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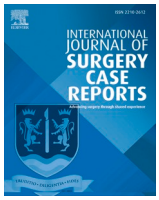

Case report

Superior mesenteric artery syndrome in a 16-year-old girl patient: A rare case report from Syria

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ABSTRACT

Introduction: Superior mesenteric artery (SMA) syndrome is a rare condition with a significant decrease in the angle between the SMA and the abdominal aorta.
Presentation of case: A 16-year-old girl presented with abdominal pain, vomiting, weight loss, and loss of appetite. Family history included allergies to contrast media. Clinical examination revealed a distended abdomen with a severely dilated stomach. Imaging showed narrowing in the duodenum. Upper endoscopy confirmed the diagnosis, and gastrojejunostomy was performed successfully. Patient recovered well post-surgery and was discharged after two weeks with resolved symptoms.
Discussion: SMA syndrome is a rare and serious condition, more common in women, young adults, and teenagers. Mental health support is crucial in managing SMA syndrome patients. Treatment involves CT scans for diagnosis, and options include conservative care or surgery, with success rates varying in studies.
Conclusion: Early diagnosis and intervention are crucial in managing superior mesenteric artery syndrome, as prompt treatment can significantly improve patient outcomes and prevent life-threatening complications. This case contributes to the literature by emphasizing the importance of considering SMA syndrome in differential diagnoses, particularly in young patients with non-specific gastrointestinal symptoms, thereby enhancing awareness and understanding of this rare condition.

1. Introduction

Superior mesenteric artery (SMA) syndrome is a rare condition first described in 1842 by von Rokitansky, and further researched by Wilkie in 1927, expanding knowledge about this rare entity [1]. The main mechanism behind this syndrome is a significant decrease in the angle between the superior mesenteric artery and the abdominal aorta. This reduced angle, typically less than 22°–28°, compared to the normal range of 38°–56°, leads to compression of the third segment of the duodenum, resulting in obstruction of the duodenal lumen [2]. Over 400 cases have been reported in medical literature, with an estimated prevalence ranging from 0.1 % to 0.3 % [3]. Constipation, vomiting, abdominal bloating, and pain are among the key accompanying symptoms of this rare syndrome [4]. SMA syndrome can lead to serious complications such as shock, gastric perforation, malnutrition, and even death, making early diagnosis and treatment essential for saving the patient's life [5]. Although specific recent advancements in diagnosis or treatment impacting outcomes have not been well-documented, the importance of early detection and intervention remains paramount. SCARE 2023 criteria have been followed in reporting this work [6].

2. Case presentation

A 16-year-old girl was admitted to the hospital with a chief complaint of abdominal pain, vomiting, weight loss, and loss of appetite. She was a non-smoker and non-alcoholic. The patient had no previous medical, surgical, or medication history. However, in the family history, it was mentioned that her parents had an allergy to the contrast media used in CT scans. Additionally, she had a sister who died due to contrast media. Her vital signs were: pulse 80 beats per minute, blood pressure

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120/80 mmHg, temperature 37C°, and respiratory rate 16 breaths per minute. The Glasgow Coma Scale (GCS) was 15/15 and the Morse Fall Scale (MFS) was over 44.

Over the past three months, the patient had experienced a steady weight loss, resulting in a current weight of 36 kg. Despite efforts to gain weight, she had been unable to do so. **On clinical examination,** the abdomen was distended with fullness in the epigastrium, and the rest of the organs examined were normal.

Laboratory investigations showed WBCs 8.4 × 10^9/L, RBCs 3.03 × 10^{12}/L, hemoglobin 15.3 g/dL, hematocrit 47.3 %, prothrombin time 10.8 s, and CRP was normal (see Table 1).

An ultrasound of the abdomen and pelvis was performed, which revealed a severely dilated stomach that was filled with fluid and showed several distended intestinal loops filled with fluid, some of which showed some peristaltic movements and others did not (see Fig. 1). The liver, spleen, gallbladder, pancreas, kidneys, and bladder were normal.

A nasogastric (NG) tube was inserted, followed by a **computed tomography (CT)** scan of the abdomen and pelvis was conducted with oral contrast (non-contrast CT scan via intravenous injection due to the patient's family history). The scan revealed gastric distension with food residue and relative narrowing in the third part of the duodenum behind the superior mesenteric vessels (see Fig. 2). The aortomesenteric distance was measured at 4 mm, and the superior mesenteric artery angle was 15°. The doctor decided to perform an upper endoscopy, which showed normal esophagus and stomach, but severe narrowing was observed after the second portion of the duodenum, with normal mucosa (see Fig. 3). Given the suspicion of superior mesenteric artery syndrome based on these findings, the decision was made to proceed with surgical intervention on the same day. Despite the patient's concerns, informed written consent was obtained to perform a gastrojejunostomy. A midline abdominal incision was made, and the omentum was dissected to access the stomach and jejunum. The greater curvature of the stomach was transected, and a gastrojejunostomy was created using a hand-sewn continuous running suture. The procedure was successful with no complications. Nutrition was resumed on the third day post-surgery. The patient was monitored until stable and discharged after two weeks of hospitalization. She regained her appetite, and all previous painful symptoms disappeared after two months of follow-up.

