CASE REPORT

Superior mesentric artery syndrome in pediatric population: An ardous manifestation of duodenitis

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SUMMARY

Superior mesenteric artery (SMA) syndrome is a rare duodenal 'sandwich' due to narrow SMA-aorta angle, particularly observed in the pediatric age group. This compression can lead to acute or chronic abdominal pain. In this case report, a 11 years old active boy came with chronic upper gastrointestinal symptoms. Initially treated with anti-reflux medication, the patient's condition worsened with the emergence of red-flag symptoms. The diagnosis was established after computerized tomography (CT) scan was done that revealed acute angulation of SMA and aorta. This case report stresses the challenges in diagnosing the disease due to its' vague symptoms manifestation and the importance of early detection for effective management.

INTRODUCTION

Superior mesenteric artery (SMA) syndrome is a rare condition where the third portion of the duodenum is compressed by the narrow angle between the SMA and the aorta. Several factors contribute to the acute angulation between the SMA and the aorta, as shown in Table I. The prevalence in pediatric patients is not well-known, but studies suggest it is rare, ranging from 0.013 to 0.30 percent in the general population. SMA Syndrome was first described by Rokitansky 1 in 1861 diagnosed by a series of upper gastrointestinal (GI) barium studies. Subsequently, in 1984, Gustafsson describe the hypotonic duodenography combined with simultaneous SMA arteriography a more accurate investigation superior to barium study. Diagnosis is usually challenging and requires a high index of suspicion due to the nonspecific presentations and its rarity, while computerised tomography (CT) is considered a gold standard for diagnosis.[1]

We describe the rare case of SMA syndrome in pediatric age group manifested as chronic duodenitis.

CASE PRESENTATION

A healthy 11-year-old boy with recurrent epigastric and umbilical pain, nausea, aggravated by food intake and constipation (Bristol Stool Form Scale 1). Tolerating orally and active. Clinical examination revealed soft, non-tender abdomen and palpable fecaloma at left iliac fossa. He was initially treated with proton pump inhibitors and laxative, with a 2kg weight drop at two weeks follow-up. He was able to tolerate solid and liquid in moderate amount. There was subsequent worsening symptoms included non-bilious vomiting, intolerance to oral intake, and weight fluctuation (27kg to 28kg). BMI: 11.7kg/m^2. Therefore, he was admitted for close observation due to mild tenderness in epigastric and umbilical regions. No mass felt on clinical examination.

His laboratory investigation was normal with an albumin of 40g/L. Esophagogastroduodenoscopy (OGDS) revealed mild gastritis with extensive duodenitis, presence of bile reflux. Maximum anti reflux medication was initiated but no improvement was noted. CT angiogram showed no dilatation of stomach and proximal duodenum. Aortomesenteric angle was 18.3 degrees with aaortomesenteric distance of 5.3mm, suggestive of Superior Mesenteric Artery (SMA) Syndrome. Nasojejunal tube was inserted with guided by OGDS for enteral feeding in hoping weight gaining. After six weeks of nasojejunal tube feeding, there has been no significant weight gain, necessitating the addition of parenteral feeding. Furthermore, due to the previous tube being noted as malposition, a gastrojejunostomy tube was inserted. The child exhibited good tolerance to enteral feeding and a successful weight gain has been attained through a blend of enteral and parenteral nutrition.

DISCUSSION

This case report scrutinized the diagnostic challenge of a pediatric patient suffering from chronic abdominal pain attributed to SMA syndrome. SMA syndrome is characterised by "sandwich" of the transverse part of the duodenum as it traverses the space between the SMA and abdominal aorta due to reduced aorta-mesenteric angle. The name "sandwich" refers to the anatomical situation where the duodenum becomes trapped between these two structures. It is hypothesized that loss of the retroperitoneal fat pad and connective tissue narrows the aortomesenteric angle, creating a mechanical obstruction of the duodenum.

In general population, predisposing factors for acute angulation between the SMA and the aorta as shown in Table I. In pediatric age group, SMA syndrome is frequently associated with low BMI, weight loss, rapid linear growth

This article was accepted: 09 September 2024
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Table I: Predisposing factors in acute angulation in SMA syndrome

Intrinsic cause	Extrinsic cause
i. Physiology of severe weight loss causing retroperitoneal fat depletion. ii. Anatomical variant such as a high insertion of the Treitz ligament. iii. Surgical alterations cause mesentric tension and compression. (i.e spine surgery or ileoanal pouch anastomosis)	i. External compression by belts or body spica casts ii. Mechanical force from intra-abdominal tumor microenviroment push against the SMA, causing the acute angle.

