Case report

An unusual cause of abdominal pain: Case report of a Superior Mesenteric Artery syndrome

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ABSTRACT

Introduction and importance: Superior Mesenteric Artery (SMA) syndrome is an occlusive vascular disease, mainly caused by a severe weight loss that leads to a reduction of the fatty cushion around the SMA. Postprandial abdominal pain, early satiety, vomits, weight loss and malnutrition are the main symptoms of SMA syndrome. Therapy consists of nutritional support, while mini-invasive surgery is indicated when conservative measures are unsuccessful.

Case presentation: A 39-years old caucasian woman reported the following symptoms: epigastric pain irradiated up to the right hypochondrium, late post-prandial vomiting and severe weight loss. During previous hospitalizations a gastroscopy, a colonoscopy, and radiographic study of food transit were performed, these exams showed a slow gastric emptying. We performed an abdomen CT scan discovering SMA syndrome typical features. A duodenjejunalostomy was performed in videolaparoscopy; the procedure was completed easily without complications.

Clinical discussion: Debilitating conditions with severe weight loss or anatomic abnormalities are the main causes of SMA syndrome. Symptoms are usually unclear and non-specific. The commonest SMA syndrome features are highlighted with contrast CT. This syndrome is often misunderstood and not considered in differential diagnosis of abdominal pain, determining a delay in diagnosis and further weight loss. An early diagnosis is useful to choose the best treatment of the case. For severe cases, surgery represents the best treatment, especially duodenjejunalostomy that is usually linked to rapid symptoms' resumptions without post-operative consequences.

Conclusion: SMA syndrome should be included in differential diagnosis of abdominal pain. Severe cases can be treated with mini-invasive surgery.

1. Introduction

Superior Mesenteric Artery (SMA) syndrome, also known as Wilkie’s syndrome, Cast syndrome or duodenal ileus, was first described by Von Rokitansky in 1842 and then better understood by Wilkie. In SMA syndrome the corner between aorta and superior mesenteric artery is too narrow and this condition determines the compression of the third part of duodenum [1].

Since SMA syndrome first description, approximately 400 cases have been reported [2], with a prevalence of approximately 0.0024–0.3% [3]. As this condition leads to an obstruction of the proximal small bowel, a delay in diagnosis is linked to notable morbidity and mortality [4]. Weight loss is both a symptom and one of the main causes of the disease. The main symptoms of this disease are postprandial abdominal pain, early satiety, vomits, weight loss and malnutrition [5]. The conservative management consists mainly of nutritional support [6]; however, surgical intervention is indicated for patients with chronic and refractory symptoms [7]. This paper reports the case of a young caucasian woman with referred abdominal pain for about two years and weight loss. This case report has been reported in line with the SCARE Criteria 2020 [8]. The diagnosis of SMA syndrome was made through many specialist consultations and radiological procedures. Because of the ineffectiveness of conservative treatments, minimally invasive surgery was proposed.

2. Case report

A 39-years old caucasian woman of 58 kg in weight and 159 cm in
height (without a noteworthy remote medical history) has been reporting epigastric pain irradiated up to the right hypochondrium and late post-prandial vomiting since December 2019. Due to these symptoms, the patient was hospitalized many times and different exams were performed, including gastroscopy and colonoscopy. However, she was discharged for functional gastropathy with home therapy based on levosulpiride.

Subsequent instrumental investigations, including radiographic examination with study of food transit (Fig. 1), showed a slow gastric emptying. During this period, the patient reported a weight loss of about 18 kg, reaching a BMI of 15.8. Acute intermittent porphyria, infectious aetiologies or other non-surgical causes were excluded.

Due to the persistence of her symptoms, the patient was admitted to our department to perform detailed diagnostic examinations, including CT scan of the abdomen (Fig. 2a-b). Meanwhile a conservative therapy with parenteral and enteral nutrition was started. The abdomen CT scan showed a gastric ptosis up to the small pelvis, a delayed emptying of the stomach, a dilation of the duodenum and an extremely acute angle between aorta and superior mesenteric artery of about 25°. The resulting diagnosis was of Superior Mesenteric Artery syndrome, for which a surgical treatment was proposed.

A duodenojunostomy was performed in videolaparoscopy. After the induction of pneumoperitoneum with a Veres needle, four trocars were placed: one in the sub-umbilical area (for camera), two 12 mm trocars in the right and left flank, and a 5 mm trocar in the sub-xiphoid area. It was evidenced a noteworthy dilation of the duodenum up to the third portion and gastric ptosis as well. The submesocolic portion of the duodenum was exposed and the third duodenal portion was anastomosed to the second jejunal loop with a stapler device (Fig. 3a-b).

The procedure was completed easily without complications. The post-operative course was regular, with resumption of nutrition in fourth postoperative day (delayed by the patient’s initial refusal due to the persistence of a subjective sensation of nausea). The pre-existing anemia (caused by the preoperative state of malnutrition) required transfusion therapy. The patient was treated and monitored until stabilization, delaying the discharge in twelfth p.o. day. During the post-operative outpatient checks, the patient reported disappearance of painful symptoms and post-prandial vomiting. After a 3-month follow-up, the patient regained 10 kg, reaching a BMI of 19.8.

3. Discussion

Superior Mesenteric Artery (SMA) syndrome is a rare life-threatening disorder. It consists of the reduction of aortomesenteric angle from normal 38–56° to 6–25°, and the decrease of aortomesenteric distance from normal 10–28 mm to 2–8 mm. This condition causes the loss of retroperitoneal fat, mesenteric fat, and lymphatic tissue, which normally determines a cushioning effect for the duodenum, protecting it from compression [9]. The incidence reaches the 0.1–0.3% and females aged between 10 and 40 years are more commonly affected [6].

