Study Of 18 Cases Of Superior Mesenteric Artery Syndrome At Tertiary Care Hospital

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Abstract: Background: Superior mesenteric artery (SMA) syndrome is a rare condition of duodenal obstruction, caused by the overlying SMA. Objective: To report on our experience with the management of SMA syndrome, drawing attention to its existence. Material And Methods: We reviewed our records to identify cases diagnosed with SMA syndrome, in the period from january 2010 to June 2018. Results: Eighteen patients were identified, six male and twelve females.Vomiting and abdominal pain were presenting complaints in all patients and history of weight loss was present in all of them. Only after radiological investigations was the diagnosis declared. Either Barium meal studies or computerized axial tomography (CT) scans with oral contrast was done. Surgery was performed in all patients with open duodenojejunostomy. Long lasting improvement was sustained in all patients except three in the surgery group who, despite initial improvement, still has infrequent attacks of abdominal pain. Interpretation and Conclusion: Although the clinical manifestations of SMA syndrome are shared with many other disease entities, it has unique radiological as well as endoscopic features, which enables a confident diagnosis to be made. Once diagnosed, conservative treatment with nutritional support should be tried first. In case of unresponsiveness, surgery may give a cure. [Suthar K Natl J Integr Res Med, 2020; 11(1):45-48]

Key Words: Arteriomesentric duodenal compression; Cast syndrome; Superior mesenteric artery syndrome; Wilkie's syndrome.

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Introduction: Superior mesenteric artery syndrome is a rare condition first described by Rokitansky in 1861 resulting from a reduced angle between the artery at its origin from the abdominal aorta and the transverse third part of the duodenum causing duodenal obstruction[1,2]. In young and otherwise healthy patients presenting with abdominal pain, nausea, anorexia, weight loss, and vomiting, the diagnosis of superior mesenteric artery (SMA) syndrome should be considered [1,3,4] . SMA syndrome differs from conditions with similar symptoms including familial neuropathic diseases, such as megaduodenum, in that it is a true obstructive condition without any underlying myopathy. In recent years, there have been numerous case reports of this condition, but in spite of this, diagnosis of this condition is frequently delayed resulting in ineffective symptomatic therapies and inappropriate investigations. This manuscript presents a series of eighteen SMA syndrome patients treated at a tertiary care hospital.

Material And Methods : From January 2010 to June 2018, eighteen patients presented to the General Surgical Unit of Sheth Vadilal Sarabhai Hospital. The unit is a tertiary care center with a population of approx 0.7 crores and five of the patients were referred from outside the normal drainage area.All the patients were examined clinically and subjected to upper GIscopy followed by barium meal study in some patients and computerised tomography with oral and rectal contrast in others. A prospective collated database was then retrospectively reviewed for demographic data, clinical presentation, diagnostic workup, treatment, and outcomes. Long-term follow up was obtained from patient review for all patients.

In this period, we were able to retrieve the files of all patients, six male and twelve females, with a mean age of 18.5 years. Presentation consistently involved persistent chronic vomiting, esophageal reflux, and abdominal pain. In six patients, this was associated with severe reflux, and in five patients, significant electrolyte abnormalities, particularly hypokalemia requiring intravenous fluid administration.The symptoms were chronic with length of symptoms ranging from between 4 to 16months (mean 10 months) prior to diagnosis.

Table 1: S&S Of SMA Syndrome

No.	Clinical Presentation	Number Of Patients
1.	Chronic Vomiting, Abdominal Pain	18
2.	Severe Reflux	6

3.	Significant Electrolyte Abnormality	5
4.	Significant Weight Loss	18

All patients had sought medical advice during this period. All had been diagnosed with reflux and treated with proton pump inhibitors. In all cases, there was a prolonged period of medical review, investigation, and treatment from the development of symptoms to final diagnosis.

Confounding the ability to accurately diagnose the condition were comorbidities including psychosocial conditions. These included alcohol abuse, eating disorders as well other illnesses such as pancreatitis. All patients had significant weight loss from the development of symptoms ranging from 10 to 15 kg (mean 13 kg) with the weight loss ranging from 20% to 30% of their body weight. Upper GIscopy were performed in all patients. Nonspecific findings of reflux esophagitis were described with only one endoscopist suspecting a possible obstruction of the third part of the duodenum (D3).

Suspicion of SMA syndrome occurred late in the disease course and only after the patients had a second opinion with the surgical team. All patients then underwent diagnostic imaging aimed at confirming the suspected diagnosis. All patients had either formal Barium meal (Ba meal) studies or computerized axial tomography (CT) scans with oral contrast which showed a dilated stomach and duodenum with an obstructive lesion in D3. In spite of the marked weight loss, all patients had serum albumin and serum proteins within normal limits.

Five patients had electrolyte abnormalities as previously noted, and in one of these patients, we elected to give 3 weeks of total parenteral nutrition (TPN) prior to surgery CT angiography was performed on all patients to assess the aortosuperior mesenteric artery angle. The angle ranged from 9° to 21° with a mean of 15° (normal 38 –68°). Scans also indicated reduction of the aortomesenteric distance with compression of the left renal vein seen in one case . Hence, all patients met the diagnostic criteria of SMA syndrome with radiological evidence of obstruction of D3, reduced aortosuperior mesenteric artery angle, and reduced aortomesenteric space.

Results: All patients were treated with mobilization and division of the fourth part of the duodenum with the end portion of the jejunum then being placed through the avascular portion of the right mesocolon and a side to side anastomosis between the third part of the duodenum and jejunum. All patients had uneventful postoperative recoveries. All patients had oral nutrition reinstated after 3 days. There were no wound infections or anastomotic breakdowns and the average length of stay was 9 days (range 7–14 days).

