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CASE REPORT

Superior mesenteric artery syndrome in an adolescent male

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Summary
This report describes a 13-year-old male patient admitted with superior mesenteric artery syndrome (SMAS) requiring urgent surgery for decompression due to worsening pain and elevated lactate levels. SMAS is a rare cause of upper gastrointestinal obstruction and in this case may have been due to a rapid growth spurt.
Keywords: rapid growth spurt, superior mesenteric artery syndrome

Case report

A 13-year-old male with no known comorbidities presented to the emergency department with a one-day history of severe epigastric pain, anorexia, nausea, and profuse bilious vomiting. On further enquiry, the patient had experienced a similar but less severe episode six months prior to the index presentation where no medical help was sought as the symptoms resolved with a fluid diet. These symptoms occurred during a time of rapid height gain. Vital signs on presentation were within normal limits. The patient had a significantly distended upper abdomen with severe epigastric tenderness and a succussion splash. All other systems examined were within normal parameters.

A full blood count noted an elevated white cell count of 16.6 x10^9, normal haemoglobin 14.9 g/dl, MCV 88.1 fl and platelets 370 x10^9 signifying a mild inflammatory response. His renal function showed a prerenal picture with normal sodium and potassium but a urea of 6.2 mmol/l and a creatinine of 102 umol/l.

A chest X-ray showed a large gastric bubble but no signs of free air in the peritoneal cavity. An abdominal X-ray revealed a grossly distended gastric outline, containing food content, and a proximal small bowel obstruction.

A nasogastric tube was passed and drained 1500 ml of bilious stomach content instantly. The patient was kept nil per mouth, with the nasogastric tube on free drainage, and intravenous fluid was administered. An urgent computer tomography (CT) scan with intravenous contrast was performed as the child was extremely uncomfortable despite analgesia and fluid resuscitation. The CT scan showed an obstruction at the third part of the duodenum with gross stomach and proximal duodenal distension (Figure 1). A clear transition point was seen between the abdominal aorta and superior mesenteric artery (SMA) origin with an

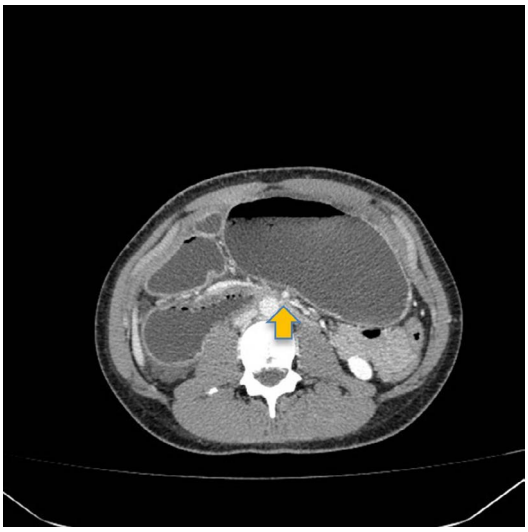

Figure 1: Abdominal CT scan noting obstruction of 3rd part of duodenum between aorta and SMA

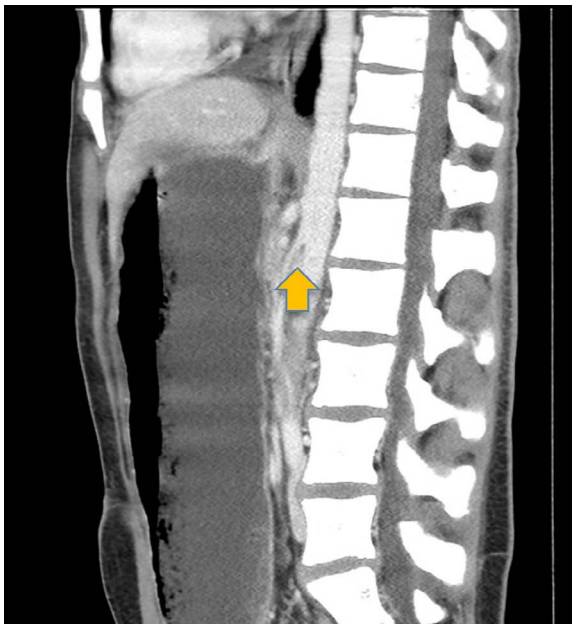

Figure 2: Sagittal view of CT scan noting aortomesenteric distance of 4 mm and aortomesenteric angle of 7 degrees

aortomesenteric distance of 4 mm and an aortomesenteric angle of 7 degrees (Figure 2).

The CT findings were diagnostic of superior mesenteric artery syndrome (SMAS).

Conservative treatment was contemplated but the patient was extremely uncooperative, repeatedly removing the nasogastric tube and refusing nasogastric or enteral feeds. His epigastric pain progressively worsened. A repeat blood gas 48 hours after admission showed a raised lactate of 8 mmol/l with all other parameters within normal limits. Therefore, the decision was to perform an urgent surgical decompression.

