

Case Report

Wilkie's syndrome: a rare chronic intestinal occlusion syndrome, to be considered in the differential diagnosis of gastrointestinal neoplasms

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ABSTRACT

Wilkie's syndrome, also known as Cast syndrome, is a very rare vascular disease with an incidence of 0.2-0.7% and most often occurs in women. The decrease of the retroperitoneal perivascular fatty cushions which causes the decrease of the aortomesenteric angle and extrinsic compression of the third portion of the duodenum which generates symptoms of intestinal occlusion and weight loss, in most of the patients this pathology can have a chronic course. We present the case of a 58-year-old male patient, with no relevant history, who presents to the emergency department due to dehydration, involuntary weight loss, abdominal distension, oral intolerance, nausea and vomiting. Medical management was started to improve general conditions, diagnostic protocol was initiated due to clinical picture of intestinal occlusion, and finally a diagnosis of Wilkie's syndrome was established with the help of abdomino-pelvic tomography with intravenous contrast, surgery was performed with exploratory laparotomy, and laterolateral duodenojejunal anastomosis in two planes, the patient had an adequate postoperative evolution with adequate tolerance to the oral route, and was discharged home without complications.

Keywords: Wilkie's syndrome, Cast syndrome, Intestinal obstruction, Midline laparotomy incision, Jejunoduodenal anastomosis

INTRODUCTION

Wilkie's syndrome also known as superior mesenteric artery syndrome (MSA), chronic duodenal ileus or Cast's syndrome.¹ It was first described by Carl Von Rokitsansky in 1861 post mortem, later studied in detail by Wilkie in 1927.⁴ It is a very rare vascular disease with an incidence of 0.2-0.7% and most of the time it occurs in women.²

It is due to the anomalous course of the MRA that originates from the abdominal aorta with an angle of less than 22 degrees. This aortomesenteric angle compresses what lies between the abdominal aorta and the MRA: third portion of the duodenum and the left renal vein, and as a consequence patients present clinical data of intestinal obstruction, postprandial fullness, weight loss and varicocele on some occasions.³

CASE REPORT

Male, 58 years old, with no pathological personal history. Admitted to the emergency department for abdominal distension and pain accompanied by nausea and vomiting on multiple occasions for two weeks of evolution, he reported involuntary weight loss of 12 kg in two months.

The abdominal X-ray on admission showed severe distension of the gastric chamber and intestinal loops, nasogastric tube was placed with spontaneous gastrointestinal output of 2000 cm³ characteristic gastrointestinal. He was admitted to the general surgery floor to continue conservative treatment. Abdominopelvic CT with IV contrast was requested, showing severe distension of the gastric chamber and intestinal loops with no transition site. Endoscopy was performed and

reported: erosive esophagitis of distal third, non-specific erosive pan-gastropathy, permeable duodenum with extrinsic compression of origin to be determined. According to endoscopic findings, the CT scan in arterial phase was analyzed in detail and Wilkie's syndrome was suspected, patient was prepared and surgery was performed.

Exploratory laparotomy was performed, findings: 3rd duodenal portion dilated secondary to obstruction by extrinsic compression of the superior mesenteric artery, intestinal loops distal to the angle of Treitz without apparent alteration, a latero-lateral duodeno-jejunal anastomosis was performed in two planes, without incidents during the surgery. During the postoperative period there were no eventualities, intestinal transit was performed and the enteral diet was started, the patient was discharged home without complications.



Figure 1: Coronal abdominal tomography demonstrating distension of the gastric chamber.

