

## Superior Mesenteric Syndrome –Acute On Chronic Presentation and A Review

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### Case Summary:

A 24-year-old Caucasian male presented with low GCS, seizures, severe vomiting and hypokalaemia initially admitted under the physicians through the Emergency Department and later shifted to the Intensive care unit (ICU). The initial diagnosis was Acute Kidney injury with metabolic hypochloreaemia and hypokalaemic alkalosis with a background of Chronic Kidney disease treated under the Nephrologist in the past. In ICU, Patient was intubated and resuscitated with intravenous fluid resuscitation and correction of electrolyte abnormalities. The patient also had aspiration pneumonia in ICU and was treated with intravenous antibiotics Total parental nutrition (TPN) for nutritional support and Pabrinex due to the risk of refeeding syndrome. The patient also developed swelling right arm and an Ultrasound of the right Arm showed right-sided jugular vein thrombosis and was treated with anticoagulants.

He had a CT (Computer Tomography) scan of the head for low GCS, which did not reveal any abnormality, Lumbar puncture and CSF analysis were normal. CT scan of the abdomen and pelvis with contrast revealed a diagnosis of superior mesenteric artery syndrome. He responded well to conservative treatment in ICU and was discharged with a referral to a tertiary center for further management.

Past history included multiple admissions over six years for vomiting and pyelonephritis. Oesophago-gastro duodenoscopy (OGD) showed gastric stasis with ulcers in the gastric antrum and duodenum. Barium meal showed a failure to progress beyond Duodenum and confirmed gastric outlet obstruction. He had lost about 2 stones over the last couple of years and was on proton pump inhibitors and Anti-emetics.

Platelets	521	Neutrophils	16.6	Urea	39.5	AKI status	3	GGT	38
RBC	6.01	Lymphocytes	1.0	Calcium	2.81	GFR	11	ALP	144
Haemoglobin	177	Monocytes	1.7	Adjusted Calcium	2.45	PT	1.2	Total Bilirubin	25
Haematocrit	0.5	Eosinophils	0.0	Phosphate	1.90				
MCV	83.4	Basophils	0.1	Total Protein	93	APTT	31.9		
MCH	29.4	CRP	72	Albumin	58	Fibrinogen	3.94		
MCHC	352	Na	126	Globulin	35	Chloride	60		
RDW	13.6	K+	3.7	Magnesium	1.06	ALT	22		
WCC	19.60	Creatinine	586	TSH	2.12	AST	35		

**Table 1:** Blood results on admission.

### Radiological findings:

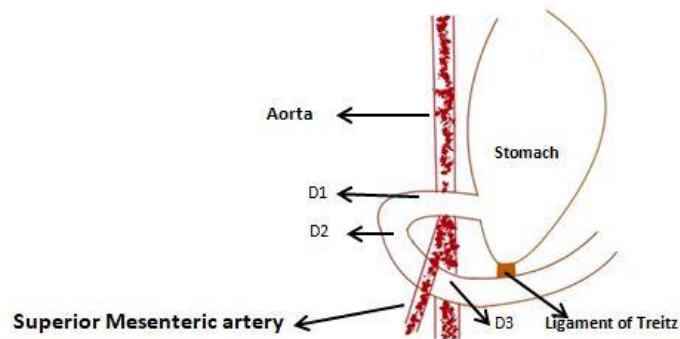
The images from the CT abdomen showed signs of SMA syndrome are as shown below.



**Figure 1:** Sagittal reformatted CT image demonstrating an Aorto-SMA angle of 11.7 degrees (18-70 degrees)

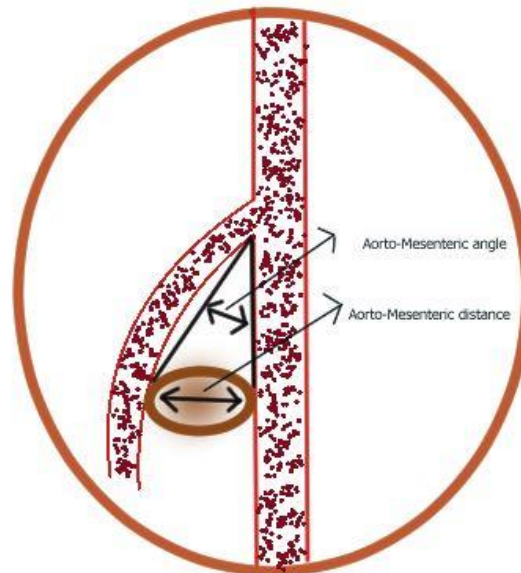


**Figure 2:** Sagittal reformatted CT image demonstrating an Aorto-SMA distance of 5 mm (10-28 mm).



**Figure 1:** Anatomy of superior mesenteric artery syndrome

D1 - First part of the duodenum, D2 - Second part of duodenum & D3 - Third part of duodenum.



**Figure 2:** Compression of the third part of duodenum by SMA. Aorto–Mesenteric angle (AMA) and Aorto-Mesenteric distance (AMD) are as shown above.

