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Superior Mesenteric Artery Syndrome (Wilkie's Syndrome): Acute Severe Presentation in a 12 - Year - Old Girl and Role of Early Diagnosis

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Abstract: ***Superior mesenteric artery, SMA syndrome is a rare condition characterized by the compression of the third part of the duodenum between the abdominal aorta and the superior mesenteric artery. This case report discusses a 12 - year - old girl who presented with intermittent abdominal pain and vomiting, later diagnosed with SMA syndrome via radiological imaging. Initial conservative management, including nasogastric decompression and nutritional support, resolved her symptoms. SMA syndrome is more prevalent in females, typically in older children and teenagers, often associated with significant weight loss. Diagnostic imaging such as CECT and MRA confirms the condition. While conservative treatment can be effective, surgical intervention like duodenojejunostomy is recommended if symptoms persist to prevent serious complications.***

Keywords: Superior mesenteric artery syndrome, Wilkies syndrome, duodenal compression, paediatric abdominal pain, duodenojejunostomy

1. Introduction

Superior mesenteric artery syndrome is an atypical condition which was first described in 1861 by von Rokitansky (1). It is also known as Wilkie's syndrome which occurs due to compression of 3rd part of duodenum between abdominal Aorta posteriorly and superior mesenteric artery anteriorly (2). It is mostly due to loss of retroperitoneal fat that normally act as cushion around SMA. Patient usually presents with chronic intermittent abdominal pain, vomiting, nausea, early satiety and anorexia. A history of weight loss is usually seen but is not necessary (3). Diagnosis is basically by radiological imaging. initially conservative management is given to all patient but if failed then a surgical intervention should not be delayed.

2. Case Report

A 12 - year - old girl presented to the paediatric emergency with intermittent abdominal pain since 4months. it was insidious in onset, dull aching in nature, on and off type around the umbilicus, non - radiating to any other site with no aggravating and relieving factor. She also complaints of vomiting 3 days back which was acute in onset, yellowish in colour, non - projectile, non - bilious and contains food particles.4 months prior to admission, the child had visited a different hospital with similar abdominal pain which resolved after taking treatment on OPD basis.

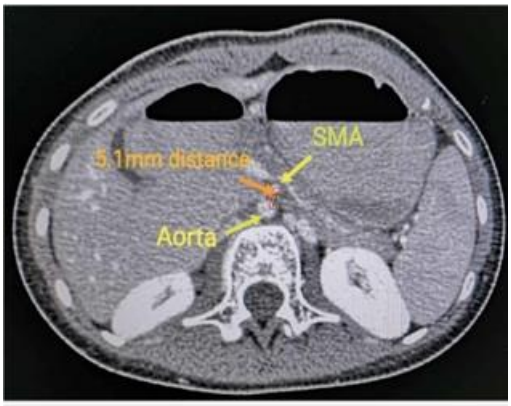

FIGURE 1: CT scan showing a reduced distance of 5.1 mm (normal 10-20 mm) between SMA and aorta, insinuating a diagnosis of superior mesenteric artery syndrome.
SMA: superior mesenteric artery

Her initial vitals signs were within normal limits. patient was of thin built with a height of 140cm and weight of 28.5 kg (BMI - 14.5). she mentioned a loss of 2 kg weight in the past 3 months. Central nervous system, cardiovascular and respiratory system was normal. On examination of gastrointestinal system, on inspection no abnormality was found. on palpation generalised tenderness was present but non guarding and abdominal masses were detected. Blood tests including CBC, Serum Electrolytes, RFT, CRP, ESR and urine routine were within normal limits. Abdominal ultrasound showed no obvious abnormality except slightly distended stomach. CECT was then performed and diagnosis of SMA syndrome was made. Her symptoms resolved after initiation of conservative management.

