CASE REPORT

Wilkie's Syndrome: A Rare Etiology of Recurrent Vomiting

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Abstract

Wilkie's syndrome is a rare cause of duodenum extrinsic compression by an excessively small aortomesenteric angle. It presents with signs of upper intestinal obstruction. Definitive diagnosis requires imaging tests. We report the clinical case of a previous healthy 17-year-old male who presented with a three years history of postprandial vomiting and 4 kg weight loss. He had no changes in bowel frequency, fever, gastrointestinal bleeding, altered eating behavior, or physical examination findings. The Wilkie's syndrome was confirmed by computed tomography angiography. After three months of conservative treatment he became asymptomatic. Weight loss and upper intestinal obstruction should raise the suspicion of Wilkie's syndrome. It requires a high level of suspicion and multidisciplinary, conservative treatment is preferred. Because of the rarity of the case, the authors alert to this entity.

Keywords: Adolescent; Superior Mesenteric Artery Syndrome/diagnosis; Superior Mesenteric Artery Syndrome/diagnostic imaging; Superior Mesenteric Artery Syndrome/therapy; Vomiting/etiology; Weight Loss

Introduction

Wilkie's syndrome or superior mesenteric artery (SMA) syndrome is a rare disease in which the angle between the abdominal aorta and upper mesenteric artery (aortomesenteric angle) is excessively acute (usually between 38 and 65º), causing compression of the third transverse portion of the duodenum in its path between these two vessels. Duodenal compression causes upper, partial or complete intestinal obstruction, and occurs more often in young people with a history of significant weight loss.[1-5]

Given the lack of specificity of the clinical findings, there is often a delay in diagnosis, requiring a high level of suspicion. It manifests with postprandial fullness, vomiting, abdominal pain and weight loss among other symptoms, and may have an acute or chronic onset.[1,2] Definitive diagnosis requires imaging tests. The treatment is conservative by opting for the surgery when the first one fails.[1] The authors describe a Wilkie's syndrome case in an underweight adolescent, aiming to alert the existence of this disease and thus preventing the diagnostic delay and its potential complications.

Case Report

A 17-year-old male, without relevant history, was observed with a three-year history of epigastralgia, occasional late postprandial vomiting (no biliary or fecal), and postprandial fullness. His symptoms improved with the trunks anteflexion and in the supine position. He reported a progressive weight loss of 4 kg. No change in intestinal transit, fever, gastrointestinal bleeding or disturbance of eating behavior. The patient presented good general condition with weight, height and body mass index (BMI) in percentile 25-50. He presented a normal abdomen: normal bowel sounds, without mass or organomegaly, without signs of peritoneal irritation. The remaining examination was irrelevant.

Obstructive syndrome (gastric or intestinal, including Wilkie's syndrome), celiac disease, gastroduodenal peptic disease, intestinal parasitosis and renal failure were the most likely diagnostics.

At the first visit a blood count was requested, ionogram (sodium/potassium 138/4.1 mmol/L), renal function (urea/creatinine 39/0.57 mg/dL), ferritin, hepatic cytolysis and celiac disease markers, venous blood gas analysis, urinary sediment examination, fecal occult blood test, and feces parasitological examination, they were all normal. Barium contrast-enhanced esophagogastroduodenal radiography was performed, which was suggestive of Wilkie's syndrome (moderate dilatation of the duodenal arch, especially of the second and third portions) (Fig. 1).

The axial tomography angiography demonstrated a reduction of the angle between the proximal portion of the superior mesenteric artery and the abdominal aorta (15º),

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resulting in marked extrinsic compression of the third portion of the duodenum (maximal endoluminal caliber of 3 mm), making frankly difficult duodenal emptying, which suggested Wilkie's syndrome (Fig. 2). It also revealed left renal vein moderate extrinsic compression, "pinched" between the superior mesenteric artery and the aorta with a minimum caliber of about 5 mm, findings suggestive of Nutcracker's syndrome.

The patient was directed to nutrition consultation to increase visceral fat and pediatric surgery to evaluate the need for surgical treatment. Since the progressive improvement, it was decided to maintain a conservative medical treatment, with a daily meal plan divided into 6-7 meals, hypercaloric (2400 kcal/day) and hyperproteic with carbohydrate supplementation (Fantomalt®). The patient became asymptomatic after three months.

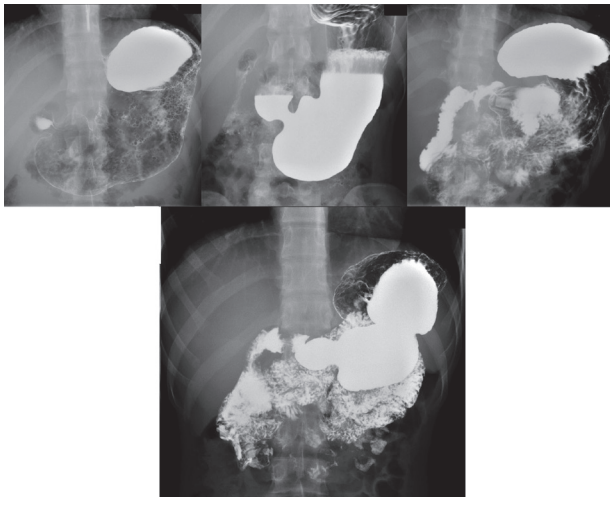

Figure 1. Above: Sequence of images of the esophagus-gastro-duodenal radiological study demonstrating moderate duodenal dilation, especially of the second duodenal portion. Below: Cinematic image reveals duodenal struggle peristalsis.

