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Case report

A case report of a 13-year-old girl diagnosed with superior mesenteric artery syndrome after undergoing spine correction with posterior fusion for rapidly progressed juvenile idiopathic scoliosis

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ABSTRACT

Introduction: Superior mesenteric artery (SMA) syndrome (SMAS) is a relatively rare disease that the etiology is closely related to the anatomy and the topography of the duodenum, aorta, and SMA.
Aim: To present and analyze the case of a patient who was diagnosed and treated for SMAS after scoliosis surgery.
Case study: A 13-year-old girl was admitted to our department for the surgical treatment of juvenile idiopathic scoliosis, with a Cobb angle of 120° in the main curvature. Postoperatively, we obtained the expected correction of 50%. Interestingly, after the treatment, BMI changed from 19 to 16, which assigned the patient to the underweight, starvation and emaciation group.
Results and discussion: On the 5th postoperative day, the patient's condition deteriorated. She suffered from abdominal pain, nausea and vomiting. We diagnosed SMAS. After conservative treatment the patient's condition improves and she was discharged from the hospital on the 16th postoperative day in a good general condition after the complete resolution of SMAS symptoms.
Conclusions: (1) SMAS can occur frequently in patients after surgical correction of the spine deformities. (2) At the curvatures of the order of 100°–120° or more, there is a significant change in the topography of the anatomical structures and their adaptation to a new

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Abbreviations: AIS, adolescent idiopathic scoliosis; LRV, left renal vein; PSF, posterior spinal fusion.

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position after surgery. (3) Special attention must be paid to young, lean patients, with BMI below 19, and the postoperative effect of an elongated axis of the spine. (4) Even if SMAS occurs, in most cases it can and should be treated conservatively.

1. Introduction

Superior mesenteric artery (SMA) syndrome (SMAS) is a relatively rare disease that was first described by Rokitansky in the 19th century, further analyzed by Wilkie in 1927, in 75 patients[1,2] and also called as Wilkie disease or cast syndrome.[3,4] The etiology is closely related to the anatomy and the topography of the duodenum, aorta, and SMA. SMAS is caused by the compression of the mesenteric vessels in the third part of the duodenum (Fig. 1). This is caused by the activation of extrinsic factors.[1,6] The main predisposing factors are weight loss and loss of the fat protection due to anorexia, severe trauma, and tumors, anatomical variants (the ligament of Treitz and SMA), and surgeries performed in the abdominal cavity.[7] The literature describes cases of this syndrome after surgical correction of the spine curvature, as well as after the treatment using a brace.[1,3,6] SMAS is a rare disease, with an incidence of less than 0.4%.[3] However, after surgical correction of the spine the incidence increases and is estimated to be 1.0%–4.7%.[1,8,9] It is a disease that often affects women more than men, in a ratio of 3:2,[10] and some authors[11,12] pay attention to a rather high number of deaths (33%), as described in the literature. There have been near 400 case reports in the English language literature since 1980.[1,4,13] These figures may not necessarily reflect the actual number of cases due to the difficulties in correct diagnosis.[2]

