

Wilkie's Syndrome in an Adolescent: A Rare Etiology of Upper Intestinal Obstruction

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Abstract

Wilkie's Syndrome (WS) was described in 1927 and its physiopathology is related to the formation of an abnormal acute aortomesenteric angle measuring between 7° and 22°. It leads to digestive symptoms due to external compression of mesentery artery against the third portion of duodenum. This is a case of WS in a young, tall and slim male patient. Three months before, he began postprandial vomiting, abdominal pain, hyporexia and weight loss. The diagnosis was made by an upper gastrointestinal series with barium contrast and confirmed by Laparotomy. Duodenojejunostomy is a well-known technique and it was successfully performed in this case.

Keywords: Wilkie's Syndrome; Superior Mesenteric Artery Syndrome; Mesenteric Duodenal Compression Syndrome; Duodenal Obstruction; Surgery; Duodenojejunostomy

List of Abbreviations: WS: Wilkie's syndrome; CT: Computed tomography; UGS: Upper Gastrointestinal Series; BMI: Body Mass Index; SMAS-like: Superior Mesenteric Artery Syndrome-Like Syndrome

Introduction

Wilkie's syndrome or Superior Mesenteric Artery Syndrome is a rare condition of upper intestinal obstruction. In the general population, the incidence is from 0.013 to 0.3% [1]. This disorder is characterized by extrinsic compression of the third portion of the duodenum due to an abnormal acute angle between the aorta and the superior mesenteric artery, or due to presence of an overlapping of the ligament of Treitz in the retroperitoneum [1]. The syndrome has been known since 1842 and was first described by Carl Von Rokitansky [2]. In 1878, Willet reported a case of death secondary to what he named fatal vomiting [3]. In 1927, Doctor Wilkie described a series of 75 patients with the so-called "duodenal ileus", and 64 of them were submitted to duodenojejunostomy that confirmed the mechanism of extrinsic duodenal compression [4]. In most cases, the onset is insidious with postprandial nausea and vomiting, pain mainly in the epigastrium and weight loss [5]. As to diagnostic exams the gold standard examination is CT angiography and 3D-reconstruction but Upper gastrointestinal series (UGS) is a simple examination and can diagnose this syndrome as well [6]. The surgical treatment of WS should be performed after a conservative treatment that includes fluid and electrolyte replacement, hydration and appropriate nutritional support. The main surgical option is duodenojejunostomy [5,7]. The aim of this study is to report a rare case of upper bowel obstruction (Wilkie's Syndrome) in an adolescent and make important considerations about its pathophysiology, diagnosis and treatment.

Case Report

A male patient, 17-year of age, slim, height 1.8 meters (5 ft 11 in), weight 54 Kg (119 Ib), and BMI of 16.7. The patient presented with postprandial vomiting and abdominal pain, cramps in the mesogastrum which began three months previously and were concurrent with hyporexia and weight loss over 17 kg during that period. He was admitted to the Division of General Surgery of the Hospital de Base, in the city of Porto Velho, Brazil and physical examination showed malnutrition status with emaciation - grade 3 - according to World Health Organization. Laboratory tests revealed some alterations - low hemoglobin and albumin. An abdominal plain CT showed relative distension of stomach and first duodenal portion without evidence of neoplasm or other abdominal abnormalities. After hospital admission, an upper gastrointestinal series was performed and showed an area of stenosis in the third portion of the duodenum, and minimal amount of jejunal loops (Figure 1A and B). The patient was submitted to exploratory laparotomy after parenteral nutrition support for three weeks and a laterolateral duodenojejunostomy was performed

on two plans, in the second portion of the duodenum (Figure 2A and B). He developed extensive pneumonia in postoperative period (fourth day) and ventilatory support in the intensive care unit for two weeks was required. Patient progressed satisfactorily, being discharged four weeks after the surgery. He was followed after the surgical procedure for 2 years with complete reversion of symptoms.

