ABSTRACT: Superior mesenteric artery (SMA) syndrome is a rare acquired disorder in which an acute angulation of SMA causes compression and obstruction of the third part of the duodenum between the SMA and the aorta. The diagnosis is usually made by exclusion. A large number of debilitating conditions leading to a loss of fatty tissue such as anorexia nervosa, malabsorption, or hyper catabolic states like burns, major surgery, severe injuries, or malignancies can cause this syndrome. To our knowledge there are no cases reported in the Indian literature, though many such cases are being diagnosed and successfully treated the world over. We recently diagnosed and successfully treated a patient of S.M.A. syndrome in Shri Aurobindo Institute of Medical Sciences, Medical College and Post-Graduate Institute. We take the opportunity to review the literature on S.M.A. syndrome.

KEYWORDS: Duodenal obstruction, SMA syndrome.

S.M.A.S. – A CASE REPORT: A 60 years old male farmer was admitted from medical O.P.D. with two weeks complaints of recurrent bilious vomiting, 2-3 hours after meals and abdominal distension. The patient was passing hard stools every second or third day. He reported significant loss of weight over last month. There were no other complaints. He was addicted to tobacco chewing and occasionally consumes alcohol.

Except for an emaciated and dehydrated look there was no abnormality on general and systemic examination. His hemoglobin was 13.3gm% with normal hematological picture; blood urea 105mg%, serum creatinine 3.88 mg%; serum proteins 4.46 gm.% with albumin 1.9 gm.%, globulin 2.56gm% (A/G ratio=0.7); serum bilirubin 0.93mg%, S.G.O.T. 48 units, S.G.P.T.46 units and I.N.R. was 1.1.

His barium study showed massive dilatation of the stomach which reached up to the pelvic brim as well as dilatation of 1<sup>st</sup>, 2nd and 3<sup>rd</sup> parts of duodenum.

His abdominal sonography revealed dilated stomach and 1<sup>st</sup>-3<sup>rd</sup> parts of duodenum with the superior mesenteric artery deviated to the left and the aorto-mesenteric distance of 12 mm.
Upper G.I endoscopy showed grade B esophagitis, small hiatus hernia and mucosal fold thickening and narrowed lumen of distal duodenum, but no intraluminal growth or stricture was seen.

A contrast enhanced C.T. scan of the abdomen (rectal and oral iodinated contrast & venous contrast) showed dilated 1st -3rd part of duodenum with aortic mesenteric angle of 23 degree and aorta–mesenteric artery distance of 12 mm. A 1.6x1.4 cm hemangioma in the posterosuperior segment of right lobe of liver was incidentally noted.
The patient improved with nasogastric suction and intravenous fluids; his blood urea came to 53mg% and serum creatinine 1.8 mg%. He however denied surgery and left against medical advice, only to return after 10 days with worsening of obstructive symptoms. After initial conservative treatment with nasogastric suction and intravenous alimentation he was subjected to surgery. The ligament of Treitz was cut, third part of duodenum was found to be compressed by the S.M.A., and side to side duodenojejunostomy was done between the 3rd part of duodenum and jejunum at 30 cm from duodenojejunal junction.

Apost-operative Gastrograffin study showed a well-functioning anastomosis.

The patient’s recovery was complicated by abdominal sepsis and hypoproteinemia but after three weeks the patient was discharged from the hospital with no symptoms of duodenal obstruction.

**REVIEW OF LITERATURE:** SMA syndrome (also known as Willkie’s syndrome, gastro-vascular syndrome, cast syndrome (due to immobilization in a body cast), chronic duodenal ileus, intermittent arterio-mesenteric occlusion or AO syndrome) was first described by Von Rokitanski by autopsy in 1861. In 1908, Laffer presented one of the first review of the condition. Wilkie in 1927 reported a detailed clinical and pathophysiologic study in a series of 64 patients and suggested treatment approaches.

Skepticism about the existence of the condition continued until the 1960s till new radiologic techniques provided adequate evidence to support the existence of SMAS. Some still question whether it is a true entity and others believe that the syndrome is over diagnosed. In the recent years the reported incidence has been on the rise, possibly due to increased physician awareness.

The defining feature of this entity is upper gastrointestinal obstruction caused by compression of the third part of the duodenum between the SMA anteriorly and the aorta posteriorly.