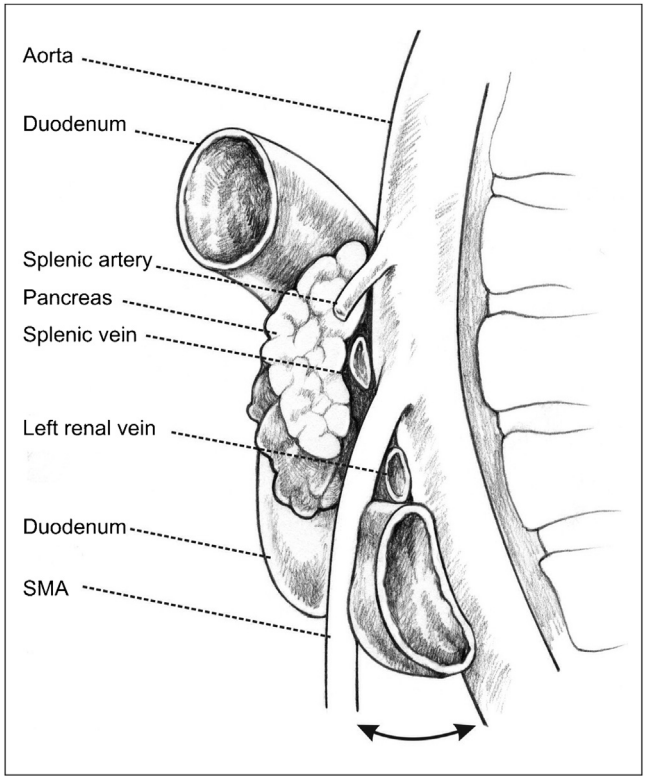

Fig. 1 – The anatomy and topography of the duodenum, abdominal aorta and superior mesenteric artery (the AP view), the SMAS pathomechanism (the LAT view), the compression of the duodenum
***Source*: Adopted from Lam et al.[5]**

2. Aim

We intend to present and analyze the case of a 13-year-old girl, with a significant deformity of the spine. After surgical correction of curvature, the girl was diagnosed and treated for SMAS.

3. Case report

A 13-year-old girl was admitted to our Department of Pediatrics, Orthopedics and Spinal Surgery for the surgical treatment of juvenile idiopathic scoliosis, which had rapidly progressed (above 15° per year).[14,15] She was previously treated exclusively with rehabilitation and a corset, without a satisfactory result. Before treating the patient with braces, X-ray, MRI, and CT examinations of the spine were carried out to rule out other pathologies.

On admission to the department, the patient's general condition was good. A clinical examination found idiopathic thoracolumbar scoliosis, with a Cobb angle of 120° in the main curvature. The Risser test was 3. Her height, body weight, and BMI before the surgery were 153 cm, 44.5 kg, and 19, respectively. The patient did not suffer from any other illnesses and was not on any medications. There was no family history of idiopathic scoliosis. Preoperative radiographs of the spine are shown in Figs. 2 and 3.

Due to the advanced spinal deformity, we discussed possible variants of the treatment with the patient and her parents, paying special attention to the potential complications that can occur with such a large curvature correction. We considered a multi-stage treatment including anterior release, cranial halo traction and posterior correction with fusion. Other proposed options covered even vertebral column resection (VCR).[16–18] After analyzing all the 'pros' and 'cons,' the parents opted for a one-stage treatment. Categorically they did not consent to the use of halo and VCR. Therefore, we planned a one-stage procedure of correction and posterior stabilization with multi-level Smith–Petersen osteotomy, informing that possible and safe correction will be from 40%

