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Case Report

WILKIE'S SYNDROME – A CASE REPORT

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Abstract

Wilkie's syndrome or Superior Mesenteric Artery (SMA) syndrome is characterized by compression of the third portion of the duodenum caused by a hyperacute angle (<22–25°) between the SMA and the aorta. This leads to the classic presenting symptoms of severe epigastric abdominal pain, postprandial fullness, early satiety and vomiting. The most common factor associated to narrowing of aortomesenteric angle is significant weight loss which leads to loss of retroperitoneal fat. A 70-year-old female presented at the clinic with a 6-month history of recurrent epigastric pain, early satiety and weight loss of 20% in the last 3 years. Past medical history included hysterectomy, appendectomy, hypothyroidism and depressive disorder. Abdominal Computed Tomography (CT) scan revealed SMA syndrome. Conservative treatment failed to relieve symptoms and the patient was submitted to a gastrojejunostomy. Diagnosis of Wilkie's syndrome is challenging and often delayed due to its insidious onset. It should be made based on history and abdominal imaging, especially CT. Treatment is usually conservative including gastric decompression, fluid electrolytes imbalance correction and nutritional support. Surgical intervention is indicated in refractory conditions to relieve symptoms. This case presented with the classic symptoms and imagiological findings and is a reminder of this differential diagnosis in intestinal obstruction..

Keywords: Bowel obstruction; Superior Mesenteric Artery syndrome; Duodenal obstruction.

Introduction

Wilkie's syndrome or Superior Mesenteric Artery (SMA) syndrome is characterized by compression of the third portion of the duodenum caused by a hyperacute angle (<22–25°) between the SMA and the aorta. This leads to the classic presenting symptoms of severe epigastric abdominal pain, postprandial fullness, early satiety and vomiting. Sometimes pain can be aggravated with lying supine and relieved in knee chest position, a maneuver that increases the aortomesenteric angle with subsequent relief of bowel obstruction. [1, 2] It was first described by van Rokitanski in 1861.[3] Incidence of Wilkie's syndrome ranges from 0.1 to 0.3%, and females aged between 10 to 40 years are

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more commonly affected. The most common factor associated to narrowing of aortomesenteric angle is significant weight loss which leads to loss of retroperitoneal fat. Superior mesenteric artery syndrome is frequently associated with severe debilitating illnesses, such as malignancy, malabsorption syndromes, Acquired Immune Deficiency Syndrome, trauma and burns.[2]

Case Report

A 70-year-old female presented at the clinic with a 6-month history of recurrent epigastric pain and early satiety. She had no vomiting or constipation and referred loss of 20% of body weight in the last 3 years. Past medical history included hysterectomy, appendectomy, hypothyroidism and depressive disorder. Physically she was emaciated, hydrated, hemodynamically stable and normothermic. Abdominal examination was painful to epigastric deep palpation without peritoneal signs or palpable masses. Laboratory studies were unremarkable. Esophagogastroduodenoscopy was unremarkable besides gastric stasis. Abdominal Computed Tomography (CT) scan revealed gastric and duodenal distension until its third portion and decreased aortomesenteric angle (17º) and distance (4 mm) (figure 1.). Conservative treatment failed to relieve symptoms and the patient was submitted to a gastrojejunostomy. A Wilkie's syndrome was intraoperatively confirmed (figure 2.). The patient did well in the postoperative period and was asymptomatic after 6 months of follow-up.

