

# Superior mesenteric artery syndrome

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## Abstract

Superior mesenteric artery syndrome is a very uncommon disorder in which acute angulation of superior mesenteric artery (SMA) at its origin from aorta results in compression of the third part of the duodenum leading to partial or complete obstruction of the duodenum. Other terminologies for this condition are Cast syndrome, Wilkie syndrome, arteriomesenteric duodenal obstruction, and chronic duodenal ileus.<sup>1</sup> The disease is extremely rare in the pediatric population. Here we report a case of SMA syndrome in an 11 year old boy who was admitted with hepatitis, subsequently developed primary peritonitis and during his admission at the hospital he developed SMA syndrome. The case is being reported for its extreme rarity, to highlight the role of imaging in establishing the diagnosis and to review the literature in the subject.

## Keywords

- Superior mesenteric artery (SMA) syndrome
- Wilkie syndrome
- cast syndrome
- pediatric

## Introduction

In this syndrome small intestine is compressed at the level of the third portion of duodenum because the space between the superior mesenteric artery and aorta is narrowed which is considered to be due to loss of the intervening mesenteric fat pad. The condition is said to occur due to acute nutritional deficiency following surgery and orthopedic trauma. SMA syndrome is one of the unusual causes of proximal GI obstruction with the reported incidence of 0.1 to 0.3% <sup>2</sup> and extremely rare in pediatric population. One pediatric study has reported the largest series of 22 cases over 20 years.<sup>3</sup>

## Case report

An 11 year old boy with jaundice and fever with the diagnosis of severe viral hepatitis was admitted to the pediatric intensive care unit. He had a very high bilirubin level (total 13.2 mg), predominantly indirect and elevated transaminase levels. On the third week of illness he developed abdominal distension, bilious nasogastric aspirate and abdominal guarding and tenderness. X-ray and CT scan showed thickened and dilated small bowel loops with a moderate amount of free fluid. With a high suspicion of intestinal obstruction or peritonitis in mind, laparotomy was carried out

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which found purulent free fluid and inflamed bowel loops and intra operative diagnosis of primary peritonitis was made as no other pathology could be found. Irrigation of the peritoneal cavity was done and a drainage tube was placed. The child was kept nil by mouth; from the fourth post operative day bilious nasogastric drainage started and the child was found to have very low serum potassium and magnesium levels. Hypokalemia did not improve in spite of repeated corrections. UGI endoscopy was performed to rule out any rare pathology affecting the duodenum due to the absence of dilated loops in the X-rays; which showed extrinsic compression of the third part of the duodenum.

A repeat CT scan was done one week after the first laparotomy and showed significant reduction in the aorta-SMA angle (25 degrees). CT scan images typically show the narrowing of SMA - aortic angle and narrowing of the duodenal lumen after development of the syndrome following acute weight loss (Fig.2, 3, 4 and 5) and a comparison picture of an age matched control (Fig.1). Since SMA syndrome was strongly suspected the child was started on parenteral hyper alimentation and supportive measures. The bilious aspirate gradually decreased, and the child improved.



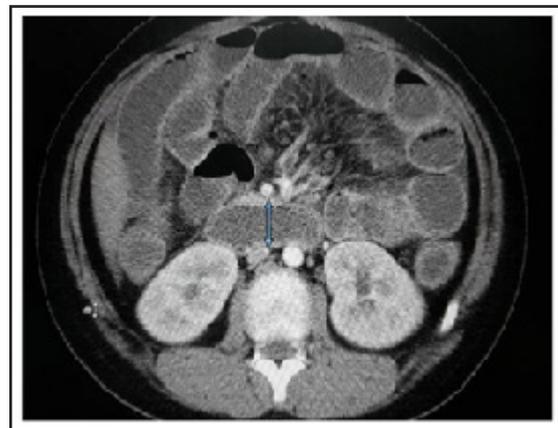
**Figure 1** Aortomesentric angle in a normal individual



**Figure 2** Aortomesentric angle before developing sma syndrome in our case



**Figure 3** Narrowed aortomesentric angle in our case with collapsed duodenum



**Figure 4** CT Section showing normal lumen of third part of duodenum before developing sma syndrome



**Figure 5** CT Scan showing critically narrowed third part of duodenum.

## Discussion

The first report of SMA syndrome took place in 1861 by Von Rokitansky. There are several reports and studies in the adult literature since then. The normal angle between superior mesenteric artery and the abdominal aorta is approximately 45°, and the third part of the duodenum crosses below the origin of the superior mesenteric artery, lying between the artery and aorta.