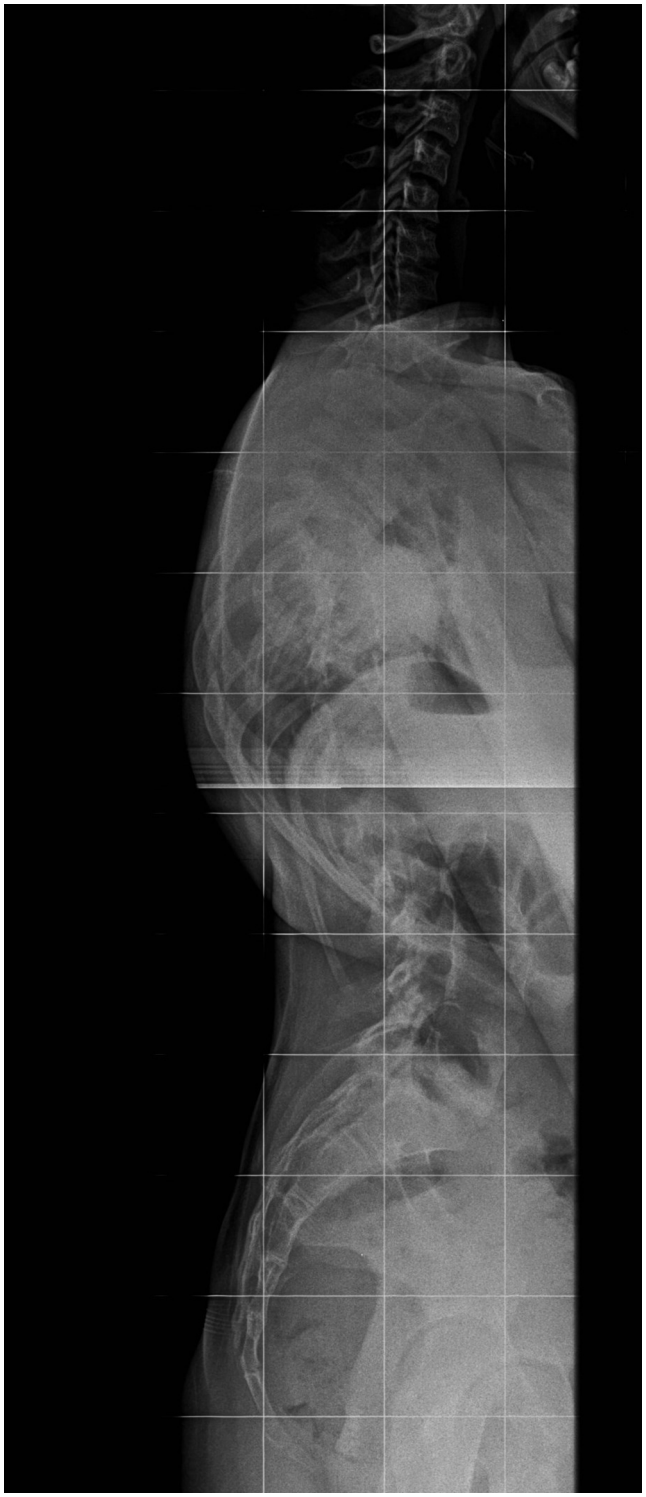

Fig. 2 – A preoperative X-ray image of the spine (AP view).

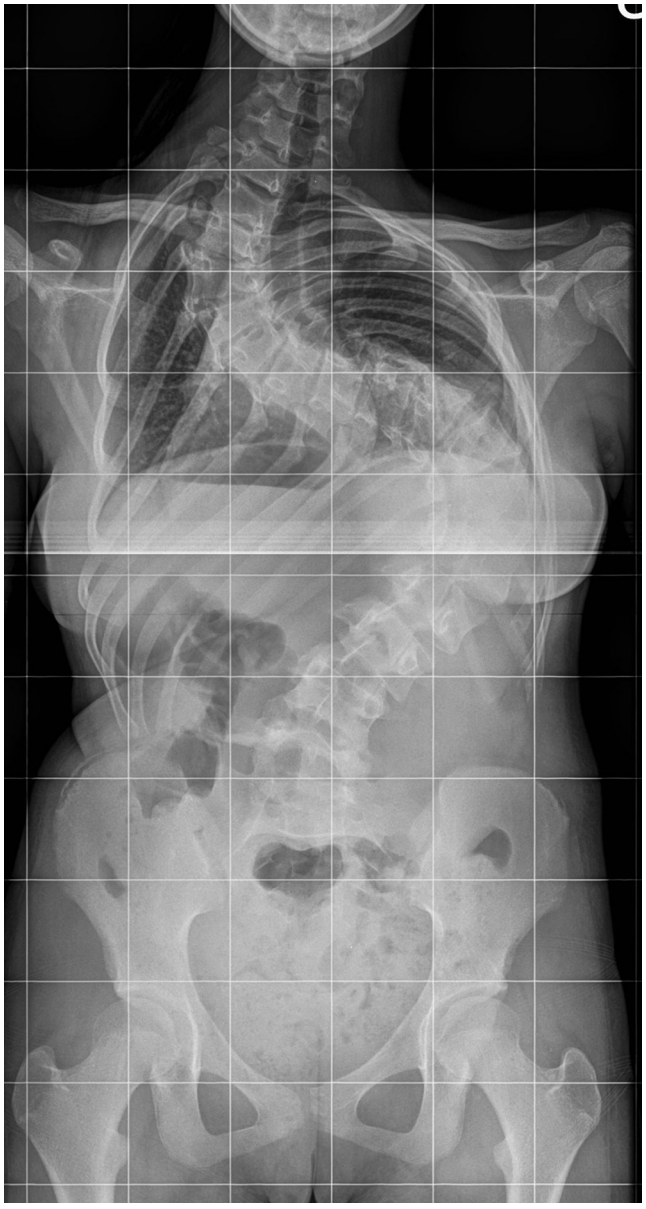

Fig. 3 – A preoperative X-ray image of the spine (LAT view).

to 60%. The parents and the patient accepted this treatment option and possible complications.