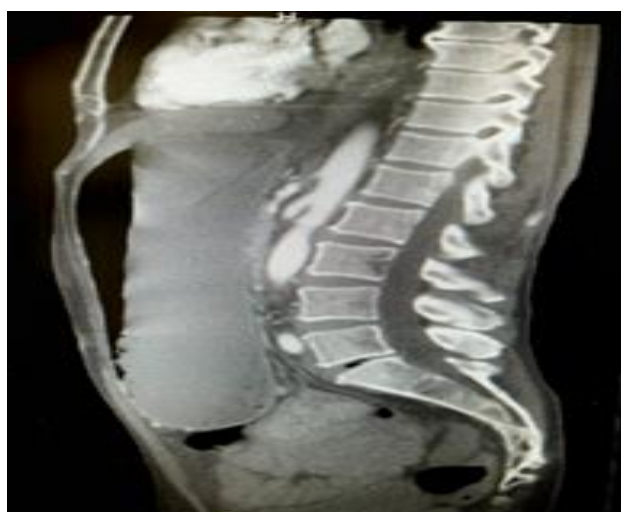


Figure 2: Sagittal abdominal tomography demonstrating distension of gastric chamber and decrease of aortomesenteric angle.

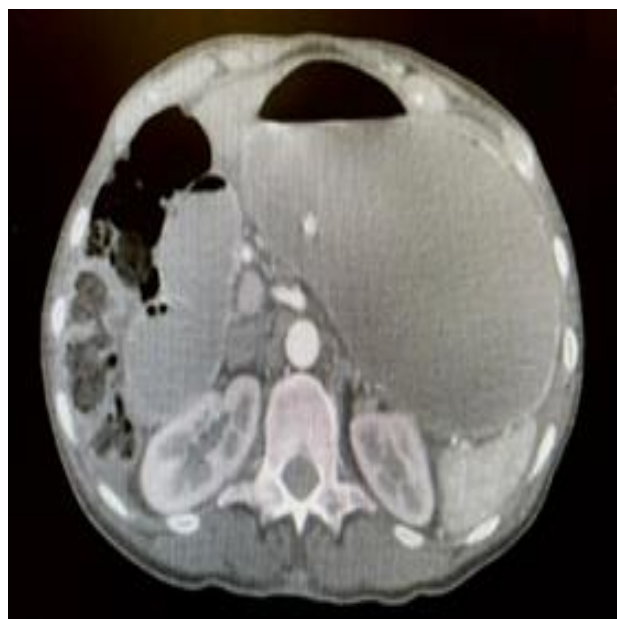


Figure 3: Axial abdominal tomography demonstrating distension of the gastric chamber and decrease of the aortomesenteric angle.



Figure 4: AP abdominal X-ray demonstrating intact gastrointestinal transit.

DISCUSSION

Several causes are responsible for this syndrome, from congenital anomalies to a hypercatabolic state or malnutrition. Regardless of the cause, the decrease of the retroperitoneal perivascular fatty conjuncts which causes a decrease of the aortomesenteric angle and extrinsic compression of the third portion of the duodenum, in spite of this, 40% of the cases are idiopathic. Compression may be partial or complete, acute or

chronic. Physiologically in an upright position, the mesenteric aortic angle measures 38-65 degrees, the normal distance of this angle is 10 to 28 mm, in this syndrome the angle is reduced from 6 to 16 degrees with a distance better than 6 mm.⁵

Due to the low sensitivity and specificity of the symptoms, high suspicion must be maintained, especially in patients who present with postprandial abdominal pain, vomiting, or recent weight loss.⁶

Regarding the diagnosis, the initial abdominal X-ray is not very specific and endoscopy shows extrinsic duodenal compression at the D2 and D3 levels; the gold standard for diagnosis is abdomino-pelvic CT with IV contrast, which shows the exact point of obstruction and rules out other pathologies.¹ Some authors consider that conservative treatment is effective in patients with recent clinical symptoms and partial obstruction; however, there are series that report failure in medical management up to 50-70%. As for surgical treatment, duodenojejunostomy is the procedure of choice with a reported effectiveness of 90%.⁴

CONCLUSION

Wilkie's syndrome has a very low prevalence, the clinical course is insidious and very unspecific, generally other types of neoplasms of the gastrointestinal tract should always be ruled out as was done with this patient, the course is benign and surgical treatment is the treatment of choice in most patients, since conservative treatment presents failures of up to 80%.

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