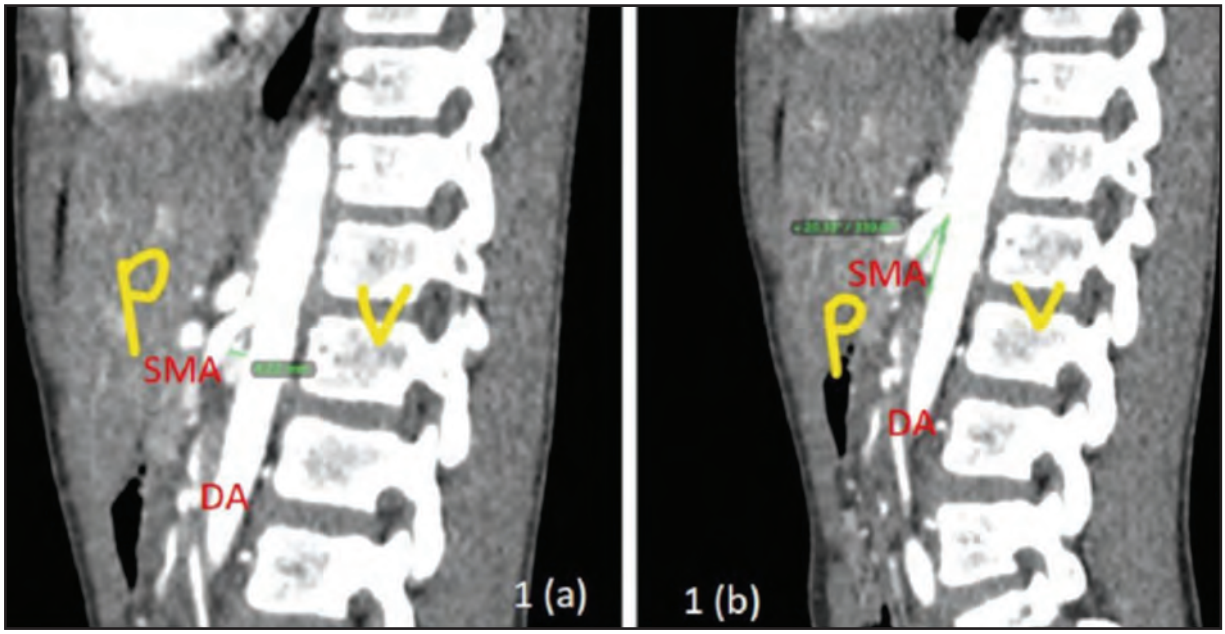

Fig. 1: Sagittal view of CT angiogram. (a) shows aortomesenteric distance of 4.22mm. (b) shows aortomesenteric angle of 20.33 degree. P = Peritoneum; V = Vertebrae; SMA: Superior Mesentric Artery; DA = Descending Aorta

spurt without weight gain are well-known risk factors of SMA syndrome. Retrospective studies conducted in western and eastern part of the world show similar figures of risk factors that develop SMA syndrome in pediatric age groups. The most common risk factor related to the development of SMA syndrome is low BMI and weight loss. Both studies also had a similar opinion that weight loss or low BMI is not a prerequisite for the development of SMA syndrome as half of study population show no recorded weight loss.[1-2] Despite having a low BMI, the patient has not experienced significant weight loss due to their active lifestyle and excellent academic performance.

A study by Ozbulbul et al. developed a theory that correlated SMA angle and visceral fat.[3] The research found that BMI correlated more strongly with subcutaneous fat than with visceral fat, and that the distance between the aorta and the SMA had a stronger correlation with visceral fat area than with BMI. In early puberty, a rapid linear growth spurt without weight gain can be a contributing factor to the manifestation of SMA Syndrome in children. SMA syndrome gets aggravated alongside malnourishment which substantially reduces the retroperitoneal fatty cushion support. Shin MS et al have shown that three out of four children with SMA syndrome, who experienced growth spurts without appropriate weight gain, responded to medical treatment.[2] Rapid linear growth without weight gain can result in an elongated mesentery as well as loss of the mesenteric root fat pad, which explains why SMAS occurred in our four patients during their growth spurt.