The procedure was performed under intraoperative neuromonitoring of the spinal cord. Postoperatively, we obtained the expected correction of 50% without any neurological deficits. The upper curvature was reduced to 57°. Postoperative radiographs are shown in Figs. 4 and 5. Both the patient and her parents were very satisfied with the result of the operation. The height of the patient after correction was 162 cm, BMI 16, and a postoperative weight of 42 kg. Interestingly, after the treatment, BMI changed from 19 to 16, which assigned the patient to the underweight, starvation and emaciation group.

4. Results

The general condition after correction of the deformity did not raise any objections and deviations in relation to other patients who underwent surgeries for idiopathic scoliosis. In the 1st two postoperative days, the patient required analgesics in the form of strong opioids. Thereafter, as per standard

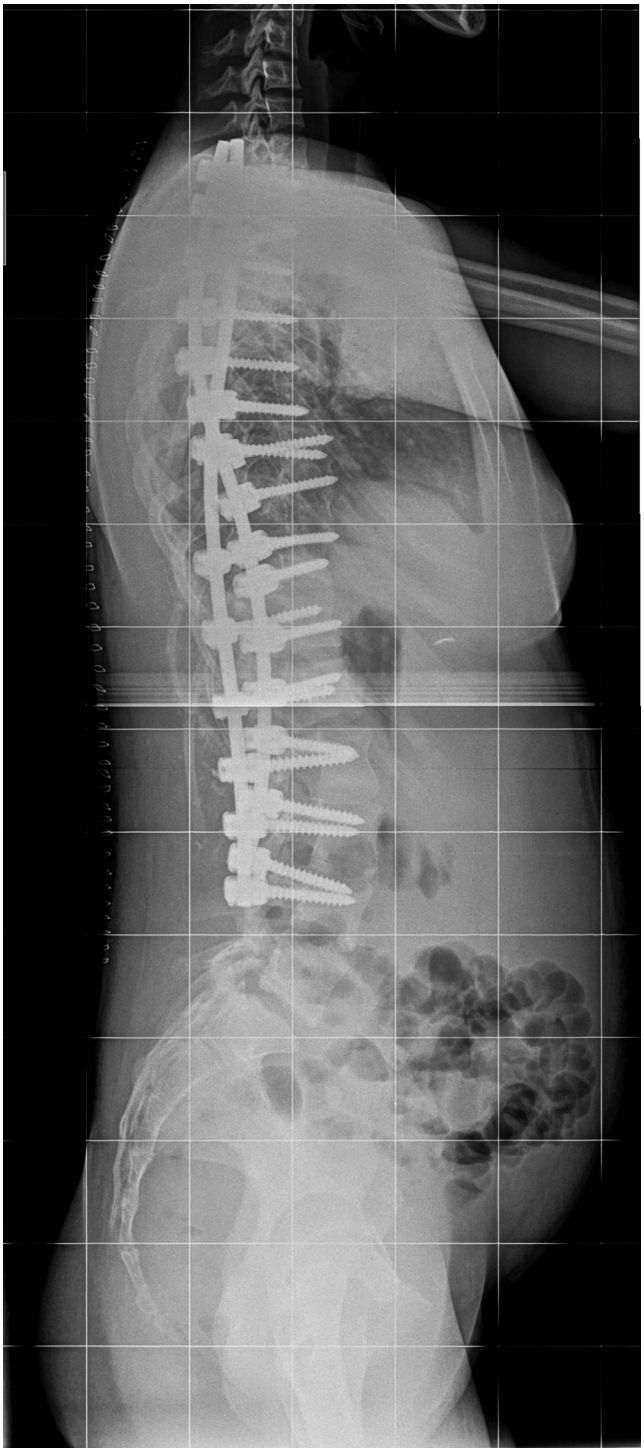

Fig. 4 – A postoperative X-ray image of the spine (AP view).