When the aortomesenteric angle decreases below 25° it could cause compression of the duodenum as it passes between the aorta and superior mesenteric artery, resulting in superior mesenteric artery syndrome.<sup>4</sup>

Also aortomesenteric distance decreases to 2-8 mm (normal is 10-20 mm) in this syndrome. Alternatively, other causes suggested in superior mesenteric artery syndrome include: a high insertion of the duodenum at the ligament of Treitz, origination of the SMA at a lower point on aorta, and duodenal obstruction due to adhesions.

SMA syndrome usually occurs after acute weight loss; such as that seen in hyperthyroidism, gastroenteritis, burns or after orthopedic and spinal surgeries. Acute weight loss precipitates the loss of fat pad in the mesentery leading to duodenal compression.<sup>5,6</sup> The child reported here had a significant weight loss of 20 percent during his three weeks of hospital stay.

While malrotation with volvulus or congenital bands are close differential diagnoses in a case of persistent or recurrent abdominal pain with bilious vomiting, clinical background and modern CT scans can give near accurate diagnosis. Conventionally hypotonic duodenography done with duodenal relaxation using Hyoscine was the gold standard diagnostic investigation. The following radiologic criteria have been suggested for the diagnosis of SMAS: dilatation of the proximal duodenum with or without dilatation of the stomach; abrupt vertical and

oblique compression of the mucosal folds; proximal to the obstruction an antiperistaltic flow of barium is seen which produces to-and-fro movements; delay in transit through the gastroduodenal region (4 to 6 hours); and relief of obstruction when the patient is placed in a position (prone or knee-chest) that diminishes the drag of small-bowel mesentery.<sup>9</sup> Several new studies have shown how CT scan is valuable in diagnosing SMA syndrome and for the follow up during resolution of symptoms.<sup>7,8</sup>

Treatment of acute onset SMAS is usually conservative. Management is focused on bowel decompression, administration of fluid and electrolytes and nutritional support or rehabilitation. Removal of the body cast is required when SMAS is associated with cast syndrome.<sup>10</sup> Once stabilized, small frequent high caloric oral feeds may be effective. Changes in feeding position, either both knees compressed to the chest or right lateral decubitus may also be useful in bypassing the obstruction. If changes in feeding position do not prove effective, a nasojejunal tube can be placed which goes past the obstruction to allow for enteral feeding. Some patients benefit from use of prokinetic agents such as Metoclopramide.<sup>8</sup> Patients who do not improve with these measures will require total parenteral nutrition.

Surgery is usually needed for patients who do not respond to conservative management and patients with chronic intermittent symptoms. Patients with acute weight loss due to trauma, surgery or burns usually respond well to conservative management, some studies report 100 percent success rate with conservative treatment.<sup>5,12</sup> Successful surgical therapies include duodenojejunostomy, gastrojejunostomy, which involve an anastomosis and cause some physiological disturbance. Excision of the ligament of Treitz<sup>13</sup> and anterior transposition of duodenum is considered to be more physiological.<sup>14</sup>

## Conclusion

SMA syndrome is a well established but very rare cause of proximal GI obstruction. The disease is even more uncommon in pediatric age group. Our experience and literature suggest that in post operative settings accompanied by acute weight loss, unexplained bilious aspirate and electrolyte imbalance might be due to SMA syndrome; after ruling out other common causes like postoperative ileus and adhesions, and CT is extremely valuable in making the diagnosis monitoring the improvement while the patient is being managed conservatively.

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