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Wilkie Syndrome, an Unusual Cause of Weight Loss: Case Report and Literature Review

Rodolfo Martín Ruiz Ravelo[1], José de Jesús Fernández Rivera[1], Juan Carlos Navarro Hernández[2], Marcela Molinar Lechuga[1], Yoatzihual Estrada González[3]

[1] Mexican Social Security Institute – General Hospital of Zone No. 33. Department of Internal Medicine. Monterrey, Nuevo León, México

[2] Mexican Social Security Institute – General Hospital of Zone No. 33. Department of Gastroenterology. Monterrey, Nuevo León, México

[3] Mexican Social Security Institute – General Hospital of Zone No. 1. Department of General Medicine. Tepic, Nayarit, México

ABSTRACT

Wilkie syndrome also known as superior mesenteric artery syndrome is an unusual and rarely cause of upper intestinal obstruction and weight loss; It is characterized by duodenal compression in its third portion caused by a narrowing of the space and angle that separates the aorta from the superior mesenteric artery. This narrowing is usually secondary to multiple etiologies, the most accepted being the decrease in mesenteric fat between these arteries.

The clinical picture and laboratory studies are usually not very specific, so the diagnosis is based on imaging methods, with computed tomography as the gold standard because it shows a decrease in the aortomesenteric angle and distance. Treatment is generally curative and can be conservative or surgical; this depends on the etiology, the time of evolution and symptoms of the case in question, so it should always be individualized based on patient characteristics.

KEYWORDS: Wilkie syndrome, Superior mesenteric artery, Weight loss, Upper intestinal obstruction, Duodenal compression.

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INTRODUCTION

Wilkie syndrome or superior mesenteric artery syndrome (SAMS) is a rare syndrome characterized by compression of the duodenum in its third portion and is secondary to a narrowing of the space and angle found between the superior mesenteric artery and the aorta [1,2]. There are multiple risk factors for the development of Wilkie syndrome, the most accepted triggering factor to date being significant weight loss, generally greater than 10 kg [3], which leads to a decrease in the retroperitoneal mesenteric fat that serves as a cushion between the aorta and the superior mesenteric artery [4,5].

The clinical picture is extremely non-specific, variable and may have a presentation of sudden onset or chronic development [6], the symptoms of upper intestinal obstruction being the most frequently presented. Like the clinical picture, laboratory findings are nonspecific and only shows hydroelectrolytic alterations when vomiting has been severe [1].

Therefore, the diagnosis falls mainly on imaging studies. There are several diagnostic imaging methods who, accompanied by clinical suspicion, can lead to the correct diagnosis, such as barium studies of the digestive tract, arteriography, among others. Currently, the gold standard is computed tomography (CT), that allows stablish the diagnosis in 100% of cases while ruling out other differential diagnoses.

The management of Wilkie syndrome is individualized, so we can opt, depending of the case for conservative treatment, based mainly on dietary hygienic measures, prokinetic and antisecretory medications, with the main objective being to weight gain [7], or we can lean toward surgical management [8].

To our knowledge, there are less than 500 cases reported at today of Wilkie syndrome, however, due to the clinical picture is similar with other common disorders that presents with upper intestinal obstruction symptoms such as neoplasms of the gastrointestinal tract, dyspepsia or

gastroesophageal reflux disease (GERD) is useful to take account in those cases where a specific diagnosis can´t be reached of the pathology causing of the said symptoms.
In this review, key points in the diagnosis and treatment of this potentially reversible pathology will be discussed.

CASE REPORT

We present the case of a 23-year-old female patient, with no family or personal history relevant to the current condition.
She reports that her condition began approximately 4 months prior to our evaluation with the presence of abdominal burning pain in epigastrium and mesogastrium with a intensity of 5/10, without irradiation, predominantly nocturnal, postprandial, especially when she ate irritating foods, as well as a partial decrease in the symptoms when adopting the left lateral decubitus position and when she intake of antispasmodics and proton pump inhibitors.
A month after the onset of the symptoms, she began to manifest early satiety and postprandial nausea, tolerating only half of the food she previously consumed. However, as time went by, she reported that the symptoms increased even after reducing food intake that leading to self-induced emesis in some occasions; She had a weight loss of up to 8 kilograms during that period of time.
A week prior to our evaluation, she presented an exacerbation of her clinical condition characterized by more intense pain, asthenia and adynamia, which didn´t subside with the previously implemented measures, as well as the presence of an episode of hematemesis of approximately 100 ml, so she decided to go to our hospital service, without presenting data of hemodynamic instability: T/A: 120/70 mmHg, HR: 70 bpm, RR 20 rpm. Her examination highlighted only epigastric abdominal pain on mid-deep palpation without signs of peritoneal irritation and the presence of a probable tumoral mass with hard consistency in epigastrium.
The following initial studies were performed: Hemoglobin 13g/dl, hematocrit 39.2%, platelets 185 K/ul, leukocytes 5.7 K/ul, albumin 4.24 g/dl, rest of the laboratories within normal ranges. Due to non-specifical clinical picture and laboratories we initially suspected that the cause of the current condition was tumoral etiology, so a simple and contrast-enhanced computed tomography of the abdomen (CT) was performed; In the CT the primary suspect diagnosis was ruled out and it evidence the presence of occlusion in the third portion of the duodenum was secondary to compression by the superior mesenteric artery (SMA) (Figures 1 and 2); given the intolerance oral intake we put a nasogastric tube however the intolerance oral intake persisted, so we decided to treat with parenteral nutrition.