### Etiology and Pathophysiology aspect of the disease:

The SMA originates at the level of the third lumbar vertebrae and forms an angle as it takes an anterior and caudal course from its origin of abdominal aorta. The space between the aorta and SMA is occupied by fat and lymphatic tissue and the loss of these tissue following certain conditions leads to compression of the relatively fixed third part of the duodenum leading to SMA syndrome [3,4]. Studies have also reported SMA syndrome is also caused by a short Treitz's ligament and low origin of SMA which caused a decline in the angle formed by the aorta and the SMA [5].The mean angle formed between the SMA and aorta varied between 38°,41.25° and 56° from studies in the literature<sup>4</sup>.The range of

angles varied between 20° to 70° [6-8] and the mean aorto-mesenteric distance between 10–28 mm [9]. Patients affected by SMA syndrome have an angle of < 25° and an aorto-mesenteric distance < 8-10 mm [9]. Our patient had an Aorto-SMA angle of 11.7 degrees and Aorto-SMA distance of 5 mm.

Ahmed et al (1997) [5] categorized the etiology of SMA syndrome into factors that predispose and those precipitate the condition as depicted below in table 2:

<b>Predisposing factors</b>
<ul style="list-style-type: none"> <li>Abnormal acute aortomesenteric vascular angle with a short aorta-SMA gap</li> </ul>
<ul style="list-style-type: none"> <li>Abnormal high fixation of duodenojejunal flexure to the ligament of treitz</li> </ul>
<ul style="list-style-type: none"> <li>Exaggerated lumbar lordosis</li> </ul>
<ul style="list-style-type: none"> <li>Unusually low origin of superior mesenteric artery</li> </ul>
<ul style="list-style-type: none"> <li>Supine position</li> </ul>
<b>Precipitating factors</b>
<ul style="list-style-type: none"> <li>Marked weight loss with subsequent loss of mesenteric and retroperitoneal fat may occur in severe wasting diseases like cancer, Burns, injuries like trauma to head and spine, Dietary disorders like anorexia nervosa and malabsorption, Post-operative state and deformity.</li> </ul>

**Table 2:** Adapted from Ahmed et al [5] showing predisposing and precipitating factors.

Ahmed et al proposed, that humans due to the erect posture naturally have a downward angle of the origin of SMA from aorta which predisposes them to SMA syndrome, unlike the quadrupeds having a right angle origin. The study logically argued about duodenum suspended by the ligament of treitz (LOT) differs in the length between different individuals and people with short LOT are at higher risk of developing SMA syndrome due to duodenum moved higher into the narrow area of the aorto-mesenteric angle. The study mentions about the hypothetical possibility of a lower position of the duodenum does not offer any advantage due to the aorta being pushed forwards as the spine curves anteriorly at the level of L4 vertebrae which will reduce the distance between aorta and SMA [5].

An initial study by Hines et al (1984) [4] and review update by Welsh et al (2007) [10] used the word predisposing factors rather than dividing into predisposing and precipitating factors by Ahmed et al. In addition to the factors listed in table 2. The studies mentioned the following factors predisposing SMA syndrome. 1) Chronic wasting disease like cancer, cerebral palsy, Paraplegia, cardiac cachexia and drug abuse 2) Trauma due to injuries like burns, Brain injury and multiple trauma. 3) A dietary disorder like anorexia nervosa and malabsorption 4) Post-operative states after bariatric surgery, ileoanal pouch anastomosis, Nissens fundoplication, aortic aneurysm repair, scoliosis surgery 5) peritoneal adhesions, Neoplasia of mesenteric root and dissecting aortic aneurysms as possible causes [10]. SMA has been reported in conditions like HIV and Methamphetamine abuse [11] in recent studies.

### Clinical presentation and management:

The clinical symptoms can present acutely or chronic with gradual onset of symptoms depending on the grade of obstruction. The diagnosis can be easily confused with SMA-like syndrome due to decreased duodenal peristalsis in conditions like pancreatitis, diabetes, dermatomyositis, Lupus erythematosus, Myxoedema, Amyloidosis and Myotonic dystrophy often procrastinating the diagnosis leading to deterioration in an already deprived nutritional status due to weight loss [5,12]. Patients presenting acutely have symptoms of duodenal obstruction like pain abdomen, vomiting, distension, abnormal bowel sounds with electrolyte abnormalities, which were aggravated by eating and relieved by adopting knee-chest and prone position as it is known to increase the angle between the aorta and SMA [13]. The subset of the population with chronic presentation shows the persistence of intermittent abdominal pain associated with anorexia and early satiety [5]. The patient above serves as an example of how the diagnosis is difficult and challenging and often doctors are misled. Having a high index of suspicion of SMA syndrome

is always helpful in treating the patients optimally and even helps further if a surgical option is deemed necessary.

