Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Hepato-Gastro-Enterology

Superior Mesenteric Artery Syndrome or Wilkie's Syndrome: About a Case Report and a Review of the Literature

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DOI: 10.36347/sjmcr.2022.v10i08.008 | **Received:** 01.07.2022 | **Accepted:** 07.08.2022 | **Published:** 11.08.2022

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Abstract **Case Report**

Superior mesenteric artery syndrome (SMA syndrome), also known as Wilkie's syndrome is a rare vascular disease due to extrinsic compression of the third portion of the duodenum between the superior mesenteric artery and the aorta. The symptoms are variable and non-specific, the treatment is first of all medical and surgery is reserved for unresponsive cases to medical treatment. We report a case of Superior mesenteric artery syndrome occurring in a 21-year-old patient, revealed by chronic epigastric pain with vomiting, the diagnosis is made by CT scan and the patient underwent laparoscopic gastro-entero-anastomosis, with good postoperative evolution.

Keywords: Superior mesenteric artery syndrome - intestinal obstruction - superior mesenteric artery.

INTRODUCTION

Superior mesenteric artery syndrome or Wilkie's syndrome is a rare pathology, due to the extrinsic compression of the third portion of the duodenum between the superior mesenteric artery and the aorta caused by the reduction of the angle between these two structures after the disappearance of the perivascular fatty tissue [1]. The symptoms are variable and non-specific, consisting of chronic epigastric pain, postprandial vomiting, nausea, early satiety, anorexia, and weight loss; these symptoms are related to the degree of duodenal compression and obstruction [2, 10]. Diagnosis is based on abdominal CT scan. The treatment is first of all medical, and surgery is reserved for unresponsive cases to medical treatment [3, 4].

Through this work we report a case of the Superior mesenteric artery syndrome in order to describe the clinical, diagnostic, and therapeutic characteristics of this rare condition.

CASE REPORT

We report a case of Superior mesenteric artery syndrome occurring in a 21-year-old patient, without any particular pathological history, who presented with chronic atypical epigastric pain associated with postprandial vomiting, and weight loss of 18 kg, with no jaundice diarrhea or constipation.

Physical examination found slight epigastric tenderness with clinical signs of dehydration, her BMI was 17 kg/m2. Laboratory tests revealed initial hypokalemia at 2.3 mmol/l (corrected to 4.1 mmol/l), with no other biological abnormalities.

Esophagogastroduodenoscopy revealed a very erosive antro-fundal mucosa without ulcerative lesions, and without detectable lesions of the duodenum. Histology results showed Moderate chronic gastritis without atrophy or intestinal metaplasia, associated with interstitial duodenitis without specific or suspicious lesion.

She underwent an abdominal-pelvic Computed Tomography (CT) that showed an aortomesenteric angle and aortomesenteric distance at 14.2° and 12.3 mm respectively and the diagnosis of Superior mesenteric artery syndrome was established (Figure 1).

Medical treatment was first undertaken; high calorie diet and prokinetic agents via a nasogastric tube with intravenous (IV) electrolyte and fluid intake, but without significant clinical improvement. The decision was therefore to operate the patient. The surgical

procedure consisted of a laparoscopic gastro-entero-anastomosis with good postoperative evolution (the patient became asymptomatic with weight gain).

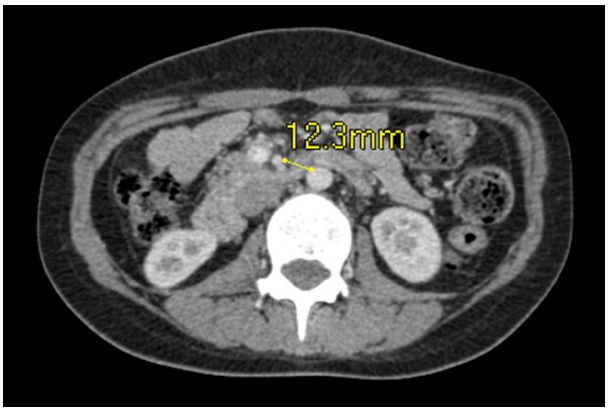

Figure 1: Transverse scannographic section showing an aorta-superior mesenteric artery distance of 12.3 mm with an angle between the aorta and the superior mesenteric artery calculated at 14.2°

DISCUSSION

Rokitansky, in 1861, was the first to describe superior mesenteric artery syndrome [5]. Since then, it has been reported in the medical literature by either incidental radiological findings or following a condition causing significant weight loss with gastrointestinal symptoms.

Superior mesenteric artery syndrome is a rare cause of duodenal obstruction; its pathophysiology is related to a reduced aortomesenteric space and angle [2, 8].

The predisposing factors to develop SMA syndrome are most often: rapid and significant weight loss, trauma/deformity of the spine (Corrective spinal surgery for scoliosis, hyperlordosis), cerebral palsy, and anatomic abnormalities (congenital or acquired) such as a hypertrophied or abnormally short ligament of Treitz attracting the third portion of the duodenum towards the apex of the duodeno-jejunal angle [3, 8].

It occurs preferentially in young female patients aged 17 to 39 years and its incidence rate is between 0.013% and 0.78% [4, 8].

SMA syndrome diagnosis is challenging and often delayed [6]. The symptoms are variable and non-specific. The clinical presentation may be in either the acute or chronic setting or evolving in insidious onset, depending on the etiology and the importance of the duodenal obstruction. The most frequent clinical signs are abdominal pain, vomiting and nausea and weight loss, often associated with epigastric distension. They are aggravated by meals and supine position, relieved when the patient is lying prone, in the left lateral decubitus, or in a knee-chest position. These positions remove pressure from the mesentery and SMA, opening the space between the SMA and the aorta [6-8]. This clinical presentation can be challenging diagnostically and poses the problem of differential diagnosis with other pathologies; hence the diagnosis is made based on clinical evidence and radiological findings [10, 11].

Advances in imaging make it possible to make the diagnosis preoperatively. The standard X-ray confirms the upper obstruction, and the CT scan calculates the angle between the SMA and the aorta, which is reduced from 7° to 22°, whereas it is normally between 25° and 60°. The aorto-mesenteric distance is also reduced and measures between 2 -8 mm, while the normal distance is 10 to 28mm. Imaging can also show gastroduodenal dilatation up to the level of the third portion of the duodenum with an extrinsic compression next to the superior mesenteric artery [7, 8].

The treatment is initially medical and conservative consisting of gastric decompression by a nasogastric tube, correction of hydro-electrolyte abnormalities with enteral/parenteral nutritional. Dietary and nutritional support measures are fundamental and include splitting meals, and posturing maneuvers during meals. Motility agents may be helpful in some patients [3, 6, 8]. Surgical intervention is indicated in cases of conservative treatment failure, a prolonged disease with progressive weight loss, and recurrent upper gastrointestinal symptoms; It is considered necessary in 75% of cases [4, 10, 11]. Surgical options include gastrojejunostomy or duodenojejunostomy, or a modification of local conditions by Strong's procedure: duodenal mobilization to lower the duodenojejunal angle by cutting the ligament of Treitz [9]. Laparoscopic duodenojejunostomy is the treatment of choice for SMA syndrome; it’s an effective minimally invasive option, with an acceptable rate of postoperative complications and favorable long-term results [10, 11].

CONCLUSION

Superior mesenteric artery syndrome is a rare entity, and is a diagnostically challenging condition. High index suspicion in cases of upper, chronic and refractory digestive symptoms in a context of significant weight loss is of utmost importance. The CT scan confirms the diagnosis, and surgery improves symptoms and quality of life.

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