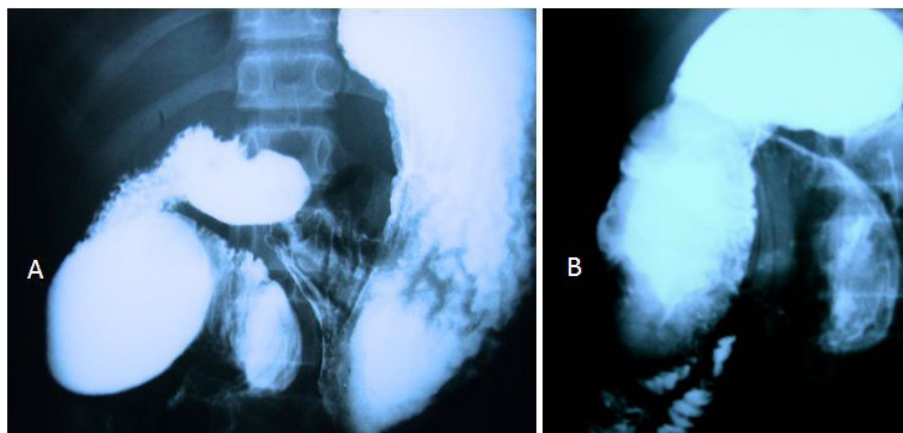


Figure 1: (A) Upper gastrointestinal series (UGS) with an intense gastric and duodenal distention. IT shows the piloric sphincter contraction during the exam; (B) Magnified image of UGS with duodenal distention – first, second and part of third portion - and minimum passage of contrast to proximal jejunal loops

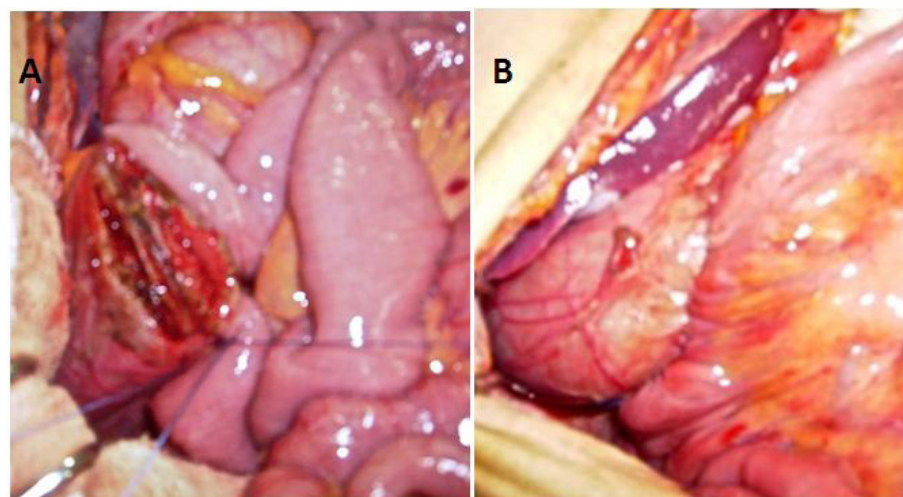


Figure 2: Duodenojejunal anastomosis made in two planes (A) Confection of first plan of anastomosis; (B) Final aspect of duodenojejunal anastomosis

Discussion

WS affects more individuals between 18-35 years with a slight predominance in females but other authors have published WS cases in the male population as well [5]. As to age group, there are reports in children and the elderly [8].

Anatomical characteristics of the region where the mesenteric vessels emerge are crucial to understand the pathogenesis of this syndrome. The origin of the mesenteric artery is in the right anterolateral wall of the abdominal aorta, anteriorly to the vertebra L1, passing anteriorly to the third portion of the duodenum. The duodenojejunal flexure is fixed in the parietal peritoneum, posterior to the ligament of Treitz, and originates from the right diaphragmatic pillar. This flexure involves the fourth duodenal portion and the duodenojejunal junction at L2 vertebra [9]. Normally an aortomesenteric angle ranges from 25° to 60° and is 10 to 28 mm distant from the duodenum [10]. The superior mesenteric artery syndrome results from the formation of an abnormal acute angle measuring between 7° and 22° and may have different causes, such as increased lumbar lordosis or use of orthotics for correcting posture [11]. Another important etiology of this syndrome is the loss of fat tissue close to the superior mesenteric artery and can occur due to malnutrition, malignancy, diabetes mellitus, AIDS, heart diseases or neuropathy, abnormal insertion of the ligament of Treitz, abdominal aorta aneurism or tumor at the root of the mesentery [6,8,12]. An infrequent cause associated to this syndrome is height growth in adolescence with no proportional weight gain [8]. This was probably the cause leading to SW in this reported case.

