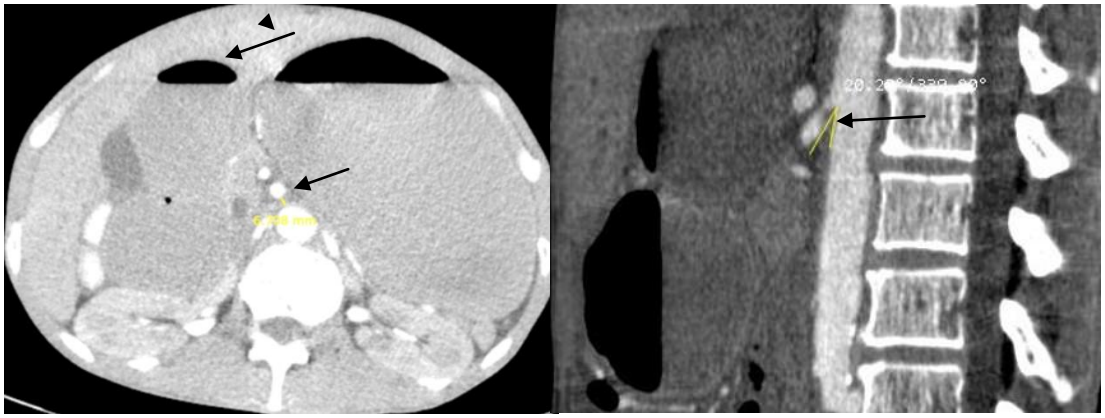

Fig.4(A,B).CTcriteria suggestive of SMAS. Selected contrast enhanced mid abdominal axial (A) and mid sagittal (B) images demonstrating reduced AMD (small vertical line in region of interest in image A), dilated proximal second part of duodenum (arrow in image A), and reduced AMA (arrow in image B).

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.

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

Diagnosis of SMAS was suggestedon 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.

DISCUSSION

Foremost depiction of the SMAS was by Carl FreiherrVonRokitansky in 1861 and laterly by Wilkie in 1927, who handed a more detailed clinical, anatomical and patho-physiological aspect of this syndrome [5]. The frequency of SMASgrounded on fluoroscopicexamination is estimated to be 0.013–0.3% [7], 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 [8]. "There is a womanish preponderance (64–66%), with 75% of cases beingin individualitiesin age range from 10 to 39 years" [9, 10].

"SMA leaves the aorta at an acute downward anglein humans. This results in the conformation of vascular angle between SMA and aorta,through which the third part of duodenum traverses"[5]. "The duodenum is typicallygirdled 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 AMAis 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 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" [10]."The patient symptoms are relieved in left lateral decubitus, prone or knee to chest position" [12].Analogous common symptoms and relieving factor was present in this case.

Causes of SMASare multiple which includeanorexia 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[12]. “Patients are often underweight, with a reported median BMI of 17.4 at diagnosis”[9].

Weight loss is not necessary for the development of the SMAS in the paediatric populationbut pubertal growth spurt is a threat factor for SMAS [13]. 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 SMASin the differential diagnosis [13].The association ofacute pancreatitis with SMAS 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 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 aopinion of SMAS is confirmed” [6].Elderly cachexicpatient 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 [16].

Conventional Barium meal studies and Hypotonicduodenography 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 [3,17].Barium meal examination shows proximal gastric and duodenal dilatation with perpendicular band of external compression at the third part of the duodenum **(Figure 2A,B).** Other findings on barium meal study 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 left lateral decubitus or prone position [7]**(Fig.3A,B)**.

In the history, Angiographic dimension of the AMA was recognised as being a more accurate investigation than routine barium meal studies [18].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

[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 AMA and AMD 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 largelyharmonious for diagnosing gastric and duodenal dilatation**(Fig.4A)**, which may be not well demonstrated by barium studies [20]. CT is of additional value in demonstrating the exact anatomic position of the duodenum and excluding other causes of duodenal obstruction [21].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 thestandard supine position in addition to thestanding position assessed the AMA,indeed in the symptomatic-free period, when gastroscopy and plain hypotonic duodenographycannot reveal diagnostic findings [22].

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

Treatment is generally conservative, and includes fluid and electrolyte correction, naso-jejunal 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 [7,21]. If conservative treatment fails, surgical procedures aiming to bypass the obstructed segment may be indicated. Options include duodeno-jejunostomy, gastro-jejunostomy or lysis of the ligament of Treitz with de-rotation of the bowel (Strong's procedure)[7].Duodeno-jejunostomy is the surgical method of choice to relieve the obstruction, with good results being reported in 79-100% of the cases [7].

Bloomberg et al [24] 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 [25] diagnosed SMAS in a 17 year old adolescent due to precipitous weight loss resulting from cannibinoid hyperemesis syndrome.AlvinaKaram et al [26] reported SMAS in a 20 year old malnourished female with nausea, vomiting, weight loss and

abdominal pain who improved with conservative treatment.Kimura Aet al [27] diagnosed SMAS in a 64 year old man with appetite loss and vomiting who underwent laparoscopic gastro-jejunostomy and percutaneous endoscopic gastrotomy with no recurrence of symptoms on follow up.

The clinical features and radiological findings in ourcase were similar to the cases reported in the literature[24-27].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 2weeks,an open duodeno-jejunostomy was performed. The patient subsequently made an uneventful recovery.

CONCLUSION

SMAS is a rare disease with non-specific symptoms, and frequentlymistaken 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 SMASis 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. For cases who have a short clinical history, conservative management with nutritional support is recommendedand cases with habitual symptoms are more likely to require surgical management if symptoms don’tameliorate 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.

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