The main cause of SMA syndrome is severe weight loss that leads to a depletion of the fatty cushion around the SMA. Etiological factors can be split into two categories: anatomic abnormality (both congenital and acquired) and debilitating condition causing a severe weight loss [10]. Congenital anatomic abnormalities include a high insertion of the angle of Treitz, or an atypical low insertion of the SMA [10]. Acquired anatomic abnormalities can arise following spinal trauma, after corrective spinal surgery, and after abdominal surgery due to tension of the small bowel mesentery. Conditions that cause severe weight loss are eating disorders (e.g. anorexia nervosa), burns or other catabolic states, and wasting conditions like malabsorptive states or neoplasia [10].

Symptoms are usually unclear and non-specific. The commonest symptoms are nausea, postprandial epigastric pain, bloating after meals, eructation, esophageal reflux, early satiety, anorexia, acute metabolic alkalosis, hypokalemia, and chronic intractable bilius vomiting that can induce severe dehydration. The patient is afraid to eat thus triggering a vicious cycle resulting in weight loss [11].

Typically, the first approach is abdominal x-ray, that usually shows a gastroduodenal distension. However, the best imaging method is the fluoroscopic one because it shows clearly not only gastroduodenal dilation and delayed gastroduodenal emptying, but also potential vertical imperfections of the third portion of the duodenum caused by vascular compression. Computed Tomography (CT) with contrast is used to evaluate aortomesenteric vasculature. A narrowed aortomesenteric angle, a decreased aortomesenteric distance, and gastroduodenal dilation with bowel calibre narrowing, are the commonest SMA syndrome features highlighted with contrast CT [12].

Usually, the first therapeutic approach is a conservative treatment made of correction of electrolyte imbalance, and nutritional support for weight regain. In acute cases a combined therapy with parenteral and enteral nutrition is necessary and successful [7].

Surgery is indicated when conservative measures are ineffective, especially for patients with a long history of progressive weight loss, and refractory duodenal dilation. There are many surgical options that can be performed in minimally invasive surgery, the most common are following described.

Strong’s Procedure consists of full mobilization of the third and fourth portions of the duodenum and the division of ligament of Treitz. After that, the jejunum is passed behind the superior mesenteric axis and it is positioned to the right of the vessels [13].

Even if a gastrojejunosotomy it could be performed, it is an unsuccessful procedure because it usually leads to a recurrence of obstruction symptoms and increase of alkaline reflux [14].

Duodenojejunostomy is the most common and successful surgical procedure. It consists in mobilizing a loop of jejunum (20 cm distal to the ligament of Treitz) and anastomosing it to the dilated second-third portion of the duodenum to decompress and bypass the obstructed area [15].

“This technique was introduced by Stanley in 1910, and then in 1998 Gersin KS and Heniford BT performed laparoscopic duodenojejunostomy for the first time. Today minimal invasive duodenojejunostomy (including robotic procedures) has a success rate of more than 90% and a complication rate of 7% [16]. The recent retrospective study of Jain...
et al. 2021, has shown the absence of intraoperative complications for 22 patients with SMA Syndromes treated with laparoscopic duodeno-jejunostomy. Only 5 patients (22.7%) presented post-operative complications treated with a conservative management. The mean hospital stay was about 7.27 ± 3.68 days, while preoperative optimization was of 3.14 days (1–7 days). During the follow-up all patients improved their symptoms, no one patient had further vomiting episodes and no one patient required re-admission for in-patient care [17].

The case reported in this paper is comparable to the trend reported Jain et al. 2021 analysis. The duodenojejunosotomy was performed easily, with no intra-operative complications and with a regular post-operative course. During the follow-up the patient reported disappearance of painful symptoms and post-prandial vomiting.

4. Conclusion

SMA syndrome is an occlusive vascular disease that is often misunderstood and not considered in differential diagnosis in case of acute abdominal pain. Radiological techniques available can easily provide a rapid diagnostic classification in order to establish prompt medical therapy.

If non-invasive therapy results inefficacious, mini-invasive surgery should be performed. Mini invasive surgery, especially duodenojejunosotomy, is usually linked to rapid symptoms resumptions with no post-operative consequences. Further studies are required to improve SMA syndrome diagnosis as well as to define standard therapies.

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Ethical approval

Ethical approval has been exempted by our institution because this is a case report and no new studies or new techniques were carried out.

Consent

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

Fig. 2. CT scan of the abdomen showing a narrowed aortomesenteric angle in sagittal plane (a) and gastroduodenal dilation with bowel calibre narrowing in axial plane (b).

Fig. 3. Videolaparoscopic procedure: evidence noteworthy dilation of the gastroduodenal tract (a); duodenojejunosotomy (b).
Referencing the checklist

This case report has been reported in line with the SCARE Criteria 2020 [8].

Author contribution

Roberto Cantella: Operated on the patient, drafting the manuscript, literature research.

Giuseppe Evola: drafting the manuscript, literature research.

Cristina Di Fidio: Operated on the patient, drafting the manuscript, literature research.

Marianna Iudica: drafting the manuscript, literature research.

Marco Patanè: drafting the manuscript, literature research.

Luigi Piazza: Operated on the patient, drafting the manuscript, literature research, Director of the Department.

Registration of research studies

Not applicable.

Guarantor

The guarantor for this case report is Roberto Cantella.

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Declaration of competing interest

All the authors certify that there is no conflict of interest regarding the material discussed in the manuscript.

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