3. Discussion

SMA syndrome is a rare syndrome that can be life-threatening and falls under various names, including chronic duodenal ileus, Wilkie's syndrome, or superior mesenteric artery syndrome [7]. SMA syndrome is more common in women than men at a ratio of 3:2, and it occurs more frequently in young adults and teenagers, which aligns with our case involving a 16-year-old girl [8]. It is characterized by the obstruction of the stomach outlet due to external compression in the third part of the duodenum by the superior mesenteric artery. The causes of this syndrome can be congenital, such as a shortened aortomesenteric distance or anomalies in the superior mesenteric artery, or acquired. Other studies have suggested a close relationship between SMA syndrome and high duodenal insertion in Treitz's ligament and trauma and spinal orthotics [9]. Patients may present with vague symptoms, making the diagnosis challenging, with key symptoms including severe epigastric pain, abdominal distension, vomiting, and weight loss [1]. Contrary to most studies, Mauney et al. reported contrasting results in their study involving 8 patients with SMA syndrome, suggesting that weight loss may not necessarily be significant in this syndrome [10]. On the other hand, several studies have emphasized the importance of focusing on the mental health of patients with this syndrome, as it is closely associated with various psychological disorders such as anxiety and depression, necessitating both physical and psychological follow-up during the recovery period [11]. Cases have been reported where SMA syndrome is accompanied by Nutcracker syndrome, leading to compression of the left renal vein and the appearance of various kidney-related symptoms such as hematuria, proteinuria, and left flank pain [12]. In some autopsies, hypotheses have been proposed regarding the mechanisms of sudden death caused by the syndrome, including volvulus, increased abdominal pressure, severe hypokalemia, and irregular heartbeats, in addition to pulmonary complications [1]. Computed tomography is considered the cornerstone for diagnosing SMA syndrome, and three diagnostic criteria must be met: 1) aortomesenteric distance less than 8 mm, 2) duodenal obstruction, and 3) superior mesenteric artery angle less than 25 degrees [13]. Contrast-induced hypersensitivity reactions, including both immediate and delayed types, pose significant diagnostic challenges, particularly in distinguishing between allergic-like and physiologic responses. The prevalence of skin and mucous involvement, such as erythema and urticaria, in up to 70 % of patients with contrast reactions complicates the accurate diagnosis and management of underlying conditions [14]. Our patient has a family history of a fatal reaction to contrast media, which further complicated the situation and necessitated various investigative approaches. This concerning family history prompted the medical team to take extra precautionary measures. As for treatment, conservative management is the preferred option for managing the syndrome, while surgical treatment is the preferred option for newborns [15]. One study showed that conservative management achieved success rates ranging from 14 to 83 %, while surgical treatment was successful in 90 % of cases [13,16]. Conservative treatment options include nasojejunal tube feeding, total parenteral nutrition, or oral intake of high-calorie fluids [17]. In our case, surgical intervention was performed through a gastrojejunostomy, while other surgical options generally include duodenojejunostomy, Treitz's ligament division, or vascular reconstruction [18]. In a previous case, surgical intervention was chosen after one month of unsuccessful conservative treatment, aligning with literature that suggests surgery is more successful in cases unresponsive to conservative management. The procedure involved performing a gastrojejunostomy using neodymium magnets to create the anastomosis, which was confirmed to be successful after 10 days [19]. In another similar case, an 88-year-old male with intractable vomiting and significant weight loss underwent successful surgical intervention through gastrojejunostomy, highlighting the effectiveness of surgical treatment in severe cases [20].

Table 1
Laboratory tests upon admission.

Investigations	At presentation	Normal range
White Blood Cells (WBCs)	8.4	4.5–13.0 × 10^9/L
Red Blood Cells (RBCs)	3.03	4.5_5.5 × 10^{12}/L
Hemoglobin (Hb)	15.3	13.0_16.0 g/dL
Hematocrit (Ht)	47.3	38.8_50.0 %
PT	10.8	9–13 s

4. Conclusion

This case of SMA syndrome in a 16-year-old girl highlights the diagnostic challenges and management strategies in the presence of a family history of contrast media allergy. The successful surgical intervention underscores the importance of considering alternative imaging techniques and individualized treatment plans in similar cases. Future research should focus on exploring the psychological impact of SMA syndrome on patients, aiming to improve both physical and mental health outcomes.

