

Wilke's syndrome. A rare cause of duodenal obstruction

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Abstract

Wilke's syndrome is a rare cause of partial or complete duodenal obstruction due to exterior compression of the third part of the duodenum by the superior mesenteric artery. It is a true syndrome with characteristic clinical picture which should be included in the differential diagnosis of chronic duodenal ileus. It is initially treated conservatively and then operatively in case of failure of the conservative treatment or if the patient is not willing to follow the medical treatment. We present a case report of Wilke's syndrome with a literature review on this pathologic entity.

Keywords cast syndrome, arterio-mesenteric duodenal compression syndrome, chronic duodenal ileus, superior mesenteric artery syndrome

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Introduction

Wilke's syndrome is a rare partial or complete duodenal obstruction of the third part of the duodenum due to compression of the anterior duodenal wall by the superior mesenteric artery. The syndrome is known since 1842 and many articles on the vascular compression of the duodenum were published in the medical literature either as case reports or as reviews. It is a true syndrome with a characteristic clinical picture which troubles patients suffering from it before a firm diagnosis is established. We present a case of a 15 year old young woman with dehydration after persistent vomiting for some months due to severe loss of body weight, who was diagnosed as suffering from Wilke's syndrome and was successfully treated operatively.

Case report

A 15-year-old female patient was urgently admitted with symptoms of dehydration, loss of appetite and persistent vomiting for 10 weeks. For four months she was vomiting two to three times weekly and this increased to three to four times daily during the last week before admission.

The vomitus consisted of undigested meals eaten recently without any blood. During the last four months the patient reported a significant weight loss (21 kg). Her past medical history was free. On admission the patient was very thin and extremely weak with signs of severe dehydration. A nasophagogastric tube was inserted and the fluid and electrolyte balance was corrected by parenteric infusion of crystalloid fluids with electrolytes. Two days later the patient was subjected to gastroscopy which showed mild inflammation of the lower esophageal mucosa and gastritis. The instrument could not pass further than the third part of the duodenum, due to extraluminal pressure on the duodenal wall. Barium meal follow through confirmed the extrinsic obstruction of the third part of the duodenum and proximal dilation of the stomach and duodenum [Fig. 1] with normal motility of the upper GI tract. CT scan of the upper and lower abdomen excluded the possibility of a tumor or an annular pancreas being the cause of the compression of the duodenum. Thus based on the history, the clinical and imaging findings of the patient, the diagnosis of Wilke's syndrome was concluded.

The patient denied the initially suggested conservative treatment and therefore she was subjected to laparotomy. The first and second parts of the duodenum were dilated and hypertrophied. The dilated duodenum was abrupted at the point where the superior mesenteric artery crossed the third part of the duodenum [Fig. 2]. These findings confirmed the diagnosis and a site to site duodenojejunostomy in two layers was performed [Fig. 3]. The postoperative course was uneventful and barium meal a month postoperatively showed unobstructed passage of the contents from the duodenum to the jejunum. One year later the patient gained the lost weight and remained asymptomatic.

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Figure 1 Barium meal showing the dilation of the first, second and third part of duodenum

Discussion

The compression of the anterior duodenal wall by the superior mesenteric artery (SMA) as a cause of duodenal obstruction was first described by Von Rokitsky at in 1861 on a postmortem case [1] and was studied in detail by D. Wilke in 1912 [2]. The vascular compression of the duodenum received during the years many names over the years, such as

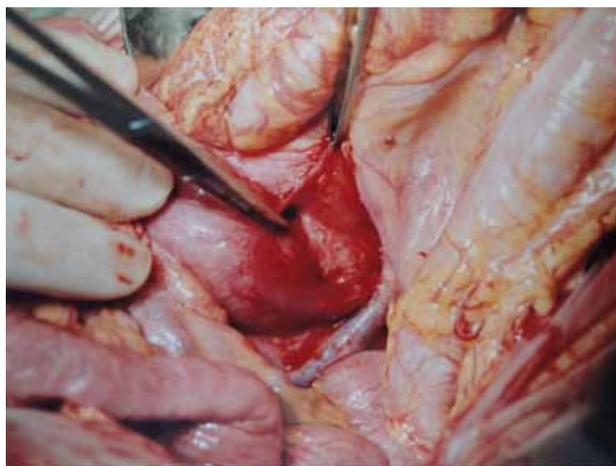


Figure 2 Operative findings of the superior mesenteric artery obstructing the third part of the duodenum