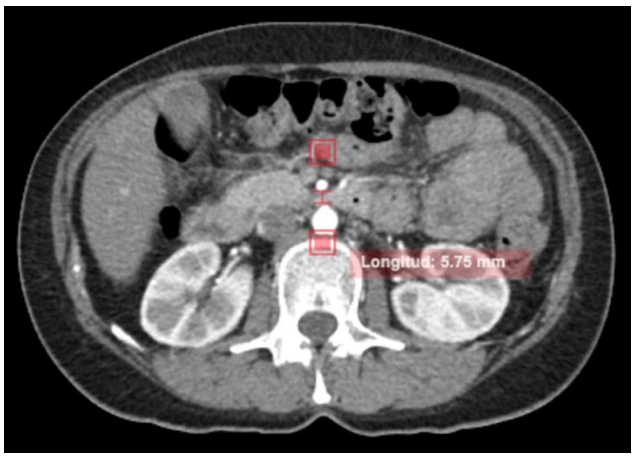

Figure 1. Abdominal computed tomography in axial projection with contrast in the arterial phase, a compression of the third portion of the duodenum and distance between the aorta and SMA less than 8mm is observed.

Within the diagnostic approach, a endoscopy was performed to rule out an intrinsic intestinal obstruction in which the obstruction site couldn´t be visualized (Figure 3), highlighting the presence of mild non-erosive esophagitis and erosive gastropathy predominantly of the body. So the Wilkie syndrome diagnosis was concluded. However given the chronicity of the condition and poor evidence of resolution with conservative treatment, surgical management was chosen and therapeutic laparotomy was carried out, confirming SAMS when aortomesenteric compression was found in the third portion of the duodenum, subsequently duodenojejunostomy was performed without present any complications presenting a good post-surgical evolution with adequate tolerance to the gradual resumption of oral intake, which is why it was decided to discharge him from the hospital.

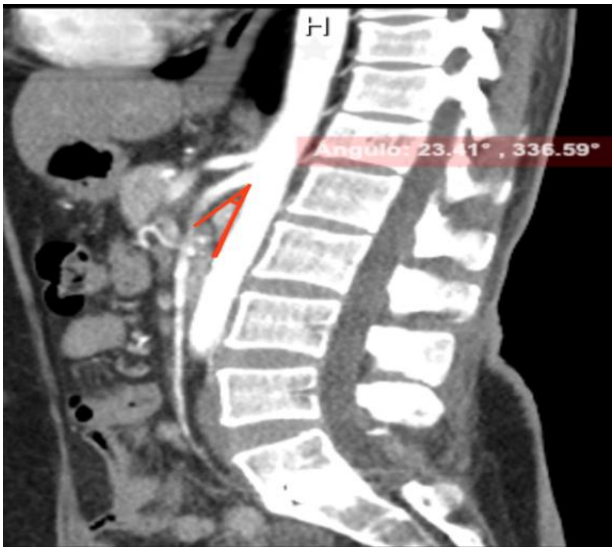

Figure 2. Abdominal computed tomography in sagittal projection with contrast in the arterial phase, a decrease in the aortomesenteric angle of 23.41º is observed.

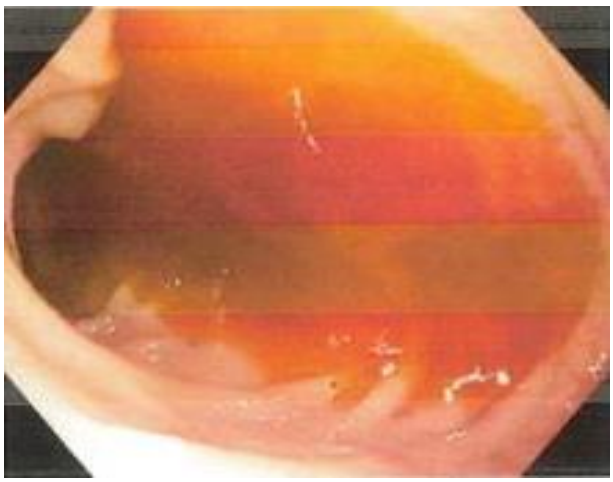

Figure 3. Upper endoscopy at the beginning of the third portion of the duodenum, no obstruction site is seen.

During her follow-up, she presented symptoms of upper intestinal subocclusion 4 months after the surgical procedure, that required readmission and was successfully resolved with conservative management after the placement of a nasogastric tube, so she was discharged without complications. Today, after two years of patient follow-up, she maintains adequate tolerance to the oral intake, completely denying having had another episode of postprandial abdominal pain, confirming the success of the therapy used.

