SUPERIOR MESENTERIC ARTERY SYNDROME - AN UNCOMMON CAUSE OF SMALL BOWEL OBSTRUCTION: A CASE REPORT

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ABSTRACT: Superior Mesenteric Artery Syndrome (SMAS) or Wilkie's disease is a rare form of obstruction of the transverse part of duodenum entrapped between superior mesenteric artery and abdominal aorta. SMAS presentation is usually non-specific and poses a diagnostic dilemma. A case of 45 year old asthenic male with no prior surgical history presented to emergency department with epigastric mass, post prandial epigastric pain and intractable vomiting for four months with significant weight loss. On radiological investigations diagnosis was made and consistent with SMA syndrome. Patient was managed conservatively for two weeks, as symptoms persisted duodenojejunostomy was performed and patient was relieved of obstructive symptoms and showed satisfactory nutritional improvement in follow up. SMA syndrome should be considered as possible etiology of small bowel obstruction in specific set of patients.

KEYWORDS: Superior Mesenteric artery; aorto-mesenetric angle; Wilkie's Disease.

INTRODUCTION: Superior mesenteric artery syndrome is an atypical clinical disorder of vascular compression affecting third part of duodenum; first described by ROKITANSKI in 1861, later WILKIE coined the term "chronic duodenal ileus" and published a detail description of seventy five cases in 1927 since then, although several cases are reported so far the disease remains a subject of controversy and many have doubted its actual incidence. Modern imaging techniques nevertheless have restricted the trend of over diagnosis. The young age with non-specific symptoms often lead to delay in diagnosis. This entity, over the years, acquired different names, such as chronic duodenal ileus, arteriomesenteric duodenal compression, Wilkie's syndrome and cast syndrome. As the diagnosis of SMAS is generally difficult so it's imperative to convincingly exclude other causes which cause duodenal obstruction. The surgery first proposed for SMAS was duodenojejunostomy by BLOODGOOD.

CASE REPORT: A 45year old asthenic male presented to our emergency department (ED) with epigastric mass, postprandial epigastric pain, intractable bilious vomiting for four months with loss of appetite and significant weight loss and with no prior surgical history. He was on antiemetics without any relief.

On examination, patient was 149cm tall with body weight of 25kg; vitals were stable with signs of minimal dehydration. Abdomen was scaphoid with epigastric fullness. No hepatomegaly or ascites present. Ausculto-percussion and succusion splash test gave an impression of gastric distension.

The patient's hematological and biochemical results were normal. Erect abdomen X-ray and barium meal series revealed double bubble sign (fig. 1) and an abrupt cut-off at the level of

third part of duodenum (D3) with proximal dilatation and delayed gastroduodenal emptying respectively (fig. 2). Upper gastro intestinal endoscopy reported gastro duodenal stasis without any demonstrable intrinsic lesions. High resolution ultrasonography and contrast enhanced CT scan abdomen detected narrowed segment of D3 with reduction in aortomesenteric (AM) angle to 22° and aortomesenteric distance to 7.3mm (fig. 3). The clinical presentation and imaging findings correlated with diagnosis of SMA syndrome.

Patient was treated conservatively with nasogastric tube decompression, small, frequent, high caloric feeding, postural advice and supplementation for two weeks, as symptoms remained refractory surgery was planned. Exploratory laparotomy was done, dilated stomach, 1st, 2nd and a portion of 3rd part of duodenum with extrinsic compression of transverse part of duodenum noted (fig. 4a). Duodenojejunostomy was performed (fig. 4b). Patient recovered uneventfully. During four months of follow up he was symptom free and with weight gain of 4kgs.



Fig. 1: Erect X ray Abdomen with double bubble sign



Fig. 2: Barium Meal Series



Fig. 3: CECT with Narrowed AM angle and AM distance



Fig. 4a: SMA compressing D3



Fig. 4b: DUODENOJEJUNOSTOMY

DISCUSSION: SMA syndrome is a rare condition and was first described by von Rokitanski in 1861.¹ Wilkie later provided a more detailed anatomical, clinical and patho-physiologic description and named it chronic duodenal ileus.² Wilkie later published case series of seventy five cases in

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1927.³ SMA usually forms an angle of approximately 45° with abdominal aorta at its origin while the third part of the duodenum crosses in between the SMA anteriorly and aorta posteriorly. Factors such as dramatic weight loss resulting in loss of retroperitoneal fat which acts as cushion around SMA, high insertion and shortness of ligament of Treitz, low origin of SMA, peritoneal adhesions, malrotation of duodenum, rapid linear growth without compensatory weight gain, scoliosis and body casting will sharply narrow the aorto-mesenteric angle to less than 25° and can cause entrapment and compression of the third part of the duodenum, resulting in Superior Mesenteric Artery Syndrome.⁽⁴⁻⁸⁾

SMA syndrome can present as acute or chronic intermittent duodenal obstruction. Postprandial epigastric pain, fullness, bilious vomiting and rapid weight loss are characteristic symptoms. Food aggravates while certain postural adjustments like left lateral position, knee chest position or prone position will relieve symptoms. As obstruction is incomplete, diagnosis is challenging, often made by exclusion.^(5,7) Barium study findings although characteristic but are not specific for SMA syndrome. Non-invasive modalities like color doppler ultrasonography, CT or MR angiography are currently useful tools for diagnosis.^(4,5,7,8) Conservative management, consisting of frequent and small feedings has been successful at times and should be tried initially. Surgery is indicated in longstanding and unresponsive cases or massive duodenal dilatation and stasis.^(5,8) Open or laparoscopic Duodenojejunostomy is the most accepted procedure with success rate of 90%.^(4, 5, and 8) Cleavage of the ligament of Treitz is another option, enabling the duodenum to drop away from the apex of the sharpened aorto-mesenteric angle.

CONCLUSION: SMA syndrome is a rare form of proximal small bowel obstruction with nonspecific clinical presentation. A high index of clinical suspicion for those with predisposing factors, and imaging are paramount for early diagnosis of SMA syndrome. Conservative management is successful in most of patients with definitive surgery reserved for refractory cases.

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