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Case Report

Intestinal Obstruction Due to Duodenal Malrotation Successfully Managed by Duodeno-jejunal Derivation on Roux's Loop

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Abstract

Superior Mesentric Artery Syndrome (SMAS) is a rare entity due to compression of the duodenum between the SMA and Aorta. Very rarely, this syndrome can be present in a pre-existing anatomy of congenital malrotation of the duodenum. We describe case of the intestinal obstruction in a 21-year-old woman due to undiagnosed congenital duodenal malrotation presenting with SMAS. The current literature, diagnostic modalities and management options are discussed.

Keywords: Congenital Duodenal Malrotation; Superior Mesenteric Artery Syndrome; Wilkie Syndrome; Arteriomesenteric Occlusion of the Duodenum

Introduction

Congenital duodenal malrotation is a digestive condition that occurs when the duodenum (the first part of the small intestine) is compressed between two vessels: the aorta and the superior mesenteric artery, due to rotation around the SMA axis which leads to superior mesenteric artery syndrome (SMAS) [1]. This result in chronic or acute duodenal obstruction: intermittent, complete or partial [2].

SMAS was first described by Von Rokitansky in 1861, who proposed that it was caused by obstruction of the third part of the duodenum due to arteriomesenteric compression [3]. Subsequently, in 1927, Wilkie published the first study of 75 patients, thus, SMAS is also known as Wilkie syndrome [4]. Some studies report the incidence of SMAS to be about 0.10.3% [3]. Among of patients described in literature, women predominate in a ratio of 3:2 at the age of 10-39 years, but may be diagnosed at any age [5].

SMAS typically occurs due to the loss of the mesenteric fat pad (fatty tissue that surrounds the superior mesenteric artery). Delays in diagnosis may result in significant complications. Depending on the cause and its severity, treatment options can include addressing the underlying cause, dietary changes (small feedings or a liquid diet), and/or surgery [6-8].

We present a clinical case of SMAS with congenital duodenal malrotation in a young woman whose diagnosis was delayed which led to a worsening quality of life. We describe successful surgical management of the patient by duodeno-jejunal Roux-en-y loop.

Case Presentation

21-years-old female presented with postprandial persistent pain in the epigastric and hypogastric region and abdominal distension. The pain started 7 months back and has gradually worsened. Patient underwent laparoscopic surgery for a left ovarian cyst at 15 years of age. She underwent a surgery for intestinal occlusion

with a laparoscopic approach (adhesiolysis) a year ago but the abdominal pain syndrome persisted and the symptomatic drug treatment was inefficient.

The clinical examination findings of the patient showed poor nutrition (BMI – 19.8 kg/m^2). Her vitals were stable. On palpation the abdomen was soft, moderately distended, peristalsis present, negative peritoneal signs. *CT* examination was suggestive of signs of SMA syndrome with marked gastric distension. (Figure 1 a, b) and compression of the third portion of the duodenum between the superior mesenteric artery and the aorta. Upper GI endoscopy showed erythematous gastropathy, duodeno-gastric reflux.

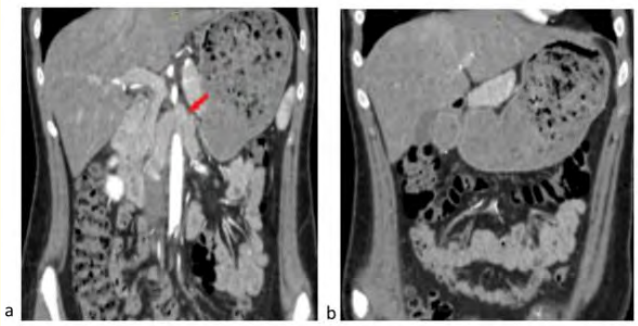

Figure 1: a) Compression of the duodenum (red arrow) by the superior mesenteric artery with b) marked gastric distension.

