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Wilkie's syndrome

An uncommon cause of small bowel obstruction in a young patient

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Wilkie's syndrome. An Uncommun cause of small obstruction in a young patient

Superior mesenteric artery (SMA) syndrome, also named as Wilkie's syndrome, is a rare, potentially life-threatening gastro-vascular disorder and an uncommon cause of proximal bowel obstruction characterized by a compression of the third and final portion of the duodenum by the abdominal aorta and the overlying superior mesenteric artery. There isn't a consensus regarding to the optimal treatment of this condition. We describe a case of Wilkie's syndrome in a young female with characteristic symptoms of upper intestinal obstruction, whose diagnosis was delayed for 2 years.

KEY WORDS: Superior mesenteric artery, Upper intestinal occlusion, Young female

Introduction

Superior mesenteric artery (SMA) syndrome, also named as Wilkie's syndrome or cast syndrome, is a rare, potentially life-threatening gastro-vascular disorder and an uncommon cause of proximal bowel obstruction characterized by a compression of the third and final portion of the duodenum by the abdominal aorta and the overlying superior mesenteric artery. Suggestive symptoms are early satiety, nausea and recurrent vomiting, abdominal distention, weight loss, and extreme "stabbing" postprandial abdominal pain. Medical treatment should be proposed first in all cases, except for cases where emergency surgery is necessary upon presentation. If medical treatment fails, or is not feasible due to severe illness, surgical intervention is requested. We describe a case of Wilkie's syndrome in a young female with characteristic symptoms of upper intestinal obstruction, whose diagnosis was delayed for 2 years.

Case presentation

A 22-year-old woman with no remarkable medical history was referred to the surgical department with a 2-years history of weight loss, early satiety and chronic postprandial abdominal pain. Physical examination revealed an underweight and long-limbed woman, with no operation scars on the abdomen; deep palpation revealed a painless palpable mass during examination in epigastrium and in right quadrants. Laboratory tests were normal. A CT scan of the abdomen and pelvis was performed and showed an acute angle of 14° (normal range 38-56°) between the SMA and the aorta, with impression upon the anterior third part of the duodenum (Figs. 1, 2). Gastrointestinal barium series showed delayed transit through the third part of the duodenum. These findings suggested Wilkie's syndrome. Then a conservative medical treatment was started, with gastric decompression by naso-gastric tube and parenteral nutrition followed by oral diet. The patient was discharged after 5 days of low diet advancement and was asymptomatic with an increased weight at 6-month follow-up.

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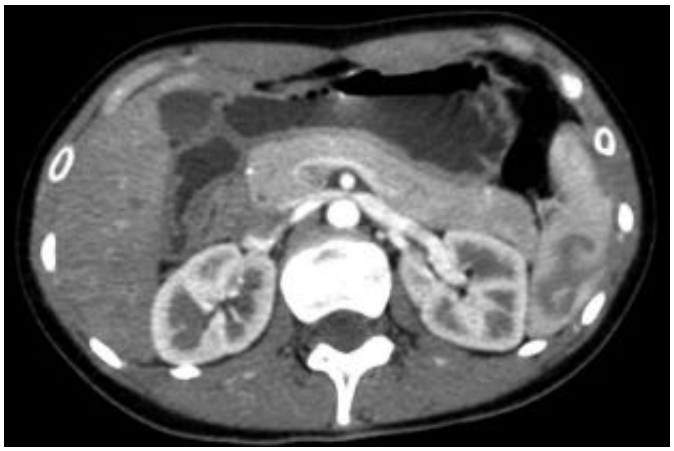

Fig. 1: Axial image through the stomach from a contrast-enhanced computed tomography of the abdomen. The gastric lumen and proximal duodenum are markedly distended and the third portion of duodenum is compressed between the SMA and the aorta. It also shows a compression of the left renal vein.

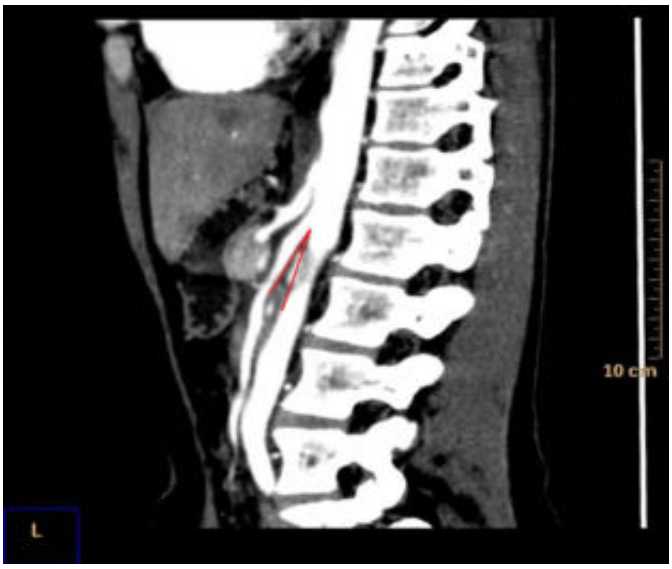

Fig. 2: Sagittal image through the stomach from a contrast-enhanced computed tomography scan of the abdomen. The horizontal segment of the duodenum narrows abruptly as it passes between the SMA and the aorta an angle of 14°.