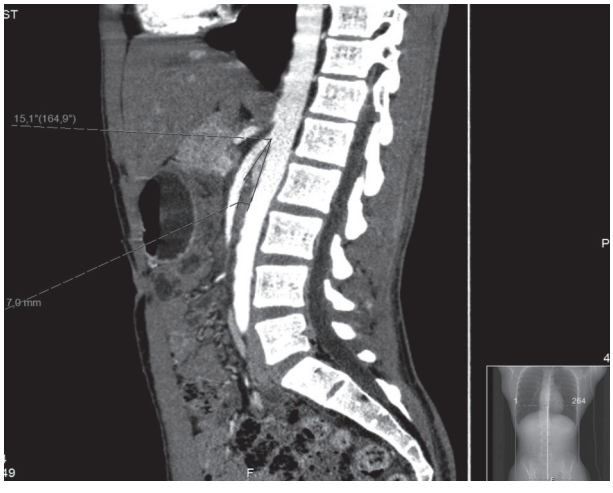

Figure 2. Sagittal section of angiography by computed axial tomography where the marked reduction of the angle between the proximal portion of the superior mesenteric artery and the abdominal aorta (15º) can be visualized, with consequent extrinsic compression on the third portion of the duodenum.

Discussion

Wilkie's syndrome is a rare disorder with an incidence that ranges between 0.013% and 0.3%.[1,2]

Superior mesenteric artery syndrome was first described by the Austrian professor Carl Freiherr von Rokitansky in 1861. Later, Wilkie provided a more detailed clinical and pathophysiologic description in a series of 64 patients and suggested treatment approaches. After that, a controversy regarding the actual existence of this syndrome started, especially because of the lack of specificity of the symptoms and the long list of differential diagnoses.[1,6] The authors' opinion is that the current advance in diagnostic imaging allows the diagnosis in situations previously considered as idiopathic.

The superior mesenteric artery branch runs from the abdominal aorta at an acute angle descending to the root of the mesentery and travelling to the duodenum, usually to the right of the midline. In humans, the aorta-SMA angle ranges from 38º to 65º, due to the erect posture, while in quadrupeds, it is nearly a right angle. The main anatomic feature of SMA syndrome is a narrowing of the aorta-SMA angle to 20º-25º, and as a result, the aortomesenteric distance decreases to 2-8 mm, rather than 10-20 mm. The symptoms severity depends on the degree of compression of the duodenum.[1] In this adolescent, the aortomesenteric angle was 15º, slightly lower than that considered normal, and it is therefore justified that he had mild symptoms.

There are several causes of Wilkie's syndrome, shown in Table 1. The most likely cause in this patient was a congenital anatomical change. Risk factors include thin body *habitus*, prolonged bed rest, abdominal surgery, and exaggerated lumbar lordosis.[3,7] In this patient, none of these risk factors were identified.

Patients with SMA syndrome may present acutely, with chronic insidious symptomatology, or with an acute exacerbation of chronic symptoms. The acute presentation is usually characterized by signs and

Table 1. Main etiology of Wilkie's syndrome

Anatomical alteration	Congenital	- Insertion too low of the mesenteric superior artery - Insertion too high of Treitz's angle
	Acquired	- Post-surgery of the spine - Spinal injury - Post-surgery abdominal - Inflammatory states
Important weight loss	Mesenteric fat loss next to mesenteric superior artery	- Catabolic state - Changes in eating behavior - Consuming diseases

symptoms of duodenal obstruction. Chronic cases may present with long-standing vague abdominal symptoms or recurrent episodes of abdominal pain, associated with vomiting. Other less common symptoms are esophageal reflux, early satiety with a sensation of fullness owing to increased gastroduodenal transit time, and gastric distension.[1] In this adolescent, the presentation was chronic insidious, perhaps due to the fact that the aortomesenteric angle is only slightly inferior to the considered norm.

A detailed history, as well as imaging findings, should highly raise the clinical suspicion for the diagnosis of SMA syndrome. A delay in this diagnosis can potentially lead to many complications, such as electrolyte imbalance, catabolic wasting, peritonitis and gastric perforation.[1] In this clinical case, there were no complications.

Barium radiography demonstrates dilatation of the first and second part of the duodenum with or without gastric dilatation, an anti-peristaltic flow of barium proximal to the obstruction and a delay of 4-6 hours in gastroduodenojejunal transit time, with relief of obstruction when the patient is placed in the prone, knee-chest or left lateral position.[1,8] In this patient, the findings on contrast-enhanced esophagogastroduodenal radiography raised the diagnostic suspicion. In fact, this adolescent's symptomatology was relieved with the anteflexion of the trunk, as referred in the literature.