The barium meal study at 30 min and 2 hours establish chronic gastric stasis (Figure 2 a, b) – at 30 min about 75% and 30% of the ingested volume of barium contrast remained in the stomach at 2 hours.

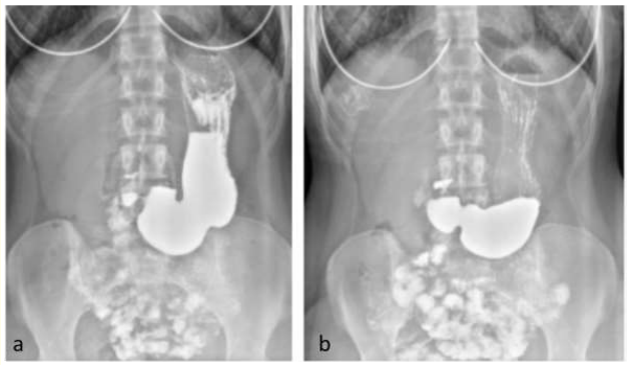

Figure 2: Barium meal study: a) 30min post oral contrast, b) 2 hours post oral contrast ingestion.

Surgical procedure

Under general anesthesia, Upper median laparotomy was performed. Kocher mobilization of the duodenum was performed: duodenal malrotation posterior to the head of the pancreas was determined (Figure 3).

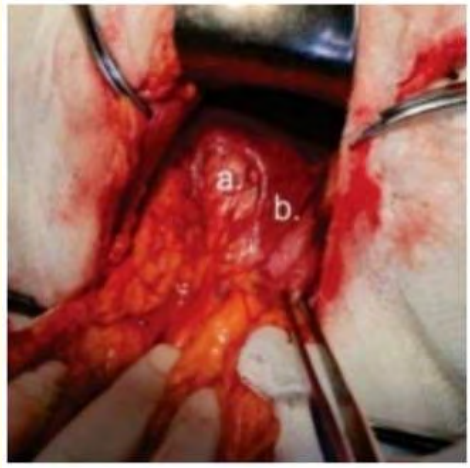

Figure 3: Intraoperative revision of the abdominal organs. a – pancreas, b – malrotation of the duodenum.

This was followed by the mobilization of the duodeno-jejunal ligament (Treitz). The causal factor of compression of the duodenum in the D3 segment through the superior mesenteric artery was established (Figure 3). Latero-lateral duodeno-jejuno anastomosis on the Roux loop was done. In order to decompress the digestive tract, we plced a naso-gastric tube and a naso- jejunal tube for enteral feeding. Intraperitoneal drains were kept at the site of the duodeno-jejunal anastomosis and the right iliac fossa. Postoperatively, the patient underwent analgesic and prokinetic treatment.

Patient had bowel sounds from post-operative day 2. On the 3rd postoperative day, enteral feeding was initiated through the nasal-jejunal tube and oral feeding was initiated on day 5. The patient was discharged on the 8th day in a satisfactory condition. The clinical picture and imaging after one month after surgery was normal.

Discussion and Conclusion

Congenital stenoses and duodenal stenoses by extrinsic compression, including the superior mesenteric artery syndrome,

are common in children (4th place after stenosis of the pylorus, intestines, colon, etc.), while in adulthood with a relatively rare incidence.

Barium digestive examination, scintigraphy of the digestive tract, selective exploration of the duodenum, together with clinical data allow to establish the presumptive diagnosis of extrinsic duodenal subocclusion, especially in malformations of the duodenal frame or duodeno-jejunal angle. Clinical-paraclinical examinations, especially CT imaging with angiography allow an early diagnosis. This case report highlights the role of angiographic CT imaging examination as a method of choice in the diagnosis of upper mesenteric artery syndrome.

This clinical case is actual from its etiopathogenic considerations. It is well known that during embryonic development the intestine supports a complex phenomenon of fixation and rotation, which can be vicious and develop various anatomical and pathological situations, and this are a potential sources of intestinal occlusion. Studies showed that the vast majority of intestinal occlusions may be secondary to an extrinsic compression of the duodenum, duodeno-jejunal angle, or vascular pedicle through pathological attachments, more commonly congenital Jackson-type [9].

