Successful Medical Management of a Child with Superior Mesenteric Artery Syndrome

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ABSTRACT
Superior mesenteric artery syndrome is a relatively rare but known condition that may affect children. This condition can be managed conservatively and that with nutrition hence avoiding major surgical intervention. A previously fit 13-year-old boy presented with non-bilious vomiting that had turned bilious, over 5 days making sub-acute intestinal obstruction likely. Barium meal and follow-through were conclusive of superior mesenteric artery syndrome (SMAS) which was confirmed with a CT abdomen. He was managed with parenteral nutrition and positioning when taking oral feeds. He made a full recovery and demonstrated weight gain and continued to remain well without vomiting on follow up. Although a rare condition, SMAS should be considered in children with acute vomiting which can be managed conservatively as highlighted by this case without any known precipitating causes. The medical condition of SMAS is a known yet underused treatment which may avoid major intraabdominal surgery. In cases such as our parenteral nutrition was used as a rescue treatment and enteral nutrition followed recovery and convalescence.

Key Words: Superior mesenteric artery syndrome, Children, Vomiting, CT abdomen

INTRODUCTION
Superior mesenteric artery syndrome (SMAS) is a rare yet recognised syndrome in children and adults alike. SMAS happens due to sudden loss of mesenteric pad of fat around the duodenum which leads to mechanical compression of the duodenum by the two large arteries Aorta and Superior mesenteric artery and hence the name Superior Mesenteric artery syndrome. SMAS has been reported in children with acute vomiting, post spinal surgery, burns and some rare occurrence of familial cases and association with coeliac axis compression syndrome have also been reported.

CLINICAL PRESENTATIONS
A previously fit and well 13-year-old boy presented to the children’s outpatients with a 5-day history of non-bilious vomiting. He was admitted for fluid management and monitoring. His initial single episode of bilious vomiting was investigated with a plain abdominal x-ray which revealed normal gas pattern and constipation and his clinical signs had not changed since admission. On admission, he was noted to be emaciated but well hydrated. His biochemical and haematological indices were all within normal limits.

The main concern was weight loss, his weight before admission was 38 and his weight currently was 34.6.

During the course of stay he started to have bilious vomits which led to a clinical diagnosis of sub-acute intestinal obstruction which was further managed with broad-spectrum IV antibiotics, nasogastric tube on free drainage and commencement of total parenteral nutrition. Barium meal and follow-through showed dilatation of 3rd part of the duodenum and no contrast flowed past this level. The contrast passed freely when the patient was placed prone raising SMAS as a strong possibility. We performed a CT scan of his abdomen (320 slices) 48 hours post barium meal and follow-through, as a large volume of barium remained in his abdomen.

The CT scan of his abdomen showed compression of his duodenum between his aorta and superior mesenteric artery with mild dilatation of proximal duodenum (Figure 1 and 2). The aortomesentric angle is narrow and measured approxi-
Mately 9.7 degrees. Critical proximal stenosis of the celiac artery by the arcuate ligament was also noted, the findings confirming the diagnosis of SMAS.

**MANAGEMENT AND OUTCOMES**

Total parenteral nutrition (TPN) was continued and oral feeds (high energy feeds) was started concurrently. After 72 hours of commencing TPN he was allowed to eat soft semi-solid diet. He tolerated diet very well and demonstrated weight gain and his TPN was gradually weaned. During the entire period of above, he was nursed in the left lateral position hence eliminating the pressure on the duodenum by the superior mesenteric artery and aiding rapid recovery. On follow up, his weight gain had remained progressive and his vomiting had completely resolved. This case demonstrates the onset of SMAS after acute weight loss following vomiting in a previously well child. It also emphasises the importance of TPN and nutritional rehabilitation in acutely unwell children and a rare condition like SMAS.

**DISCUSSION**

Superior mesenteric artery syndrome is a rare condition with an incidence 0.013% to 0.3%. SMAS results from compression of the duodenum between the abdominal aorta, posteriorly and superior mesenteric artery, anteriorly thought to be due to loss of duodenal fat pad. The condition was first described in von Rokitansky in 1842.SMAS was associated with orthopaedic casting by Willet in 1878.Wilkie in 1927 formally characterized SMAS in a series of 75 patients. Typical cases of SMAS present after orthopaedic surgical casting or after spine surgery and after acute weight loss. Girls are more commonly affected than boys. The common causes of acute weight loss are hyperthyroidism, anorexia nervosa or gastroenteritis. The symptoms of SMAS are nausea, upper abdominal pain, early satiety, anorexia. The symptoms described by patients are similar to small bowel obstruction. The upper abdominal pain described by nearly half of the patients is located in the epigastric region. The diagnosis of SMAS is confirmed by upper gastrointestinal radiography and CT abdomen has also been used to confirm it. Rarely patients have been diagnosed by laparotomy.

There are two large single-institution series published by Burrington and Wayne in 1974 and a paediatric case series by Vincent Bank and Steve Werlin in 2006. The latter study adds that low BMI is not a risk factor for SMAS and only 50% of their cohort had weight loss before diagnosis. Their median length of time from onset of symptoms to diagnosis was only 5 days. Patients with neurological injury are at high risk of acquiring SMAS when compared to the general population.

The management of SMAS is normally conservative. The recent advances in both enteral and parenteral nutrition have made a significant impact on the management of SMAS. The following need to be the mainstay of treatment and meticulously monitored during the treatment and recovery phase of SMAS i.e. bowel decompression, fluid and electrolyte balance with aggressive nutritional support and rehabilitation. Changes in feeding posture such as right or left lateral decubitus position may also facilitate quick recovery.

If medical treatment fails to achieve resolution of symptoms surgical intervention becomes necessary. Surgery becomes the mainstay if there is dilatation and/or stasis of small bowel. The surgical procedures that have been used are duodenojunostomy, gastrojejunostomy, or resection of the ligament of Treitz. Nasojejunal feeding has also been used as a feeding adjuvant in the management of SMAS. The complications of SMAS are dehydration and electrolyte imbalance particularly refeeding syndrome during the recovery phase. All patients with SMAS make a complete recovery and death from SMAS is rare.

This case highlights a rare but fully treatable cause of vomiting in a previously well 13-year-old child. Early diagnosis and aggressive nutritional management are of paramount importance and surgical intervention should only be reserved to patients who are refractory to medical therapy or if laparotomy is required at presentation.

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Figure 1: CT scan abdominal part of subject with Superior mesenteric artery syndrome.

Figure 2: CT scan abdomen of subject with Superior mesenteric artery syndrome.