Lee et al (2012) study of a large case series of 80 patients described the patients to be mostly females (53/80) with a median age of presentation at 28 years. The median body mass index at diagnosis was 17.4 kg/m<sup>2</sup>, with a range of 10 to 22.1 kg/m<sup>2</sup>. About 50 % of the patients in the study were associated with mental and behavioral disorders (21.3%), infectious disorders (12.5%) and disorders of the nervous system (11.3 %). The symptoms noted were vomiting (70%), Nausea (66.3%), Abdominal pain (65%), Anorexia (33.8%), and Postprandial fullness (33.8%) [14].

Biank et al (2006) case series of twenty-two pediatric patients found a majority of patients were female (64%) with presenting symptoms of abdominal pain (59%), vomiting (50%), nausea (40%), early satiety (32%) and anorexia (18%). SMA syndrome in children most often present acutely rather than chronic with no weight loss as a predisposing factor and medical management being more successful [15].

The investigations available for diagnosis include X-ray, Barium swallow, Ultrasound Doppler, Computer tomography (CT), Magnetic resonance imaging (MRI) angiography and Endoscopy for diagnosis. Plain X-ray might show normal or dilated stomach with diminished bowel gas findings [16]. Ultrasound Doppler can be used to measure the mesenteric angle after overnight fasting during normal expiration at about 2 cms below the origin of the SMA in right lateral, standing and supine position and proved to be effective tool and could be used in epidemiological screening to diagnose suspected cases of SMA syndrome [9]. But, Ultrasound can be operator dependent and might not be suitable to be done as emergency settings.

Barium swallow might show proximal duodenum dilatation, but also the classic abrupt termination of the swallow in the third part of the duodenum with obstruction, which is relieved when the patient takes a left lateral decubitus position [4]. Radiologic criteria for the diagnosis of SMA syndrome comprises of 1) Dilatation of the first and second parts of the duodenum, with or without gastric dilatation, 2) Abrupt vertical and oblique compression of the mucosal folds, 3) Anti-peristaltic flow of contrast medium proximal to the obstruction, (iv) delay in transit of 4–6 h through the gastroduodenal region, and 4) relief of obstruction in a prone, knee-chest or left lateral decubitus position [4,17]. CT scan of the abdomen with contrast helps not only diagnosing SMA syndrome but also to exclude the differential diagnosis. CT scan can determine the aorto-mesenteric angle, distance, the grade of duodenal compression, the extent of the loss of adipose and lymphatic tissue at the AM angle, Aneurysms, dilatation of left renal vein, Neoplasia and lastly can be used for planning

before surgery [10,18-20]. Endoscopy might reveal distended stomach and proximal duodenum and also rule out gastric or duodenal ulcer disease, intestinal intraluminal obstruction and conditions mimicking SMA syndrome [10].

Conservative management is the preferred line of management of patients with SMA syndrome and preferably involve a multi-disciplinary team approach of working closely with gastroenterologists, radiologist, Dieticians and surgeons with an objective of improving the nutritional status and weight gain, which in turn can increase the adipose tissue and increase the aorto-mesenteric angle for alleviation of symptoms. The conservative approach encompasses measures like Nasogastric tube insertion for decompression of the stomach and the duodenum, Feeding through nasojejunal tube or total parenteral nutrition (TPN), correction of electrolyte abnormalities, Appetite stimulations, Anti-emetics, prokinetic agents and postural therapy like left-lateral or prone position, when the patient is able to tolerate oral feeds which can relieve the symptoms if the patients have oral feeding [3,10,18,21]. The conservative approach has an estimated success rate of 85% and also works in patients with symptoms less than a month [15]. Patients who fail with the conservative approach are considered for surgery. Surgical options include Strong's procedure (a division of the ligament of Treitz and positioning of the duodenum to the right side of SMA preventing compression) [22]. The procedure has fallen out of choice as the procedure aggravated the symptoms and technically difficult or impossible to mobilize duodenum due to adhesions and the interference of short vessels from the inferior pancreaticoduodenal artery [7,23]. Few studies have reported Gastrojejunostomy can be performed, if severe gastric distention is present with good results [24]. However, Lee et al (1978) study showed, in spite of having Gastrojejunostomy, which provided adequate decompression of the stomach. The procedure failed in relieving duodenal obstruction [25]. The standard practice presently followed is laparoscopic duodenojejunostomy with a success rate of 80-100% with added benefits of minimal blood loss, decreased post-operative pain, better cosmetic outcome, early recovery period, faster discharge rate [11,26,27].

### Conclusion:

SMA syndrome is one of the rare disease presenting with symptoms which are atypical, insidious in onset and pose a diagnostic dilemma unless the doctors have a high degree of suspicion in a population presenting with weight loss, Nonspecific abdominal pain and early satiety. Prompt recognition of symptoms with an early diagnosis with CT scan and involvement of a multidisciplinary team of Gastroenterologist, Radiologists, dieticians, and surgeons (General and vascular) should form the objective of improving the nutritional status and weight gain with the alleviation of symptoms. If a conservative approach does not work, then surgical options should be exercised.

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