Early puberty is a crucial stage in children's development, as boys undergo significant changes, including increased muscle mass and reduced body fat, leading to thinning of the aortomesenteric space. This strongly supports the theory of puberty-related development during middle childhood or early adolescence. This case supports previous findings that weight loss is not necessary for pediatric SMA syndrome development. The rare presentation of SMA syndrome in children without weight loss may be linked to inadequate weight gain relative to height growth, leading to decreased visceral fat and predisposition to SMA syndrome.[3-4] We postulate that, two exacerbating factors for SMA syndrome in the patient were rapid linear growth spurt without weight gain and duodenitis symptoms. SMA syndrome is typically a diagnosis of exclusion, considered after extensive evaluation. Patients may present with persistent or intermittent gastrointestinal tract obstructive symptoms, requiring a thorough examination.

Patients with SMA syndrome may experience various upper gastrointestinal (UGI) symptoms like vomiting, nausea, epigastric pain, early fullness, and postprandial discomfort. The presentation can be acute or chronic, leading to non-specific symptoms and delaying diagnosis. In a Korean study from 2003 to 2013, the duration of symptoms before diagnosis ranged from 1 to 730 days (median 68 days), and the age at diagnosis ranged from 8.5 to 16.2 years (median 11.9 years).[2] The wide range of symptom manifestation duration for SMA syndrome is likely due to its non-specific symptoms, leading to many patients receiving OGDS for common UGI symptoms. While OGDS helps differentiate UGI diseases, it cannot diagnose SMA syndrome. A study by Kim JY et al.in 2021 suggests that endoscopy to the third part of the duodenum provides clues for determining the need for SMAS evaluation.[4] The exam should note three signs: [1] vertical or oblique narrowing of the third part of the duodenum during air insufflation for at least 15 seconds, [2]

marked dilation of the first and second part of the duodenum, and [3] presence of a bile lake in the stomach. Zhang R et al. hypothesized that SMA syndrome leads to weak acid reflux, causing reflux symptoms and duodenitis due to compression of the duodenum.[5] Patients with SMA syndrome are thin, may suffer from gastroptosis and delayed gastric emptying, leading to increased intragastric pressure and bile reflux duodenitis. After months of anti-reflux medication, red-flag symptoms worsened, leading to persistent post-meal vomiting due to oral intolerance.

Evolution of radiological techniques from serial upper GI studies to non-invasive CT scan, now considered the gold standard for diagnosing SMA Syndrome. This advancement allows for earlier detection and prompt treatment initiation. Shin MS et al. studies in 2013 and 2021 showed consistent cutoff values for diagnosis in adults (SMA angle <22°-25°, SMA-aorta distance <8 mm).[5] However, no consensus exists for diagnosing children, as pediatric cases can have varying cutoff values. This study also proposes the utilization of aorto-mesenteric distance instead of the aorto-mesenteric angle, given the varying anatomy of the superior mesenteric artery (SMA) and the absence of consensus on angle measurement.[2]

The treatment of SMA Syndrome involves bowel rest, fluid maintenance, electrolyte balance, nutritional support, and rehabilitation to improve weight, reduce intestinal obstruction symptoms, and address precipitating factors. Nutritional support is the major component of conservative treatment for aortomesenteric angle compression. This can be done with enteral nutrition that includes taking frequent small meals of nutritious liquid and lying on the left side or prone after eating. Anti-reflux medicines with a prokinetic can help further alleviate symptoms. Naso-jejunal feeding is an enteral feeding to provide nutritional support, and parentral feeding is an option when enteral feedings are not tolerated.[1-2,5] Shin et al. achieved a remarkable outcome in conservative treatment of SMA syndrome, with 83.3% of patients showing weight gain after treatment.[2] Such treatment should be instituted for at least six weeks before considering surgical intervention. Successful surgical therapies have included duodenojejunostomy, gastrojejunostomy, or resection of the Ligament of Treitz or Strong's procedure.

CONCLUSION

SMA syndrome is a rare condition which presents as chronic abdominal pain in children. To properly diagnose this challenge, clinicians should look into OGDS and contrast-enhanced CT. This case study has revealed cases where growth spurts and duodenitis can serve as catalysts for the onset of SMA Syndrome, and as such, it should be taken into consideration when approached with recurring abdominal complaints. In recent times, the medical approach to treatment has made significant progress, favoring nutritional treatments over the traditional reliance on surgical methods.

ACKNOWLEDGEMENT

The authors extend their heartfelt gratitude to the patient and his mother for their invaluable cooperation and consent in sharing this case report.

DECLARATION

The authors have no conflicts of interest to declare.

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