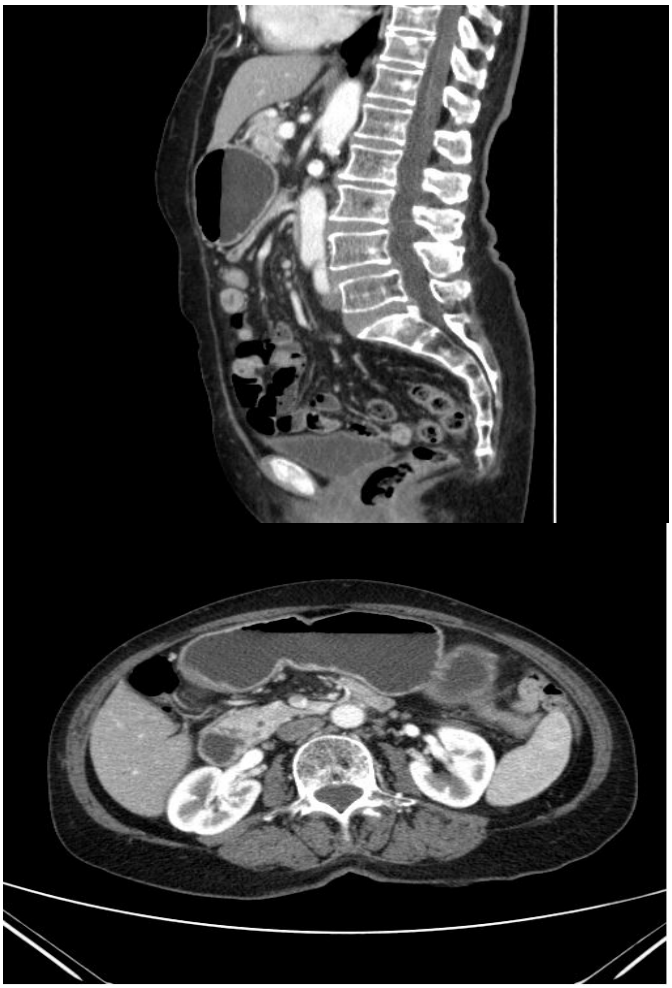

Figure 1: Aortomesenteric angle of 17º and aortomesenteric distance of 4 milimeters with compression of third duodenal portion.

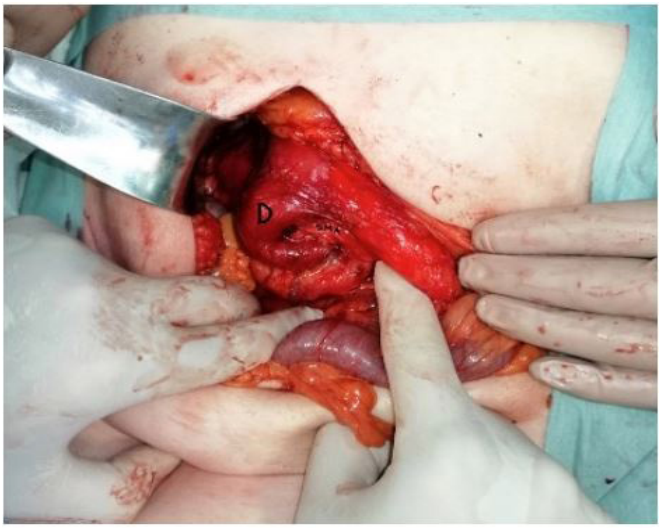

Figure 2: Intraoperative confirmation of Wilkie's syndrome. (D- Duodenum; SMA – Superior Mesenteric Artery).

Discussion

Diagnosis of Wilkie's syndrome is challenging and often delayed due to its insidious onset.[2] It should be made based on history and abdominal imaging, especially CT. It can show vascular compression of the duodenum, with a proximally distended duodenum and stomach, as well as the angulation of the SMA onto the aorta. Barium studies might show duodenal dilatation and sometimes gastric dilatation with slow gastroduodenojejunal transit.[1, 2] Treatment is usually conservative including gastric decompression, fluid electrolytes imbalance correction and nutritional support either through total parenteral nutrition or nasojejunal tube feeding.[2] Ultimately, the goal is restoration of retroperitoneal fat in order to increase the angle and prevent the rare but severe complications, as gastric perforation.[1-3] Surgical intervention is indicated in refractory conditions to relieve symptoms. Duodenojejunostomy is the procedure of choice with a success rate up to 90%. Other surgical procedures like Strong's procedure (division of the ligament of Treitz with mobilization of the duodenum) or a gastrojejunostomy have been reported in literature but are associated with increased postoperative complications like blind loop syndrome and recurrence of symptoms.[1, 2]

Conclusion

This case presented with the classic symptoms and imagiological findings and is a reminder of this differential diagnosis in intestinal obstruction.

Declarations

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Conflict of Interest: All authors declare no conflict of interest to declare.

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