All patients had an excellent clinical outcome gaining between 3 and 9 kg at 6 months review. Postoperative imaging with Ba meal studies and endoscopy showed good emptying from the stomach and duodenum and no evidence of further obstruction.Three patient developed an incisional hernia for which they had incisional hernioplasty.One patient died due to severe head injury in road traffic accident after 5 years.Five patients had episodes of abdominal pain which relieved with medical treatment.All other patients remain well (follow-up range 5–68 months).

Table 2. Follow Of 18 Patients Of SMA Syndrome

No.	SYMPTOMS	Number of patients
1.	Severe abdominal pain	5
2.	Incisional hernia	3
3.	Death	1
4.	Symptom-free	9

Discussion: Wilkie described the clinical and pathophysiological characteristics of the syndrome as well as its management approach, in a series of 64 patients giving the syndrome its eponym “Wilkie's syndrome”[4]. Many other eponyms, including chronic duodenal ileus, megaduodenum, aortomesenteric artery compression, arterio-mesenteric duodenal obstruction, cast syndrome, and chronic duodenal pseudo-obstruction, have also been used[2,4,5]. The diagnosis of SMA syndrome requires correlation of clinical symptoms and radiological evaluation. Patient may present with chronic intermittent abdominal pain, vomiting, belching, postprandial epigastric fullness and

gastric outlet obstruction, On radiological investigation, contrast study show dilatation of the stomach and proximal duodenum with an abrupt cut-off across its third part and aortomesenteric angle [4,6,7].

Ultrasonography findings include to and fro movements across the duodenum ,with facilitation of the flow through the jejunum and elongation of the aortomesenteric distance when the patient assumes the right recumbent position. Although its confirmatory role, USG was not used in any of our patients to substantiate the diagnosis, probably due to its inability to provide clear anatomic details, compared to contrast [4,6].

The SMA takes off from the abdominal aorta at the level of the first lumbar vertebra with an average angle of 42.4° (range 18° to 70°) and a distance of 10–28 mm. Suspended by the ligament of Treitz, which is attached to its 4th part or to its junction with the jejunum, the duodenum crosses the abdomen at the level of the third lumbar vertebra. A narrow aortomesenteric angle of 15.2° (range 1°–40°) and a narrow aortomesenteric distance of 2 to 8 mm have been observed in individuals with SMA syndrome [1,4,7]. CT scan sagittal view is preferred for diagnosis. Thinning out of the fat pad between SMA and aorta, consequently upon weight loss, narrows the aortomesenteric angle and distance, thereby compressing the duodenum and thus producing the clinical manifestations of the syndrome. Other factors include an abnormally low origin of SMA, excessive lumbar lordosis, and hypertrophied or shortened ligament of Treitz or its multiple attachments to the duodenum [2,5]. High fixation of the duodenum by the ligament of Treitz or an anomalous SMA crossing directly over the aorta as the latter transects the duodenum has also been incriminated.

As noticed in our series, upper gastrointestinal series utilizing contrast will show dilatation of the stomach and duodenum down to the 3rd part, with a sudden cut-off distally which conforming to the anatomical position of the superior mesenteric artery. In addition, CT scan will show the anatomical relationship between SMA and aorta across the third part of the duodenum, resulting in the abrupt cut-off. On sagittal reconstruction of the CT images, a narrowed aortomesenteric distance and angle can be easily depicted, substantiating the diagnosis[4,6]. Unrelieved , duodenal perforation may ensue. Once patient diagnosed with SMA syndrome, medical management and surgical options do exist. It is intuitive to start with the medical lines first which include decompression of the stomach and duodenum with a ryle's nasogastric tube, correction of nutritional and electrolytes deficiencies, through TPN, or preferably, if possible, enteral feeding with a nasojejunal tube past the point of compression, which facilitates the nutritional management while avoiding TPN complications[1,2,4].

When tolerated, oral feeding may be resumed. This helps build up the fat cushion between the SMA and aorta and, hence, reversing the situation. SMA may be dragged by the small bowel mesentery, to drop off the duodenum when the patient assumes the prone or right recumbent position which can be proved by ultrasound studies. This might bring about symptomatic relief till the fat pad builds up. If medical management failed, Patient may requires surgical intervention. This may take the form of gastrojejunostomy or duodenojejunostomy, by the open or laparoscopic means .

Releasing of ligament of treitz from its attachment to duodenum, mobilisation of 3rd and 4th part of duodenum and releasing the compression, has also been reported. More recently, robotic duodenojejunostomy has been utilized with success[4,5]. In our study, all the patients underwent duodenojejunostomy out of which five patients had episodes of abdominal pain and three developed incisional hernia and the rest had good post operative recovery.Other pathological conditions with similar clinical presentation, including diabetic gastroparesis, scleroderma with duodenal involvement, hereditary megaduodenum, megaduodenum due to aganglionosis, have been rarely reported. Published articles are mainly case reports and, rarely, small case series. This limits our understanding of the disease.

Conclusion: The SMA syndrome should be considered as a potential diagnosis in young adults who present with a history of persistent postprandial vomiting and weight loss. Although the clinical manifestations of SMA syndrome are shared with many other disease entities, it has unique radiological as well as endoscopic features, which enable a confident diagnosis to

be made. Once diagnosed, conservative treatment with nutritional support and positioning should be tried first. In case of unresponsiveness, surgery may give a lasting cure.

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