The specialized literature frames this case in a variant of the undiagnosed during childhood abdominal congenital anomalies. It is demonstrated that partial or intermittent occlusion present a particular degree of difficulty in the diagnosis [10].

Intestinal malformations, especially duodenal malformations, are usually the prerogative of pediatric surgery. If in childhood the symptomatology of the duodenal malformations is often non-specific, being able to mimic any type of abdominal pain syndrome, in adults the symptomatology usually takes on the appearance of an intestinal occlusive syndrome. In the given case, the diagnosis of congenital duodenal malrotation was neglected in childhood, even if the patient had signs of abdominal pain syndrome.

The individualization of this case consists in the severity of the complication of the benign malformative disease by extrinsic duodenal subocclusive syndrome, as well as by the particular surgical resolution - duodeno-jejunal derivation on the loop of Roux with satisfactory result.

Important etiologic factors that may precipitate narrowing of the aortomesenteric angle and recurrent mechanical obstruction are described in detail in a series of publications and summary articles [11-13]. Patients with intestinal malrotation who were not diagnosed until adulthood may present with a variety of chronic symptoms, including nausea, vomiting, diarrhea, vague abdominal pain, early satiety and bloating, dyspepsia, and peptic or duodenal ulcer disease.

Diagnosis of SMAS is based on interpreting clinical symptoms alongside radiological testing which can confirm its presence. Various imaging modalities can be used: plain film x-ray, barium x-ray, endoscopy, computed tomography (CT), Doppler ultrasound, and magnetic resonance angiography (MRA). CT scan is helpful in diagnostics in that it allows for measurement of aortomesenteric (AO) angle which aids in confirmation of SMA syndrome [12,13].

Laboratory tests are usually nondiagnostic and it is noted that electrolyte disturbances as well as protein and albumin levels can still be normal despite associated weight loss. While it is rare, SMAS is important to consider because the delay in diagnosis can result in significant morbidity and mortality from malnutrition, dehydration, electrolyte abnormalities, gastric pneumatosis and portal venous gas, gastrointestinal hemorrhage and gastric perforation [14-16].

The differential diagnosis includes anorexia nervosa and bulimia. In addition, SMAS should be differentiated from other causes of megaduodenum or duodenal ileus, including diabetes mellitus, collagen vascular conditions, and chronic idiopathic intestinal pseudoobstruction, etc. Mechanical obstruction secondary to peptic ulcer disease or duodenal web should also be considered [17].

Conservative initial treatment is recommended in all patients with SMAS; this includes adequate nutrition, nasogastric decompression, and proper positioning of the patient after eating. Nutritional support through hyperalimentation is of great importance with conservative therapy in an attempt to increase the mesenteric fat pad, thus increasing the AO angle and improving symptoms. Many patients will fail conservative therapy and ultimately require surgical intervention [17-19]. Surgery is indicated when conservative measures are ineffective,

particularly in patients with a long history of progressive weight loss, pronounced duodenal dilatation with stasis, and complicating peptic ulcer disease. Options for surgery include a duodenojejunostomy or gastrojejunostomy to bypass the obstruction or a duodenal derotation procedure (otherwise known as the Strong procedure) to alter the aortomesenteric angle and place the third and fourth portions of the duodenum to the right of the superior mesenteric artery [20,21].

A number of authors have concluded that laparoscopic duodenojejunostomy is safe and effective and should be considered the optimal treatment for patients with SMAS. The authors add that shorter hospitalization, low morbidity, and the high success of laparoscopic enteric bypass make this approach favorable to traditional open techniques [21-23].

Conflict of Interest/Financial Disclosures

None to declare.

Ethical Approval Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images, both orally and in writing, in accordance with the principles of the Declaration of Helsinki on Medical Research Involving Human Subjects.

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