Informed consent

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

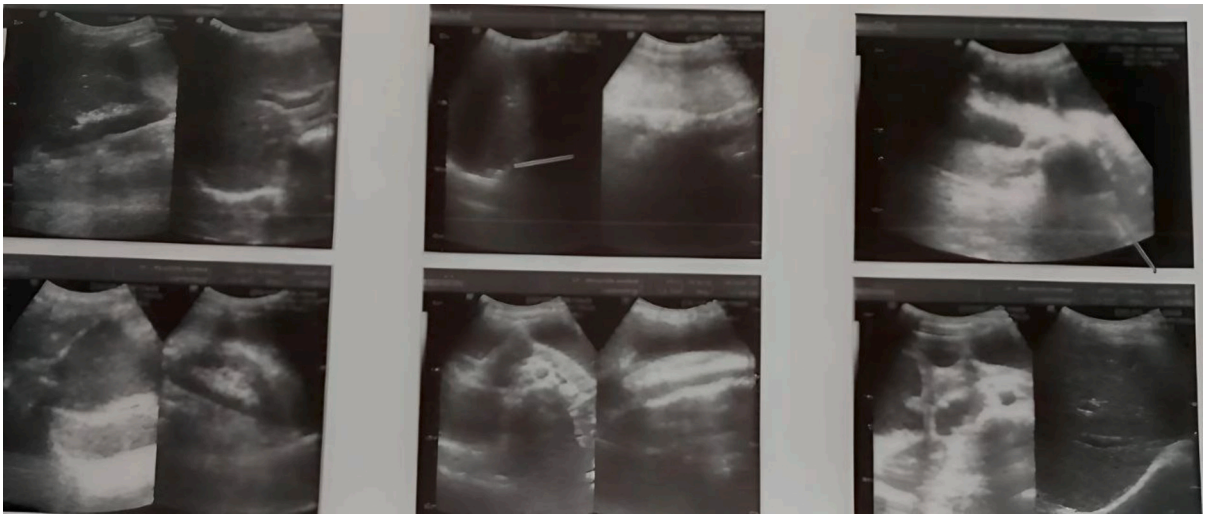

Fig. 1. Abdominal ultrasound imaging of a 16-year-old girl shows a greatly distended and fluid-filled stomach, while the liver, spleen, pancreas, gallbladder, kidneys, and bladder appear normal.

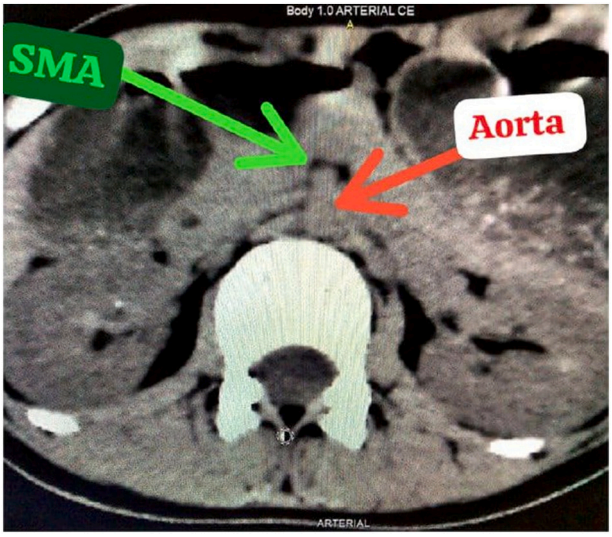

Fig. 2. CT of the abdomen revealed compression of third part of the duodenum between superior mesentery artery and aorta. The red arrow indicates the aorta, while the green arrow indicates the superior mesenteric artery.

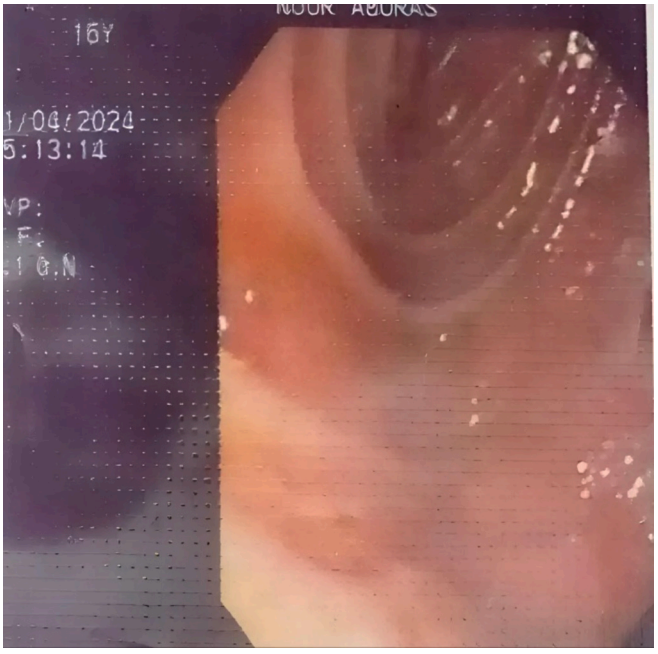

Fig. 3. Upper gastrointestinal endoscopy shows very severe stricture after the second segment of the duodenum.

Ethical approval

Ethical approval was provided by the Ethical Committee NAC of Aleppo University.

Guarantor

Dr. Hamdah Hanifa.

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Author contribution

HH, HA, LM, SK, OB, have participated in writing the manuscript, and reviewing the literature. Hamdah Hanifa critically and linguistically revised the manuscript. Mamdouh Alkhaled conceived and supervised the conduct of the study.

All authors read and approved the final manuscript.

Conflict of interest statement

The authors declare that they have no competing interests.

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Data availability

No datasets were generated or analysed during the current study.

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