

## Superior Mesenteric Artery Syndrome: A Case Report

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### Introduction:

**S**uperior mesenteric artery syndrome (SMAS) is the result of compression of the transverse part of the duodenum due to narrowing of angle between the SMA and the abdominal aorta. It is also known by a variety of other names like Cast syndrome, Wilkie syndrome, arteriomesenteric duodenal obstruction, and chronic duodenal ileus.<sup>1</sup> Till date, only 400 cases have been reported in the medical literature.<sup>2</sup> Diagnosis of the syndrome depends on high index of clinical suspicion and the radiological features of the syndrome. The radiological appearances of this syndrome on barium meal are non-specific although it has conventionally been used. Examination of the aortomesenteric angle and aortomesenteric distance on multidetector computed

### ABSTRACT:

Superior mesenteric artery syndrome (SMAS) is a rare condition caused by compression of the third portion of the duodenum between the superior mesenteric artery (SMA) and the aorta, causing symptoms of duodenal outflow obstruction. We report a case of SMAS in a young emaciated female with well-documented diagnostic imaging findings.

**Keywords:** Duodenal obstruction; abdominal pain; superior mesenteric artery syndrome; imaging

tomographic (CT) scan can conclusively diagnose this syndrome.<sup>3</sup> Depending on the severity treatment can be either is conservative or operative. Here we report such a case of SMA syndrome in a young patient with history of rapid weight loss.

### Case report:

A young female presented with features of chronic gastric outlet obstruction. She had a prolonged history of recurrent pain in the epigastrium, on and off fever and multiple episodes of vomiting. She also gives history of significant loss of weight in the past 6 months. She was on empirical antitubercular treatment for a pulled up caecum which was seen on a barium meal follow through earlier. On

examination, the patient was emaciated and afebrile. Her vitals were stable. The abdominal examination revealed a slightly distended upper abdomen. There was no relief of obstruction in the left lateral decubitus or prone position. Blood investigation revealed a normal cell counts except for low haemoglobin (8.3 g/dl). Erect plain abdominal radiograph was normal. Barium study done by us showed a distended stomach with an abrupt interruption in the contrast flow at the level of third part of duodenum (D3) (Figure 1).

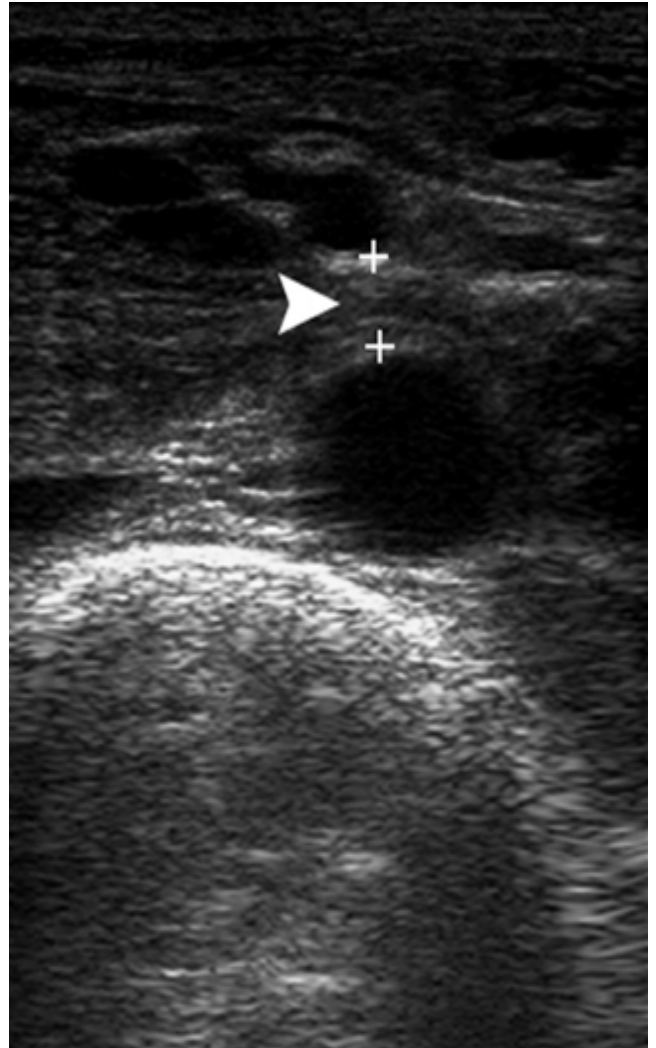
**Figure 1:** Barium follow-through shows a distended stomach and dilated duodenum up to its proximal third part.



Ultrasonography revealed a narrowed angle and short distance between the superior mesenteric artery and the aorta and a compressed third part of duodenum (Figure 2a). The imaging findings and clinical presentation correlated the diagnosis of SMAS.

**Figure 2:** Axial USG (a) and CT (b) images showing a narrowed aortomesenteric distance with compressed third part of duodenum between the superior mesenteric artery and the aorta (arrowhead). CT shows distended stomach

and first and second part of duodenum (small arrow).