**Origin and anatomical relations of superior mesenteric artery:** SMA is a direct branch of abdominal aorta distal to the coeliac trunk and passes anterior to the third part of the duodenum before supplying branches to various parts of the gut. Normally, fat and lymphatic tissues around the SMA provide protection to the duodenum against compression by the artery.
Normally, the aortomesenteric angle and aortomesenteric distance is 25° to 60° and 10 to 28 mm respectively. In SMA syndrome, usually associated with conditions of severe weight loss, the fat cushion around the SMA is diminished, hereby increasing the acuteness of the aortomesenteric angle to 6° to 15° and reducing the distance between the two to 2 to 8 mm.6

Many causes for SMAS have been suggested. Conditions like increased spinal lordosis, application of a body cast, short ligament of Treitz, a high insertion of the duodenum at the ligament of Treitz, a congenital low origin of the superior mesenteric artery and compression of the duodenum caused by peritoneal adhesions as a result of duodenal mal-rotation may also precipitate this syndrome. Rarely, SMA syndrome has been reported after abdominal surgery.7-15

Clinical presentation: Patients usually have chronic insidious symptomatology but may present with an acute exacerbation of chronic symptoms. The symptoms are usually long-standing and vague, with early satiety and anorexia and recurrent episodes of abdominal pain, often associated with bilious vomiting. Less often there is an acute presentation characterized by signs and symptoms of
duodenal obstruction and rarely "stabbing" postprandial abdominal pain due to both the duodenal compression and the compensatory reversed peristalsis. Weight loss is common because patients often regurgitate their food or become afraid to eat. Weight loss can in turn exacerbate the condition.

Symptoms can be relieved by postural changes in the prone or knee-chest position, suggesting vascular compression. However relief with postural change is inconsistent, its absence does not rule out vascular compressive syndromes. Moreover, peptic ulcer disease coexists with SMAS in up to 25% of cases.

Whether duodenal obstruction precedes the peptic ulcer or the peptic disease contributes to the arteriomesenteric duodenal compression is uncertain.

**Diagnosis:** The diagnosis of SMA syndrome is based on clinical symptoms and radiologic evidence of duodenal obstruction.

Plain radiograph demonstrates a dilated, fluid- and gas-filled stomach. Barium radiography shows dilatation of the first and second part of the duodenum, extrinsic compression of the third part, and a collapsed small bowel distal to the crossing of the SMA.

Contrast-enhanced CT scan or magnetic resonance angiography (MRA) enables visualization of vascular compression of the duodenum and precise measurement of aortomesenteric distance. Both these procedures are noninvasive and reportedly equivalent to conventional contrast angiography, which has previously been suggested as the reference standard for establishing the diagnosis.

Endoscopic examination may visualize a pulsatile extrinsic compression, suggestive of this condition.

**Treatment:** Traditionally, treatment has consisted of conservative measures such as nasogastric decompression and hyper alimentation followed by oral feeding with frequent small meals. Posturing maneuvers lying in the left lateral decubitus position during meals and motility agents may be helpful
in some patients. Medical treatment is usually successful in patients with a short history and a relatively minor degree of duodenal stasis shown radiologically.\textsuperscript{23}

Surgery is to be considered if conservative treatment fails along with a long history of indigestion, progressive weight loss, pronounced dilatation and stasis of the duodenum.\textsuperscript{24, 25} Duodenojejunostomy is effective in the majority of patients.\textsuperscript{4} A 7-year follow-up study of 16 patients treated with duodenojejunostomy found that outcome was regarded as excellent by 3 patients, good by 6, satisfactory by 5, and poor by 2 patients.\textsuperscript{26} Although effective, conventional surgery may not be suitable for debilitated patients. Laparoscopic minimally invasive duodenojejunostomy has been used since 1998.\textsuperscript{27, 28} Laparoscopic lysis of the ligament of Treitz with mobilization of the duodenum is another minimally invasive approach.

Hutchinson and Bassett and Munns and colleagues both reported 100\% success rates without any surgical intervention.\textsuperscript{5, 29} However; both these series were in orthopedic patients who had an acute postoperative episode. Biopsy specimens of the duodenum and jejunum should be obtained before surgery to exclude infiltrative, infective, neurologic or malignant causes for the duodenal dilatation and chronic idiopathic intestinal pseudo-obstruction.\textsuperscript{17, 30, 31} Duodenojejunostomy from the second portion of the duodenum to the jejunum has been considered the surgical treatment of choice in many surgical publications.\textsuperscript{22, 23, 32, 33} Other procedures include gastrojejunostomy, Roux-en-Y duodenojejunostomy and anterior transposition of the third part of the duodenum—the last technique being an effective and permanent direct approach.\textsuperscript{34} Lysis of the ligament of Treitz is specially recommended in children as it does not include any intestinal anastomosis.\textsuperscript{16, 35 and 36}

CONCLUSION: With the advent of newer imaging techniques Superior mesenteric artery syndrome has been clearly defined. As it is frequently associated with a wide range of predisposing conditions, clinicians must consider this syndrome in such a setting. More number of patients is likely to be diagnosed in future with better awareness of the disease. Conventional surgical procedures have resulted in good results but may be associated with operative risk in very debilitated patients. Minimally invasive laparoscopic surgical treatment may replace the conventional operative procedures in future.

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