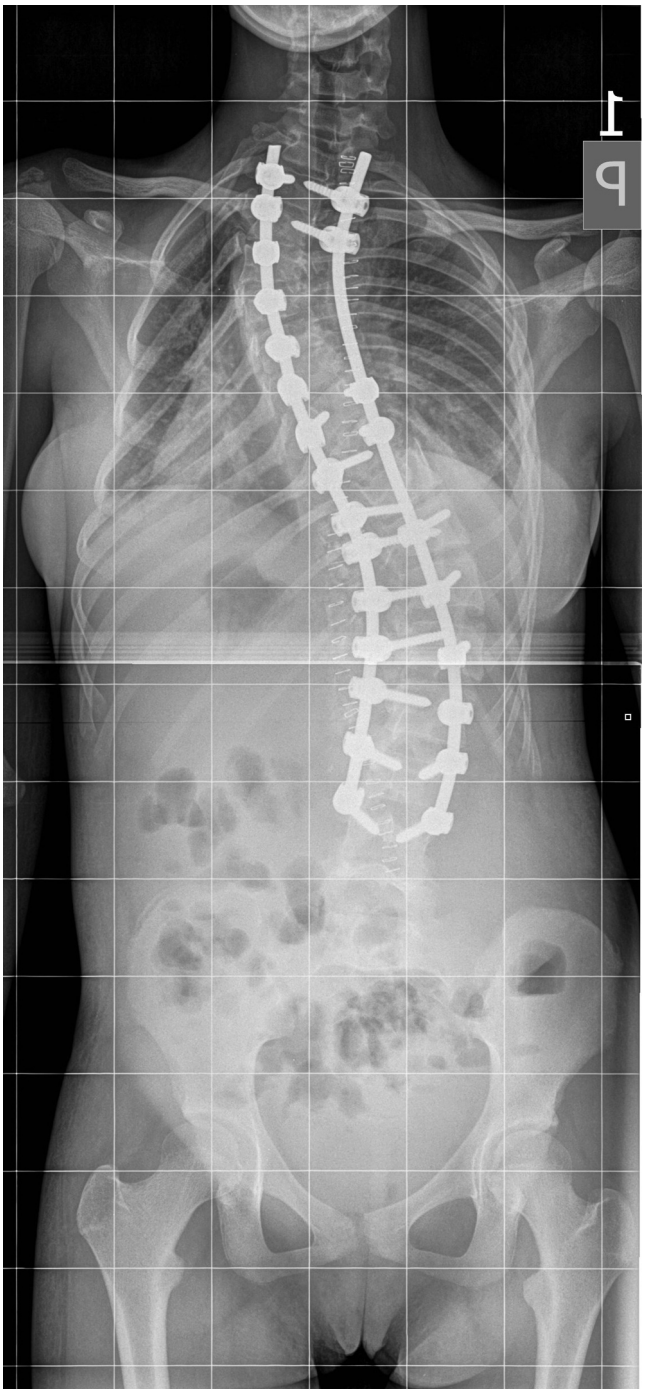

Fig. 5 – A postoperative X-ray image of the spine (LAT view).

treatment procedure, verticalization was carried out along with gradual walking. The patient had no gastrointestinal tract symptoms. On the 3rd postoperative day, she was taken off the narcotic drugs, drains were removed from the wound and an oral analgesic treatment was administered, according to the 'analgesic ladder.' On the 5th postoperative day, the patient's condition deteriorated. She suffered from abdominal pain, nausea and vomiting (7–8 times a day).

We excluded the most likely causes, i.e. negative effects of medications, an infection of the gastrointestinal tract, intestinal obstruction, appendicitis, and potential diseases of the urogenital system. The patient consulted with a surgeon, gynecologist, gastroenterologist, and a specialist in infectious diseases. Given the nature and the extent of the surgery, as well as exclusive gastrointestinal tract symptoms, we took into account SMAS and thromboembolic disease of the blood vessels of the abdomen. We performed an abdominal CT with