DISCUSSION

Wilkie syndrome was described for the first time more than 100 years ago by Rokitansky [9]. Its incidence is extremely low, being approximately 0.013 to 0.3%.

It affects women more frequently than men (3:2) and its usual age of presentation is between 10 and 30 years [10, 11]. The main determinant is the narrowing of the duodenal lumen caused by a decrease in space between the SMA and the aorta, being the weight loss the precipitating factor of excellence; The other risk factors are: HIV infection, neoplasms, diabetes mellitus, bariatric surgery, vascular surgery, or idiopathic in up to 45% of cases, among others [12, 13] (Table 1).

The symptoms reported by the most of patients coincide with a upper intestinal obstruction manifested by postprandial epigastric pain, early satiety, abdominal distension, nausea, bile vomiting, heartburn, regurgitation and weight loss; Some positions can alleviate the symptoms, with left lateral decubitus position being the most frequently adopted because it tends to release duodenal compression, as was observed in the case of our patient [14].

Due to the unclear clinical manifestations, the differential diagnosis is wide, among that stand out: mesenteric ischemia, GERD and other causes of upper intestinal obstruction such as abdominal neoplasms [15], as was the case of our patient in whom the initial diagnostic suspicion was an abdominal tumor.

Given the no specificity of SAMS, we depend on imaging studies to reach a definitive diagnosis; Among them are barium studies that shows gastric or even duodenal dilation with slow gastroduodenojejunal transit, arteriography, which was previously the diagnostic method of choice, and endoscopy in which pulsatile extrinsic compression and narrowing at the level of the third portion of the duodenum [5], and besides is useful since it can rule out an intrinsic cause of the occlusive symptoms [16]. However, at this time the gold standard is CT, having a sensitivity and specificity close to 100% when the following criteria are met: 1) abrupt obstruction in the third portion of the duodenum 2) an aortomesenteric angle less than 25◦ (normal 38◦ to 65◦) 3) distance from the aorta to the mesenteric artery equal to or less than 8 mm (normal between 13 and 34 mm) and sometimes 4) high fixation of the duodenum by the ligament of Treitz or anatomical variants of the superior mesenteric artery [17, 18, 19].

The treatment depends of the chronicity and severity of the condition, however, the initial recommendation is conservative treatment because it has a success rate of up to 85% [20]. It is focused on relieving the symptoms caused by the obstruction and correct the precipitating factors (Decrease in perimesenteric fat), an objective that is achieved opting for the enteral route after the placement of a nasogastric tube for adequate rehydration and nutrition of the patient by adopting special postures like left lateral decubitus that release the obstruction and cause opening of the aortomesenteric angle due to mechanical effects [21], in cases of refractoriness, feeding is carried out via nasojejunal tube placed by endoscopy or, if it fails, initiation of parenteral nutrition (PNT) in which it is prepared for surgical treatment [22]. Surgery is indicated in patients with the following characteristics: 1) failure of conservative treatment, 2) long-standing disease with progressive weight loss and duodenal dilation with stasis, and 3) complicated peptic ulcer disease secondary to bile stasis and reflux [23].

There are several surgical approaches including duodenojejunostomy, gastrojejunostomy and section of the Treitz ligament to mobilize the duodenum (Strong's technique) [24, 25]. Laparoscopic duodenojejunostomy is the surgical technique of choice with long-term success rates of up to 90% [26, 27].

Therefore, it´s always necessary opt for the most convenient therapeutic approach for the patient, whether conservative or surgical, always individualizing each case and taking into account the cause of the syndrome, the general condition of the patient and the time since the symptomatology onset until the moment of diagnosis.

CONCLUSION

As previously mentioned, Wilkie syndrome is a pathology rarely reported in clinical practice, mainly due to its low diagnostic suspicion, above all in patients who presents

Table 1. Risk factors for the development of Wilkie syndrome [12].

Risk factors for the development of Wilkie syndrome
Lost weight syndromes
Diabetes Mellitus type II
Neoplasms
HIV Infection
Endocrinopathies
Intestinal malabsorption
Eating disorders
Anorexia
Bulimia
Surgical causes
Orthopedic surgery for scoliosis
Bariatric surgery
Vascular surgery
Severe trauma
Head trauma
Polytrauma
Spine trauma
Deformities
Severe scoliosis
Severe hyperlordosis
Idiophatic

With the characteristic symptoms that being very non-specific are often confused with more common pathologies like dyspepsia among others. Therefore, we thought that this entity is extremely underdiagnosed, especially in those patients who despite receiving optimal treatment for other causes that cause similar symptoms, remain symptomatic. For this reason, we recommend that patients with persistent symptoms attributable to upper intestinal obstruction without any cause undergo a CT scan in order to rule out Wilkie syndrome, because despite of being a rare entity it has a potentially correctable cause with a high rate of recovery post-treatment, that could benefit the quality of life of these patients.

CONFLICT OF INTERESTS

The authors have declared no conflicts of interest.

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