*Figure 2a*

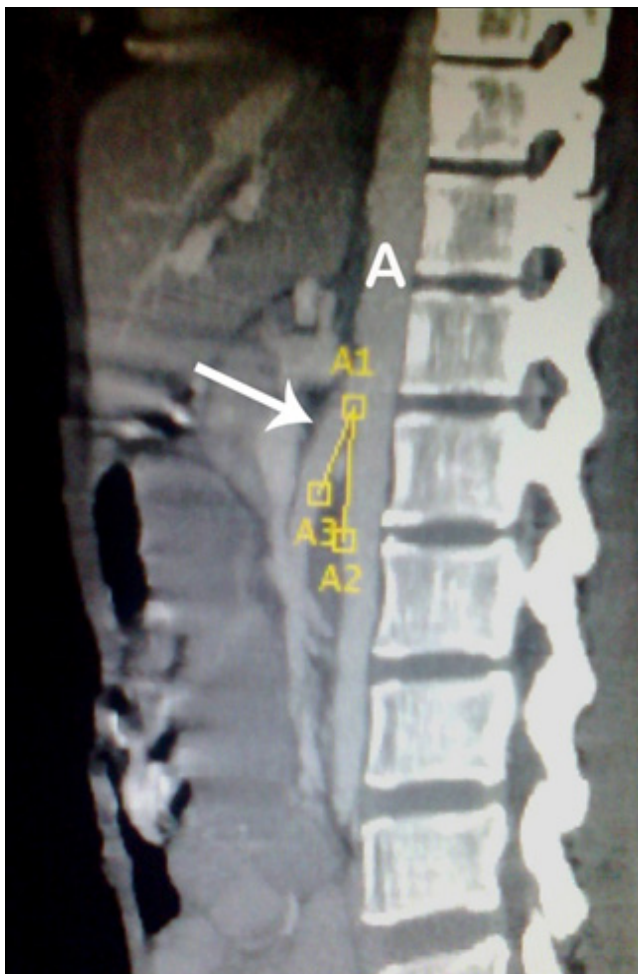
CT was advised to confirm the findings and to rule out other abdominal pathology. In the arterial phase of the CT, the distance between the abdominal aorta and the SMA was reduced (4.5 mm), the third part of the duodenum was compressed and the stomach and proximal duodenum were dilated (Figure 2b).



*Figure 2b*

However, the distal bowel loops were filled with contrast suggesting partial the obstruction. A few enlarged calcified retroperitoneal nodes were also seen around the duodenum. On the reconstructed multiplanar reformatted (MPR) image (Figure 3) the angle between the SMA and the aorta was narrowed ( $17^\circ$ ).

**Figure 3:** Multiplanar reformatted CT image showing a narrowed aortomesenteric angle. The third part of the duodenum is compressed between the SMA (arrow) and the aorta (A).



On the basis of the clinical history and the imaging findings, a diagnosis of SMA syndrome was made. The patient was managed by duodenojejunostomy which improved her obstruction, appetite and subsequently the cachexic status as well. In post-operative follow up, patient was symptom free.

#### Discussion :

The incidence of the SMAS is as low as 0.2%.<sup>3</sup> The SMA usually forms an acute angle with the abdominal aorta, and the third part of the passes between them. Normally surrounding

fat and lymphatics maintains this angle and provide protection for duodenal compression. In SMA syndrome., aortomesenteric angle is more acutely narrowed to  $7^\circ$  to  $22^\circ$  (normal range  $25^\circ$  to  $60^\circ$ ) and SMA aorta distance is reduced to 2-8 mm (normal range 10 to 28 mm).<sup>4,6</sup> There are several factors that can reduce the angle; the most common cause is significant weight loss leading to loss of the mesenteric fat cushion. Other causes include an abnormal high insertion of ligament of Treitz, a short ligament of Treitz, congenitally low origin of the SMA, peritoneal adhesions and duodenal malrotation.<sup>7</sup> In our case the syndrome was precipitated by an excessive weight loss.

Usually young females are affected and the symptomatology is commonly chronic as in our patient. The acute presentation is uncommon and usually occurs in young, thin women.<sup>6</sup> Characteristic clinical symptoms include early satiety, nausea, postprandial fullness and epigastric pain, weight loss, and bilious vomiting. The symptoms are often relieved by postural changes. Diagnosis depends on high index of suspicion as symptoms can be non-specific.

Conventional barium meal and hypotonic duodenography are important for the diagnosis of SMA syndrome. On these studies although radiological appearances are characteristic but not specific for SMAS. In patients with inexplicable abdominal pain, to suspect a SMA syndrome, ultrasound is valuable tool for a screening of reduced aortomesenteric angle.<sup>8</sup> Earlier, angiographic measurement of the aortomesenteric angle was noted as being a more accurate investigation than routine barium meal examination to suspect a SMA syndrome. In the current days, non-invasive modalities like ultrasonography, CT or magnetic resonance (MR) angiography are useful tools for diagnosing SMAS.<sup>4,6,9</sup> Recently, Konen et al have highlighted the advantage of 3-dimensional CT angiographic reconstruction study in the diagnosis of SMAS.<sup>10</sup> CT criteria for the diagnosis of SMA syndrome include the aortomesenteric angle (the angle between the SMA and the aorta, which is measured on sagittal MPR images) of less than 22 degrees and the aortomesenteric distance (the shortest distance between the walls of the SMA and the aorta at the level where the third part of duodenum passes between them) of less than 8-10 mm.<sup>11</sup> The initial treatment of this condition is generally conservative. In our patient, there was significant reduction of the aortomesenteric angle and distance. Surgery is indicated in longstanding and unresponsive cases or in massive duodenal dilatation and stasis.<sup>5,9</sup>

**Conclusion:**

SMAS is a rare clinical condition that is clinically a diagnosis of exclusion. However, ultrasonography and multidetector CT are proven to be useful imaging modalities for its diagnosis and management.

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