CASE REPORT

Familial superior mesenteric artery syndrome

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SUMMARY

Superior mesenteric artery syndrome (SMAS) has an incidence of 0.1–0.3%. It is caused by a narrow angle between the aorta and the superior mesenteric artery, resulting in the clamping of the third portion of the duodenum. It is more common in females (2:1), with no racial differences. It is also more frequent in patients under 30 years and may have a congenital or acquired aetiology. There is only one case of the familial form described in the medical literature. We present a case of a 20-year-old woman, with recurrent episodes of epigastric discomfort and vomiting since she was 10 years old. After clinical suspicion, diagnosis was achieved after digestive endoscopy, upper gastrointestinal transit and abdominal MR. The patient was kept under parenteral nutrition for 17 days before surgery. She was discharged 8 days after surgery and the postoperative course was uneventful.

BACKGROUND

Superior mesenteric artery syndrome (SMAS) is a rare disease. There is only one case of the familial form of SMAS described in the medical literature.

Diagnosis is complex and requires a high level of suspicion.

CASE PRESENTATION

A 20-year-old woman, repeatedly diagnosed as having bulimia and anorexia, was referred by her gastroenterologist, to a surgery outpatient care unit due to recurrent episodes of epigastric discomfort and vomiting since she was 10 years old.

No abnormalities were detected on clinical examination besides a body mass index of 18.9 kg/m².

The patient's mother had undergone surgery for SMAS at the age of 16 years.

INVESTIGATIONS

Blood tests showed hypoalbuminaemia and a decrease in total protein count, with no other significant changes.

Following are the procedures performed and their findings:

- ▶ Upper endoscopy (July 2012): stasis oesophagitis with linear erosion of the lower third. Gastric distention with stasis content. Pylorus permeability. Deformed and enlarged bulb enabling the transition to D2.
- ▶ Upper gastrointestinal transit (August 2012): hypotonic stomach with delayed emptying. Contrast accumulation in a 10 m diameter structure in the right iliac fossa, corresponding to the dilated duodenum (figure 1).
- ▶ Abdominal MRI (October 2012): gastric and duodenal distension, the latter in the transition from

Figure 1 Upper gastrointestinal transit: hypotonic stomach with delayed emptying. Contrast accumulation in a 10 cm diameter structure in the right iliac fossa, corresponding to the dilated duodenum.

D3–D4. Duodenal stricture between D3–D4, posterior to the superior mesenteric artery. The angle formed between the superior mesenteric artery and the aorta was 13° (figures 2–4).

DIFFERENTIAL DIAGNOSIS

- ▶ Inflammatory bowel disease with duodenal stricture
- ▶ Gastric neoplasm (antrum)
- ▶ Duodenal or small intestine tumour
- ▶ Pancreatic neoplasm.

Figure 2 Abdominal MRI: gastric and duodenal distension, the latter in the transition from D3–D4.

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Figure 3 Abdominal MRI: duodenal stricture between D3–D4, posterior to the superior mesenteric artery.

TREATMENT

The patient was kept under parenteral nutrition for 17 days in order to optimise her nutritional status before proceeding to surgery. Intraoperatively, both the D3–D4 duodenal clamping by the mesenteric artery and the absence of fixation of the entire colon were observed (figure 5). An intestinal bypass was performed consisting of a Roux-en-Y in anterior position in relation to the superior mesenteric axis.

Figure 4 Abdominal MRI: sagittal view with dilated duodenum.

The patient was discharged 8 days after surgery and the post-operative course was uneventful.

OUTCOME AND FOLLOW-UP

The patient remains asymptomatic 1 year after surgery, with no nausea and no vomiting, and has had progressive weight gain.

DISCUSSION

SMAS, also known as ‘Cast’ syndrome, Wilkie’s syndrome, ileus chronic duodenal or mesenteric root syndrome, is a rare disease, first described in autopsies, in 1861, by Carl Freiherr Von Rokitsansky.

This syndrome is caused by the clamping of the third portion of the duodenum by an abnormally narrow angle formed between the superior mesenteric artery and the aorta. Normally, this angle is approximately 25–60°, but in pathological situations it may be <6–15°.¹

The SMAS has an incidence of 0.1–0.3%.

It is more common in women (2:1) and in those under 30 years, and has no racial differences.²

This syndrome can have either a congenital or acquired aetiology. The congenital form is due to incomplete rotation of the duodenum, short Treitz ligament and high or low origin of the superior mesenteric artery. The acquired form is due to extreme weight loss, osteoarticular pathology of the spine and oesophagectomy.^{3 4}

There is only one case in the literature of the familial form of this disorder, which was described by Ortiz *et al.*⁵

There are several surgical techniques described, such as duodenojejunostomy (performed in this patient), gastrojejunostomy and Strong’s operation. All these techniques can be performed laparoscopically.

The first duodenojejunostomy for SMAS was performed in 1908, by Stavely. It is the most frequent surgical procedure and has a success rate of about 80%. Duodenojejunostomies have the best results in severe cases, having had significantly better outcomes than gastrojejunostomy and Strong’s procedure.

Figure 5 D3–D4 duodenal clamping by the mesenteric artery and dilated duodenum in the left of the figure.

Gastrojejunostomy has failed to completely release duodenal obstruction, leading to persistence of symptoms, complicated with blind loop syndrome, gastric bile reflux and ulceration. Strong's operation consists of dividing the ligament of Treitz with mobilisation of the transverse and ascending duodenum

Patient's perspective

One year after this surgery, I may say, it clearly changed my life for the better. The postoperative period was wondrous, without any pain, and with me being able to eat almost everything a few days later. Sometimes I still feel some abdominal discomfort after meals, but it is very uncommon for me to vomit or to wake up in pain. Also, I was finally able to put on some weight.

Learning points

- ▶ High level of suspicion.
- ▶ Enhance the patient's symptoms.
- ▶ Appropriate diagnostics.

for caudal displacement of the duodenum. This procedure does not violate the bowel and has been correlated with an earlier postoperative recovery. However, Strong's operation can be difficult or impossible due to adhesions and because caudal displacement of the duodenum cannot always be achieved owing to interference with short vessels from the inferior pancreaticoduodenal artery to the duodenum.⁶

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