3. Discussion

SMA syndrome was first described by von Rokitansky in 1861 as an autopsy finding.1 Further, Wilkie published a detailed anatomical and pathophysiological description of

this rare cause of upper GI obstruction in 1927 (2). It is due to the obstruction of third part of duodenum between the SMA anteriorly and the aorta posteriorly (3). It is an uncommon entity with an incidence of about 0.013 - 0.3% in literature (4, 5). This syndrome is more common in females and occurs usually in older children and teenagers. It is generally associated with history of acute weight loss in majority of the patients leading to loss of fatty cushion around SMA secondary to various catabolic states like burns, eating disorders, chronic illness, neoplastic diseases and malabsorptive states (6, 7). However, other causes include an abnormally high insertion of the ligament of Treitz, abnormally low insertion of the SMA, following corrective spinal surgery or spinal trauma (7). Patients usually present with classic symptoms of upper GI obstruction like nausea, vomiting of bilious or partially digested food particles, early satiety, eructation and recurrent epigastric pain and fullness. The presentation can be chronic, intermittent or acute as result of partial or complete duodenal obstruction. These symptoms are generally relieved on lying prone or left lateral decubitus position and gets aggravated in supine position. The diagnosis of SMA syndrome is based on clinical symptoms and further substantiated on radiological findings. Upper GI dye study will reveal a dilated stomach and duodenum till second part and cut off at the third part of duodenum with no mucosal irregularity.

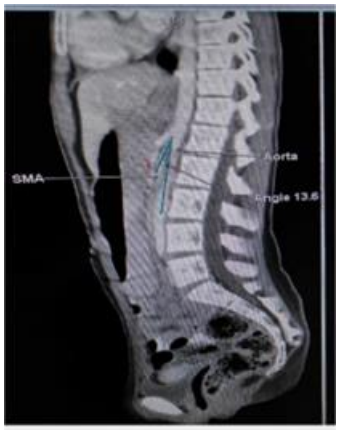

Figure 2: Showing decreased angle between SMA and Aorta

Contrast enhanced computed tomography (CECT) abdomen and Magnetic resonance angiography (MRA) are useful diagnostic investigations and provide better visualisation of a narrow a aortomesenteric angle between 6 - 250 (normal angle between 38 - 560) and reduced aortomesenteric distance between 2 - 8mm (normal 10 - 20mm) associated with this condition (6). Initial conservative management includes nasogastric decompression, intravenous fluid and electrolyte correction, promotility agents like metoclopramide, prone or left lateral position, surgically inserted nasojejunal feeding tube, or peripherally inserted central catheter for total parenteral nutrition administration should be given as a trial (4). Some patients respond to this conservative management and their symptoms gradually improves after weight restoration. However, in patients with severe symptoms and not responding to initial conservative management as in our case surgical intervention should be planned (8). Duodenojejunostomy, the most common operation for SMA syndrome, was first proposed by Blood good in 1907 (5). Other surgical options include gastrojejunostomy, Roux en Y duodenojejunostomy, anterior transposition of the third part of duodenum and division of the ligament of Treitz with mobilisation of the duodenum (6, 7, 8). The use of laparoscopic surgery in mobilisation of duodenum and lysis of the ligament of Treitz has also been reported (9, 10). Surgery in SMA syndrome has an advantage over conservative management in severe cases as it gives immediate and complete relief of the symptoms and allows early weight gain in these patients. It also reduces the chances of potential delayed complications viz. electrolyte imbalances, aspiration pneumonia, catabolic wasting, acute gastric perforation and peritonitis in chronic recurrent cases.

4. Conclusion

Superior mesenteric artery syndrome is a rare condition that can present in an emergency with acute upper gastrointestinal obstruction. It usually presents in older children with history of significant weight loss in majority of the patients. A trial of conservative management should be given to all patients. However, if the initial conservative treatment fails, one should not delay surgical intervention so as to prevent further fatal complications. Duodenojejunostomy is the procedure of choice and is highly effective in managing acute severe form of SMA syndrome.

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