Intraoperative findings were a grossly enlarged stomach with patchy areas of ischemia at the fundus but no signs of necrosis. An open Roux-en-Y duodenojejunostomy was performed. The patient did well postoperatively and was tolerating a full ward diet and passing stool on day four. He was discharged five days postoperatively.

At six-weeks follow-up the patient was symptom free and had gained 1.6 kg.

Discussion

SMAS, otherwise known as Wilkie's syndrome, cast syndrome or chronic duodenal ileus, is a rare cause of upper gastrointestinal obstruction. Characteristically the third part of the duodenum is compressed by the aorta and SMA causing the typical symptoms of early satiety, anorexia, abdominal discomfort or pain, nausea, and vomiting. The normal SMA to aorta angle has been described to range from 38–56 degrees and 10–20 mm in length but in SMAS the angle is decreased ranging from 6–25 degrees and the length 2–8 mm.[1,2] The patient had an angle of 7 degrees and 4 mm in length, which is likely why his obstruction was so severe.

Risk factors for SMAS are associated with conditions or situations leading to a decreased angle between the aorta and SMA, most notably significant weight loss causing loss of the fat pad normally supporting the aorta and SMA angle, also seen in adults with malignancy, AIDS, malabsorption syndromes, following trauma or burns, anorexia nervosa and after bariatric surgery.[3]

In the paediatric population, however, weight loss is not always responsible for SMAS. Case reports have described instances of SMAS occurring in paediatric patients with insufficient weight gain relative to height growth. This is often seen in pubertal males with an increase in lean body mass with associated loss of adipose tissue. Corrective spinal surgery has also been implicated as a cause of SMAS as they increase the length of the spine cranially and displace the origin of the SMA superiorly therefore decreasing the angle between the SMA and aorta, but in the vast majority of cases the cause of SMAS remains unknown. Presentation typically occurs in adolescents and young adults in the age range of 10–39 years and is more commonly seen in females than males.[4-7]

This patient reported quite significant height growth in the last year, weighing 54.55 kg and a length of 179 cm on admission, equalling a BMI 17.0. This is presumably the reason for the presentation of SMAS within this patient.

Conservative treatment of this condition has been described with total parenteral feeding and nasojejunal feeds with high calorie liquid feeds. The aim is to encourage weight gain to restore the fat pad, reverse the loss of angle and decrease the distance between the aorta and SMA. This is done for six to eight weeks, and if symptoms do not respond then surgery is recommended.[8] This was not a feasible option for this patient as he was in significant discomfort with a high lactate and refusing a nasogastric tube or central venous line placement for feeding.

Surgical treatment options include gastrojejunostomy, duodenojejunostomy or lysis of the ligament of Treitz. Surgical management has been noted to have a success rate of > 92% with 0% recurrence at 12-month follow-up. Compared to medical/conservative management which has a success rate of 71.3% with a recurrence rate of up to 15.8% over the same period.

Gastrojejunostomy has largely been abandoned as a treatment for SMAS due to its high complication rate. These complications include dumping syndrome, blind loop syndrome and marginal ulceration. Appel et al. had a 100% success rate with open duodenojejunostomy on 36 patients with SMAS in 1976, a marked increase as opposed to lysis of the ligament of Treitz with 79% success rate. Lysis of the ligament of Treitz, first described in 1958, has the major advantage of circumventing the need to anastomose the gastrointestinal tract and the risks associated with anastomotic leakage. The first successful laparoscopic duodenojejunostomy was performed by Gersin and Heniford in 1998.[1,9]

Currently, laparoscopic duodenojejunostomy has been seen as the surgical treatment of choice for adults due to its high success rate and minimally invasive nature. Mauney et al.[4] published a case series which proposed that laparoscopic duodenojejunostomy is a safe and effective treatment in paediatric patients with SMAS. The case series also highlighted that patients were symptomatic for a median of 10 months with trials of conservative treatment before surgical intervention, as compared to this case.[4]

As seen in this patient, SMAS is a rare disorder with possible deadly consequences if not diagnosed early and managed appropriately. The diagnosis, however, relies on clinical suspicion from the treating clinician as well as access to imaging to confirm the diagnosis. As demonstrated in the case above, a recent growth spurt in a patient with inadequate weight gain in relation to height might precipitate the onset of symptoms of SMAS. Earlier consideration for surgical management may be indicated, as it is shown to be safe and effective, compared to guidelines recommending conservative management for six to eight weeks.

Conflict of interest

The authors declare no conflict of interest.

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Ethical approval

The authors declare that this submission is in accordance with the principles laid down by the Responsible Research Publication Position Statements as developed at the 2nd World Conference on Research Integrity in Singapore, 2010. All procedures were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008. Informed written consent was obtained from the patient in the case report.

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