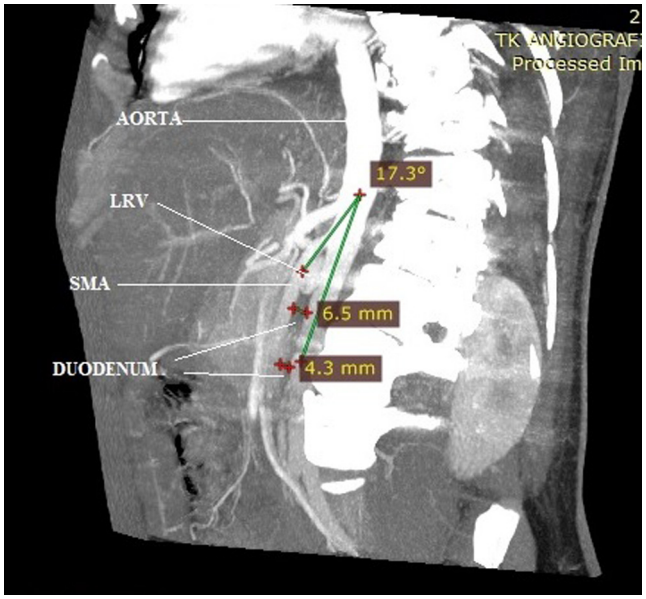

Fig. 6 – An image of abdominal CT scan with contrast, the angle of the SMA ramification from the aorta is approximately 17°, the level of the duodenum is of 4.3 mm to 6.5 mm between the SMA and the aorta. SMA – superior mesenteric artery; LRV – left renal vein.

contrast, which confirmed SMAS (the CT result along with marked structures and measurements are shown in Fig. 6). The angle of the SMA ramification from the aorta was 17.3° (normal range 38°–56°), and the distance between the SMA and the aorta at the level of the duodenum was from 4.3 mm to 6.5 mm (normal range 10–20 mm).

We used conservative treatment and parenteral nutrition for 5 days. On the 11th postoperative day, the patient's condition improves enough that she was able to take fluids and oral food. Initially, the patient received an easily digestible diet, controlled by a dietician. She returned to a normal nutrition on the 15th postoperative day. The patient was discharged from the hospital on the 16th postoperative day in a good general condition after the complete resolution of SMAS symptoms.

5. Discussion

SMAS most often occurs in young, slim, tall patients, with an asthenic body type.[10–12] BMI may also be a characteristic factor, which was seen in our patient.[2,13,19] However, under adverse conditions, it can occur in any age group, in both the sexes.[11,12] In most cases described in the literature, the presence of SMAS was associated with the anatomy and topography of the duodenum, aorta and SMA.[1,2,20] The location of the duodenum resembles a horseshoe where the raised bank faces the right side and the concave part embraces the head of the pancreas. The uppermost section of the duodenum is slightly expanded and is called the bulb (the first part of the duodenum). It is adjacent to the liver and the gallbladder. Then, the duodenum gradually narrows and is directed downwards (the descending part or the second part), embraces the head of the pancreas and then horizontally (the inferior part or the third part) crisscrosses the spine toward the mesenteric vessels. Thus, it forms an upper and lower folding. On the other hand, the duodeno-jejunum bend is directed slightly upward and moves into the jejunum (the ascending part or the fourth part). The third section of the duodenum extends in the vicinity of the abdominal aorta and the SMA, and lies exactly in front of the L1-L2 vertebrae.[7,21,22] When branching off the aorta, SMA moves forward and down at an angle of approximately 45° (normal range 38°–56°), with a normal structure and anatomical relations. Thus, when extrinsic factors act, the third part of the duodenum between the aorta, SMA and the spine is compressed. This causes partial or complete obstruction of the duodenum. The syndrome can also be caused by surgical complications associated with the impaired function of the peritoneum, duodenum, weakened abdominal wall and anorexia.[1,2,6,20,23] SMAS often accompanied by a spinal deformity treated by wearing a corset or after surgical correction of the spine.[1,2,6,8,20,23–25] The spatial relationships of the deformed spine changes during the three-dimensional correction of deformation, and thus it elongates in varying degrees in the sagittal plane.[24,25] In our case, the axis of the spine after correction increased by almost 10 cm. During derotation of the deformation, the blood vessels (aorta, SMA) tighten and stretch. A degree of the narrowing of the angle between the SMA and the aorta can cause acute or chronic clinical symptoms. The anatomical relations after the operation are shown on the CT scan (Fig. 6). It is therefore important to avoid the occurrence of SMAS at the planning stage. The parents were not been convinced to treatment in several stages especially a preoperative cranial traction, but halo traction allows for progressive curve correction during the preoperative period and can better protect the patient from several complications including the SMAS.[14–18]

