

A Model Example: Coexisting Superior Mesenteric Artery Syndrome and the Nutcracker Phenomenon

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Abstract

Case Report

Aorto-mesenteric clamp syndrome or Wilkie's syndrome is a rare cause of upper intestinal obstruction resulting from compression of the third portion of the duodenum as it passes between the superior mesenteric artery and the aorta. Clinically, it is characterized by intermittent vomiting, nausea and abdominal pain related to the degree of duodenal compression. The diagnosis is based on abdominal computed tomography. Medical management with decompression using a nasogastric tube and adequate nutritional support are initially recommended. Surgical treatment is indicated in case of failure of conservative management.

Keywords: Aorto-mesenteric clamp syndrome, pain, pathology.

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INTRODUCTION

Aorto-mesenteric clamp syndrome or Wilkie's syndrome is a rare pathology due to extrinsic compression of the third portion of the duodenum between the superior mesenteric artery and the aorta in connection with the reduction of the angle between these two structures. The symptomatology is nonspecific. The treatment is first of all medical, its failure requires the use of surgery.

CASE REPORT

This is a 19 year-old boy, from a non consanguineous marriage, followed in the pediatric ward for chronic vomiting with delayed height and weight. Its history of the disease goes back to 4 years by the appearance of intermittent abdominal pain with vomiting.

Postprandial food and transit disorders such as diarrheaconstipation alternation. All of this evolving in a context of unstated apyrexia and weight loss. On clinical examination, the patient presented a slight ulcer, megaduodenum and eating disorders tenderness of the right hypochondrium with skin folds indicating dehydration. His weight was 30kg for a height of 146cm. The patient benefited from several pediatric consultations where the diagnoses of chronic inflammatory bowel disease, hiatus hernia and celiac disease were raised. The biological assessment revealed a hypochromic microcytic anemia.

The ultrasound showed shrinking of D3 (Figure 1). Then the CT scan showed a decrease in the distance between the aorta and the superior mesenteric artery measured at 3mm (Figure 2 & 3) with an aorto-mesenteric angle less than 15 degree confirming the diagnosis of aorto-mesenteric clamp syndrome. After a short preoperative preparation (decompression by a nasogastric tube, rehydration), an internal digestive bypass of the type of laparoscopic gastro-jejunostomy was performed. The post-operative consequences were simple. The resumption of food was gradual around the third postoperative day and discharge from the hospital was on the ninth day. The patient has since been regularly followed in consultation. With a follow-up of 6 months, she is asymptomatic and he gained weight.

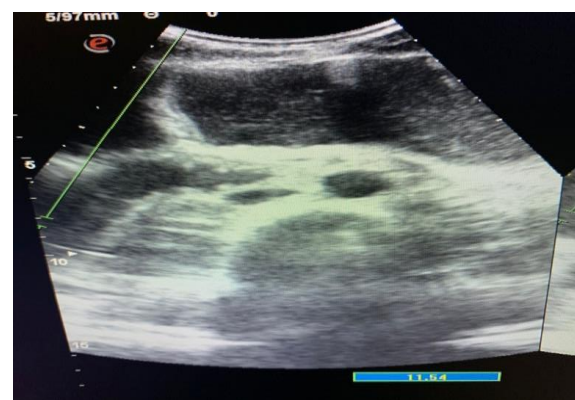


Figure 1: Abdominal ultrasound—reduced angle between abdominal aorta and superior mesenteric artery

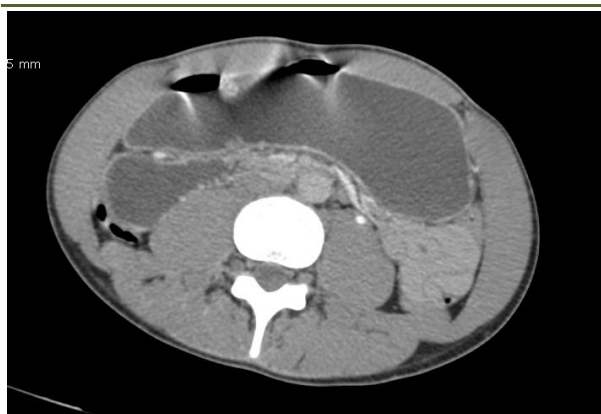


Figure 2: CT scan of abdomen demonstrating duodenal compression between the superior mesenteric artery and aorta



Figure 3: CT scan of the abdomen and pelvis showing impingement upon the left renal vein by the superior mesenteric artery

DISCUSSION

First described by Rokitansky in 1861 and then studied in detail by Wilkie in 1921, aortomesenteric pincer syndrome is a rare cause of duodenal obstruction associated with a wide range of predisposing factors [1]. It is caused by extrinsic compression of the third portion of the duodenum by the superior mesenteric artery or one of its branches against the aorto-spinal plane. This obstruction can be partial or complete, acute or chronic, congenital or acquired [2]. The prevalence of aorto-mesenteric pincer syndrome varies between 0.013% and 0.78%. It occurs preferentially in young female patients. No racial and ethnic predisposition has yet been identified [2-4]. The factors favoring the occurrence of this pathology are most often: rapid weight loss leading to a decrease in the thickness of adipose tissue in the aortomesenteric space, correction of scoliosis, spinal hyperlordosis, cerebral palsy and anatomical abnormalities such as an enlarged Treitz ligament or abnormally short attracting the third duodenal portion towards the apex of the duodenojejunal angle and thus promoting compression of the digestive segment by the superior mesenteric artery [4]. Nevertheless, 40.4% of cases occur without

an obvious triggering factor [2] and our observation seems to fall.

Etiology and the importance of the duodenal obstruction. The most common clinical signs are abdominal pain, bilious vomiting and nausea, often associated with epigastric distension. They are aggravated by eating and supine, relieved by left lateral decubitus and sitting. This symptomatology poses the problem of differential diagnosis with other pathologies such as pancreatitis [3]. The diagnosis, suggested clinically, is confirmed by the abdominal angiography which shows a dilation gastroduodenal up to the level of the third portion of the duodenum sitting in an extrinsic compression opposite the superior mesenteric artery. The superior mesenteric aortaartery distance is less than 8mm and the aortomesenteric angle is less than 22°. The therapeutic management is initially medical aimed at relieving the symptoms of the obstruction. It includes gastric decompression by a nasogastric tube, the correction of fluid and electrolyte disturbances Supplementation.

Nutritional parenteral or enteral route helps to gain weight and restore aortomesenteric adipose tissue which in normal individuals, moves the superior mesenteric artery in front of Aorto-mesenteric clamp syndrome is a rare and poorly understood pathology, posing diagnostic and therapeutic difficulties. Its diagnosis is based on clinical and radiological arguments. The implementation of appropriate conservative measures and the definition of surgical indications are essential for the prevention of complications.

CONCLUSION

Wilkie syndrome and nutcracker syndrome are rare compression syndromes. The diagnosis and treatment of Wilkie syndrome is controversial. The initial management should include gastric and duodenal decompression, treatment of fluid and electrolyte imbalance as well as reversal of weight loss using enteral and parenteral feeding. Surgery may be appropriate in some patients who continue to have debilitating symptoms after a good trial of conservative therapy.

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