This syndrome does not always present with specific clinical characteristics, thus reducing initial clinical suspicion. The diagnosis may be delayed, leading to severe malnutrition and associated complications, such as infections [13]. In this case report, the patient developed severe pneumonia after the surgical procedure and one of the factors that interfered in that complication was the

nutritional status [14]. Based on BMI, the patient was classified as having severe emaciation with a BMI of 16.7 kg/m² in addition to other findings of malnutrition.

The onset of symptoms may be acute with intense abdominal pain, persistent vomiting and bloating. In most cases, the onset is insidious with postprandial nausea and vomitus of varied intensity, pain - mainly in the epigastrium - and weight loss. The symptoms may be alleviated in left lateral decubitus and in fetal position [9].

Among the possible differential diagnoses, it is worth mentioning the so-called “superior mesenteric artery syndrome-like syndrome” (SMAS-like) with acute dilation of the duodenal arch of unknown etiology and secondary to some diseases, such as lupus, amyloidosis, dermatomyositis, scleroderma or myxedema. SMAS-like shows no change in the angle or in aortomesenteric distance, or even duodenal compression by the extrinsic mesenteric artery [9,14].

Angiography is the main diagnostic examination and enables measuring the aortomesenteric angle, but the gold standard examination is CT angiography and 3D-reconstruction [6,10]. Several exams can be performed to rule out other causes of upper intestinal obstruction, including computerized tomography of the abdomen, upper oral contrast series and esophagogastroduodenoscopy [6,9]. In this case, abdominal CT did not suggest SMAS and UGS demonstrated duodenal obstruction. It could be because Barium was used in the second exam. It is a heavy material and it could produce increase of dilatation in the digestive tract before the point of obstruction better than other materials.

Surgical treatment of WS should be performed after a conservative treatment that includes fluid and electrolyte replacement, hydration and appropriate nutrition support [14]. Surgery is indicated in cases of failure of clinical management, weight loss with no chance of adequate nutrition recovery, and extensive duodenal dilation [8]. It is crucial to understand the historical sequence of surgical procedures for this syndrome since the first operation until now. The first surgical option was gastrojejunostomy described by Stavely in 1908 [15]. Duodenal mobilization with an ample section of the ligament of Treitz, was described by Strong in 1958 [16]; Massoud was the first surgeon to perform the Strong technique by videolaparoscopy in 1997 [17]. In the following year, Gersin and Heniford conducted the first videolaparoscopic gastrojejunostomy [18]. Gastrojejunostomy with anterior transposition of the third portion of the duodenum is indicated in cases with gastric dilation and/or peptic ulcer that hinder the mobilization of the duodenal arch [19]. Another technique, proposed by Yang, consists of a circular duodenal drainage method with triple anastomosis: duodenojejunal, gastrojejunal and jejunojunal. It is indicated for patients with gastric atony, duodenal dilation greater than 8 cm H₂O or when the intraluminal pressure is above 15 cm H₂O [20]. In this case report, laterolateral duodenojejunosomy was performed through laparotomy but a lot of cases are being reported by laparoscopic approach including robotic assistance as well [5,21]. Duodenojejunosomy represents the major surgical procedure, with approximately 80% resolution and it is easy to perform and enables appropriate inspection of the abdominal cavity.

After operative procedures, some complications can occur in patients with upper intestinal obstruction. One of them is pneumonia and it occurred in this reported case. Some reasons can be related with the pathophysiology of it - use of nasogastric tube, pulmonary micro aspiration, gastric colonization by pathogen bacteria and immunologic system mechanism alterations due to malnutrition [22]. In this case, the patient had severe pneumonia and ventilatory support was needed for ten days. The bacterial pathogen *Enterococcus Spp* was identified by bacteriological culture method. Fortunately the patient had an adequate therapy with antibiotics (Vancomycin) and progressed satisfactorily. His case has been followed for 2 years and he has remained free from symptoms.

Conclusion

Wilkie's syndrome or Superior Mesenteric Artery Syndrome should always be considered in the differential diagnosis of cases suggesting high intestinal obstruction including younger populations. Delayed diagnosis may lead to severe consequences for patients and duodenojejunosomy continues to be the main procedure for this syndrome even though there are several surgical options.

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