Contrast-enhanced axial tomography or magnetic resonance angiography enables visualization of the vascular compression of the duodenum and precise measurement of the aortomesenteric angle. Endoscopic examination may visualize a pulsatile extrinsic compression suggestive of this condition.[1] Indeed, in this adolescent, the diagnosis was confirmed by axial tomography angiography.

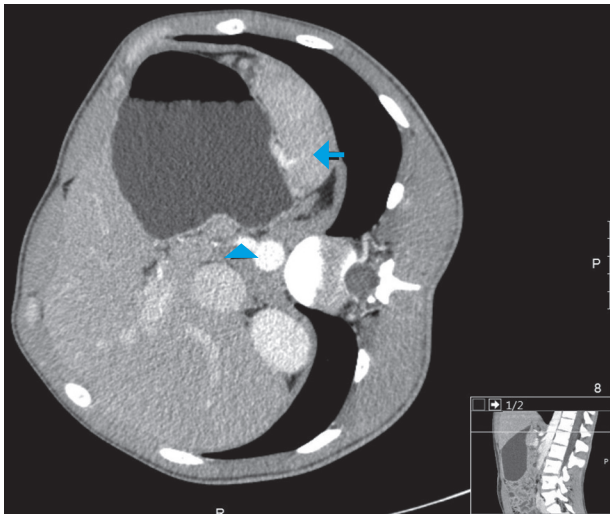

Figure 3. Cross section of angiography by computed axial tomography where the proximal portion of the superior mesenteric artery (arrowhead) and extrinsic compression can be seen on the third portion of the duodenum (with a maximum endoluminal gauge of 3 mm), with consequent upstream dilatation (arrow).

Conservative treatment consists of measures such as gastric decompression, parenteral nutrition and/or post-pyloric feeding when possible, followed by oral diet as tolerated. Posturing maneuvers during meals and motility agents may be helpful in some patients. Surgery may be considered if conservative treatment fails. The duodenojejunostomy is the operation of choice to relieve the obstruction, with a success rate of 90%. Another less invasive surgical option, known as Strong's procedure, involves lysis of the ligament of Treitz with the mobilization of the duodenum; however, this operation had a failure rate of 25%.[1,9,10]

The priority treatment is medical, which aims at weight gain for the mesenteric fat increase and consequent increase in the aortomesenteric angle. Since this patient had progressive regression of the symptoms, the conservative medical treatment was adopted with a hypercaloric and hyperproteic diet.

The prognosis is variable, as it depends on the degree of duodenal compression and the response to therapy. In this patient, since there was a regression of symptoms with conservative treatment, a good prognosis would be expected, however, requiring medical surveillance and a carefully implemented food plan for life. On the other hand, this adolescent may manifest Nutcracker syndrome clinic throughout his life, which reinforces the need for vigilance.

Wilkie's syndrome should be considered in the differential diagnosis in children or adolescents with weight loss and symptoms suggestive of upper intestinal obstruction and/or gastric distension. The diagnosis requires a high level of presumption, its approach is multidisciplinary, and the treatment is preferably conservative. Since these cases are rare, the authors aim to highlight this syndrome.

WHAT THIS STUDY ADDS

- The extrinsic intestinal compression causes should be considered in all age groups, including the adolescent.
- Clinicians should always be aware of the clinical condition of vomiting associated with weight loss and consider the possibility of an obstructive condition.
- Digestive symptoms may be due to underlying vascular disease.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Confidentiality of data
The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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Síndrome de Wilkie: Uma Etiologia Rara de Vómitos Recorrentes

A síndrome de Wilkie é uma doença rara em que há compressão do duodeno por um ângulo aortomesentérico excessivamente agudo. Tem apresentação compatível com obstrução intestinal alta e o diagnóstico definitivo requer exames imagiológicos. Os autores descrevem o caso clínico de um adolescente de 17 anos, sem antecedentes patológicos, com história de três anos de evolução de enfartamento, vómitos pós-prandiais ocasionais e perda ponderal progressiva de 4 kg. Sem alteração do trânsito intestinal, febre, hemorragia gastrointestinal, alteração do comportamento alimentar ou alterações no exame físico. A investigação foi sugestiva da síndrome de Wilkie, confirmada por angiografia por tomografia computorizada. Ficou assintomático após três meses de tratamento conservador.
Perante um quadro clínico de perda ponderal e obstrução intestinal alta deve considerar-se a síndrome de Wilkie no diagnóstico diferencial. O diagnóstico exige um elevado nível de suspeita, a abordagem é multidisciplinar e o tratamento conservador é o preferencial. Dada a sua raridade, os autores pretendem alertar para a existência desta entidade.

Palavras-Chave: Adolescente; Perda de Peso; Síndrome da Artéria Mesentérica Superior/diagnóstico; Síndrome da Artéria Mesentérica Superior/diagnóstico por imagem; Síndrome da Artéria Mesentérica Superior/tratamento; Vómito/etiologia