Figure 3 Duodenojejunal anastomosis

superior mesenteric artery (SMA) syndrome, arterio-mesenteric duodenal compression syndrome, the cast syndrome and chronic duodenal ileus. The reported prevalence to in the general population varies between 0.013% and 0.78% [3]. Behind the SMA and in front of the body of L4-L5 vertebrae lies the third part of the duodenum. The narrow part of the SMA contains the uncinat process of the pancreas and the left renal vein. The length and the attachment of the ligament of Treitz, the level at which the duodenum crosses the vertebral column and the level of origin of the SMA from the aorta influences the angle of origin of the aforementioned artery [3,4]. The normal SMA-aorta angle of origin ranges between 20°-70° whereas in Wilkie's syndrome it is very steep ranging from 6° to 15° [4,5]. The Wilkie's syndrome tends to affect young female adults aged between 17-39 years old. Probable predisposing factors include immobilization in the supine position or application of body casts, considerable and rapid weight loss such as in intensive diets or in anorexia nervosa, malrotation or paraduodenal hernias and abdominal aorta aneurysm [6-8]. The main symptoms of Wilkie's syndrome are epigastric discomfort and pain followed by vomiting which as the disease progresses they become more severe, frequent and causing fluid and electrolyte disturbances with weight loss [9].

The diagnosis of the disease is confirmed by radiological imaging [10]. The barium meal will show obstruction of the third part of the duodenum with proximal dilation of the organ and a clear cut line which demarks the obliteration of the duodenal lumen by external compression of the SMA. These radiographic features usually disappear when placing the patient in the knee – chest position as well as after application of pressure at the lower abdomen (Hayes maneuver) [5]. Colored Doppler ultra-sound may help in measuring the angle of origin of the SMA and CT angiography as well as MRI will show the distended first, second and third part of the duodenum as well as the cut off line of the obstructed lumen caused by the SMA [11,12] excluding other causes of duodenal obstruction (tumors, annular pancreas etc).

It is believed by most authors that an attempt for medical treatment should precede operative intervention. Medical treatment may be successful in patients with a short history, moderate symptoms and incomplete duodenal obstruction. It is possible that in patients who are not yet suffering acutely from vascular compression, a high calorie diet which produces gain of weight might alleviate or even remove the symptoms. It consists of frequent meals of fluid or blenderized food, diluted to the consistency of soup and then the patient lying on the left side prone or in the knee - chest position. Total parenteric nutrition has also been used but eventually 50-70% of all cases will come to surgery [13]. In our case, the patient refused the conservative treatment and therefore we immediately proceeded with the surgical treatment.

Surgical intervention is indicated in by failure of the medical treatment, preference of the patient for surgical correction rather than following strict medical treatment and the presence of associated disease such as peptic ulcer and pancreatitis.

The operative options include duodenojejunostomy, section of the ligament of Treiz and relocation of the duodenojejunal junction (operation of Strong), gastrojejunostomy, duodenal anterior replacement [14]. Duodenojejunostomy was first introduced by Starley in 1910 and over the years it has become the most frequent treatment with success rate of 90% [14]. The operation of Strong is mostly indicated in infants but with a high failure rate. Gastrojejunostomy presents the risk of peptic stomal ulceration. All these operative modalities can be laparoscopically approached although experience is limited to case reports and more data is required to recommend this operative option as the standard of care [13,15,16]. Wilkie's syndrome is a rare cause of chronic duodenal ileus with no specific symptoms and diagnosis depends on a high index of suspicion. It is treated conservatively in the early stages while duodenojejunostomy is the operative treatment of choice.

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