Discussion

Superior mesenteric artery syndrome, also known as Wilkie's syndrome, is a rare condition of upper intestinal obstruction in which the third part of the duodenum is compressed by the overlying, narrow-angled superior mesenteric artery against the posterior structures; it mainly affects females, whereof about 75% of cases is stratified in age between 9-30 years-old [1-3].

Some cases of Wilkies's syndrome have been described in neonates and infants [4-7]. SMA syndrome was first described in 1861 by Carl Freiherr von Rokitansky in victims at autopsy, but remained pathologically undefined until 1927 when D.P.D. Wilkie published the first comprehensive series of 75 patients, describing a condition defined as "chronic duodenal ileus" [8].

The main anatomical feature of this syndrome is the narrowing of the angle between the SMA and the aorta, which is normally 38° to 56°. In SMA syndrome, this angle is typically decreased to 6° to 25° and as a result, the aorto-mesenteric distance decreases to <10 mm, from normally 10 to 28 mm [9,10]. SMA syndrome can present in two forms: chronic/congenital or acute/induced. Congenital etiologies include an unusually low insertion of the SMA or high insertion of the angle of Treitz dislocating the duodenum to a cranial position. Acquired anatomic abnormalities can occur following corrective spinal surgery such as scoliosis surgery [11], spinal trauma, and after abdominal surgery such as total proctocolectomy and ileal J-pouch anal anastomosis due to tension of the small bowel mesentery [12], left nephrectomy [13]. SMA syndrome could occur after rapid weight loss from anorexia nervosa, trauma, brain injury, immobilization with a body cast (cast syndrome), neoplastic diseases and malabsorptive states, that could lead to a severe reduction of thickness of the adipose tissue in the aorto-mesenteric space [14].

Considering the rarity of this syndrome, the diagnosis can be difficult and it is often made after ruling out other more common causes and after performing diagnostic procedures such as esophagogastroduodenoscopy and colonoscopy. High clinical suspicion is warranted and diagnosis is based on clinical evidence supported by radiological findings. Barium radiography demonstrates dilatation of the first and second part of the duodenum with or without gastric dilatation, anti-peristaltic flow of barium proximal to the obstruction and a delay of 4–6 h in gastroduodenojejunal transit time [15]. Contrast-enhanced CT or magnetic resonance angiography enable visualization of the vascular compression of the duodenum and precise measurement of the aorto-mesenteric angle and distance [15].

However, radiological examinations sometimes do not provide adequate diagnostic and clinical elements and this can lead to consider this syndrome as a psychological disorder, such as anorexia nervosa, which leads to a delay of treatment resulting in increased morbidity.

Nowadays, there isn't a consensus regarding to the optimal treatment of this condition. The treatment of SMA should begin with conservative measures consisting in gastric decompression, parenteral nutrition and/or postpyloric feeding when possible, followed by oral diet as

tolerated. Surgery is only indicated when conservative therapy has failed or when a diagnosis is difficult to differentiate. Surgical options that have been proposed include mobilization of the duodenum by section of the ligament of Treitz, allowing the duodenum to fall away from the aorta (Strong's procedure) in congenital abnormalities [16], several types of reconstruction such as latero-lateral duodeno-jejunostomy [17] or Roux-en-Y reconstruction, recently also by laparoscopic [28-21] or robotic surgery [22], gastro-jejunostomy [23], anterior transposition of the duodenum[24], duodenal derotation [25] and duodenal circular drainage operation [26]. Infrarenal transposition of the superior mesenteric artery has also been described [27]. According to scientific literature, duodenojejunostomy remains the operation of choice to relieve the obstruction, with a success rate up to 90% [28].

Conclusion

SMA syndrome should be considered among the possible causes of upper intestinal obstruction, especially in young females. The diagnosis is made difficult by poor peculiarities of the symptoms and must be taken into consideration when excluding all other possible causes of intestinal occlusion. SMA syndrome is successfully treated medically. Surgical intervention should be reserved for those individuals refractory to medical therapy or when laparotomy is required at the time of presentation.

Riassunto

La sindrome di Wilkie, definite anche sindrome del compasso aorto-mesenterico definita sindrome dell'arteria mesenterica superiore, è una causa rara di occlusione intestinale alta legata alla compressione da parte dell'arteria mesenterica superiore sul terzo duodeno per un'anomalia di angolazione aorto-mesenterica. Questa sindrome colpisce prevalentemente giovani donne. Esistono condizioni predisponenti nelle forma acquisita che determinano una riduzione del cuscinetto adiposo retroperitoneale che si interpone nell'angolo superiore compreso tra il margine superiore del duodeno e la superficie inferiore dell'arteria mesenterica superiore. La diagnosi non è agevole. Le metodiche radiologiche svolgono un ruolo chiave nell'orientamento diagnostico. La terapia è prevalentemente medica. Un trattamento chirurgico si rende necessario in caso di fallimento della terapia medica o in casi particolari di urgenza.

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