SMAS is difficult to diagnose since it has no specific symptoms. It usually manifests itself through symptoms such as bloating, pain in the upper abdomen after a meal, nausea, vomiting, decreased appetite and weight loss.[6,23] Paradoxically, it may create the vicious circle mechanism and an aggravation of these symptoms.[1,2,6–9,20,23,26] Postoperative obstruction reveals a similar clinical picture as the SMA syndrome, so differentiating the latter from other diseases and making a correct diagnosis is difficult. General anesthesia, analgesics, acid–base and electrolyte balance, and potential damage to the visceral nerves can mask the postoperative obstruction.[1,2,6,20] These symptoms usually occur one day after surgery, and the clinical picture reveals itself three to five days later. A correct diagnosis is based mainly on the exclusion of other diseases and additional imaging tests. The best methods include the contrast examination of the upper gastrointestinal tract with barium and a CT scan after the administration of contrast medium.[9,19,21,27,28] In rare cases, it may be desirable to perform gastrointestinal endoscopy to eliminate pathologies inside the lumen of the gastrointestinal tract.[19,21,22,27–29] A Doppler ultrasound may also be used in diagnosis.[7,21,22,24,25,27–29] However, our center prefers an abdominal CT scan with contrast, as the most accurate diagnostic tool: angle of SMA

ramification from the aorta below 25° (normal range: 38°–56°), when measured in the sagittal plane; the distance between the SMA and the aorta at the level of the duodenum less than 8 mm (normal range 10–20 mm).

We can consider the fact and argue with the other authors[3,7,10–13,19,21,22,24,25,27–31] on whether in the case of our patient's scoliosis of 120° if we have used the SMAS diagnostic criteria (the value of the angle of ramification, the distance from the duodenum) which are adapted to healthy patients, with normal anatomy. As a result of the deformation, there is a significant change in the topography of the anatomical structures and their adaptation to the new situation.[26,32,33] Thus, we should know the value of the angle of the SMA ramification in the adapted position to make a proper correct diagnosis of SMAS. It would be useful to perform a CT scan in patients at risk before and after surgery in order to know the location and to evaluate the organs and vessels of the abdominal cavity. Alternatively, if any clinical signs occur, we ought to do another CT scan of the abdomen to assess the displacement of the structures after surgical correction of the spine as compared to the preoperative image. In our case, this angle was 17.3° with an undisturbed anatomy, which met the diagnostic criteria. By knowing the value of the angle of curvature in scoliosis, height and BMI before and after surgery, 'the extension of the backbone' by almost 10 cm resulting in the tightening of the blood vessels, and common clinical symptoms, we could diagnose SMAS, even though we did not take measurements before the surgery.

The conservative treatment of SMAS usually starts by removing causative factors such as a corset, if it was used. The treatment includes: intensive fluid therapy, replenishment of electrolyte deficiencies, anti-emetics, and if necessary the limitation of oral feeding and feeding through a nasogastric tube or even total parenteral nutrition.[1,2,6,20,23] In most cases, the complete recovery period is 7–14 days,[8–9,26] but the literature also report chronic conditions as well as those diagnosed 6 or more weeks after surgery.[12,30,34]

Surgical intervention is rarely necessary. It is used when the conservative treatment gives no effects in 4–6 weeks or when other pathology coexists, such as peptic ulcer, pancreatitis, etc. The surgery is reserved for patients with a chronic SMA disease process.[10,21,22,28,29] The most common treatments used in SMAS surgery include: anastomosis bypass and duodenal derotation.[1,2,10,20–22,28,29] The surgery is not required in complications after spinal surgery.[3,11–13,31,34]

6. Conclusions

1. SMAS can occur frequently in patients after surgical correction of the spine deformities, but atypical symptoms and drugs can mask the potential symptoms, it may be undiagnosed.
2. First, special attention must be paid to young, lean patients, with BMI below 19, and the postoperative effect of an elongated axis of the spine, which increases the patient height.
3. Even if SMAS occurs, in most cases it can and should be treated conservatively, but the mere diagnosis does not exclude the coexistence of other diseases, thus the entire clinical picture of the patient must be taken into account.
4. The problem of diagnosis in large deformations remains unsolved and at the curvatures of the order of 100°–120° or more, there is a significant change in the topography of the anatomical structures and their adaptation to a new position after surgery.

Conflict of interest

None declared.

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