LETTERS TO THE EDITOR

Wilkie's syndrome – a rare cause of gastrointestinal obstruction

To the Editor,

Wilkie's syndrome, also known as superior mesenteric artery (SMA) syndrome, is a rare cause of upper bowel obstruction. It is considered a rare vascular disease, caused by the anomalous angle between the SMA and the abdominal aorta (<22º), responsible for the compression of structures located between both, such as the duodenum and left renal vein.

A 19-year-old female patient underwent to the emergency department multiple times due to nausea, vomiting and weight loss. Full blood count, electrolytes, and albumin were normal. The patient was admitted for further investigation and treatment. An abdominopelvic ultrasonography was performed and revealed a mild left pyelocaliceal dilation without an obstructive cause. She didn't have hematuria or proteinuria, and her creatinine level was within normal range. A computed tomography (CT) scan identified an aorto-mesenteric angle of 18.2° and an aorto-mesenteric distance of 6 mm, compatible with Wilkie's syndrome (Figs. 1a,b). Compression of the duodenum by the SMA was also observed (Fig. 1c). Additionally, the left renal vein was compressed by the superior mesenteric artery. At this point, it was assumed that the symptoms of gastric outlet obstruction were attributable to mechanical duodenal obstruction. First, a conservative approach was tried. As she didn't tolerate enteral nutrition, it was initiated an hypercaloric diet through parenteral nutrition, according to her basal metabolic rate. This treatment was performed for 8 weeks. Although a significative weight gain was achieved, frequent episodes of upper gastrointestinal occlusion occurred when diet was attempted. After this period, a gastroduodenal transit showed a dilated stomach and a mechanical obstruction at the level of duodenum, so it was decided to perform a laparoscopic duodenojejunostomy, which went without complications. She restarted oral feeding with good tolerance and was discharged without symptoms. At present moment, she's waiting for a urology appointment. One month after hospital discharge, she underwent to the emergency department due to a new episode of vomiting. An esophagogastroduodenoscopy was performed, which did not show any abnormality. After this episode, she maintained asymptomatic in subsequent follow-ups.

Wilkie's syndrome is a rare cause of duodenal obstruction, caused by external compression from SMA between this vessel and the aorta. This disease is more frequent in young people

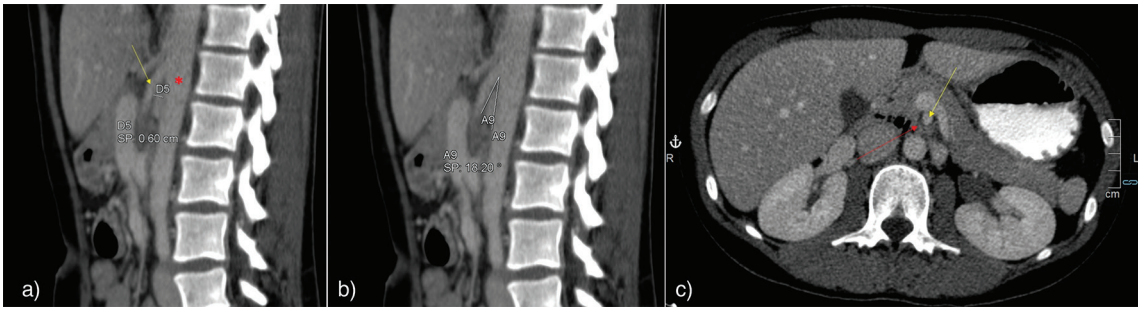

Fig. 1. a) Sagital abdominal CT scan. The aortomesenteric distance corresponds to the distance between aorta (red asterisk) and superior mesenteric artery (arrow). In this patient, this distance is reduced (6 mm). b) Sagital abdominal CT scan. In this patient, the angle is reduced (18,2º), which is typical of Wilkie syndrome. c) Axial abdominal CT scan. Identification of vascular compression of the duodenum (red arrow) by superior mesenteric artery (yellow arrow).

and has a slight female predominance [1]. It may be caused by congenital or acquired conditions. Severe weight loss and subsequent loss of mesenteric fat between the SMA and the aorta is one of the major causes. The weight loss may be due to dietary conditions, hypermetabolism and systemic conditions causing cachexia. Other causes are orthopedic and bariatric surgery, and aortic artery aneurism. Short ligament of Treitz, a low origin of the SMA, spinal deformity and malrotation of the intestine or SMA are possible congenital causes [2]. Symptoms are non-specific and depend on the grade of duodenal compression, and include nausea, vomiting, early satiety, postprandial discomfort, bloating and weight loss. Symptoms may present acutely, as a life-threatening dilation of the stomach, or chronic [3]. Diagnosis is made based on clinical symptoms and radiological evidence of the disease. In recent years, CT scan, MRI, abdominal ultrasonography, endoscopy, and endoscopic ultrasonography have been used to diagnose this disease. CT scan is considered as a reference standard for establishing the diagnosis, as it can demonstrate the aortomesenteric angle (and distance accurately), gastroduodenal dilation and vascular compression of the duodenum. The normal aortomesenteric angle is between 38 and 56º. When it decreases to 6-22º, extrinsic compression of duodenum can occur. The normal aortomesenteric distance is 10-28 mm and decreases to 2-8 mm in SMA syndrome [4]. Treatment may be conservative or surgical. Conservative management consists of a high-calorie diet and is usually advocated as a first approach, although this is based on limited evidence. Precipitating factors should be identified and treated. Surgical therapy is indicated when conservative management fails. The most traditional surgical approach was bypass of the 3rd duodenal part by the creation of a duodenojejunostomy with division of the ligament of Treitz. A laparoscopic approach has been successfully implemented with satisfactory results [5-7]. Duodenojejunostomy was our surgical team choice due its proven efficacy in relieving duodenal obstruction and its low recurrence rate. Gastrojejunostomy was not selected due to a higher risk of complications compared to duodenojejunostomy, particularly afferent loop syndrome and inadequate gastric emptying. SMA syndrome management, involving prior medical therapy followed by surgery, can be performed in a single hospitalization, yielding favorable outcomes in intermediate and long-term follow-up. While physiologically compromised patients require intensive preconditioning, they achieve excellent long-term results [8].

Mesenteric artery syndrome is a serious and rare condition that should be rapidly diagnosed and treated to prevent complications. Due to non-specific signs and symptoms, this clinical entity should be suspected in patients with rapid weight loss or other specific risk factors. Early diagnosis and treatment are essential to a successful outcome.

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Endoscopic ultrasound-guided drainage with lumen-apposing metal stents: a colorful spectrum of solutions

To the Editor,

Due to the continuous evolution of technologies and dedicated devices, in few years endoscopic ultrasound (EUS) has transitioned from a diagnostic method to a therapeutic one. This evolution has led EUS to be essential not only in diagnosis but also in the treatment of more complex patients, thus becoming an alternative to traditional surgical or radiological approaches [1, 2]. It involves demanding endoscopic procedures that necessitates cognitive and technical skills that go beyond those typically required for standard endoscopic practices [3]. Interventional EUS is now an accepted treatment modality for drainage of various locations, such as abdominal collections, bile ducts, gallbaldders and, more recently, small bowel loops [4]. Although radiologic and ultrasound guidance play a crucial role in procedural accuracy, the most immediate confirmation for the operator remains the visual outflow of fluid from the target organ, with color and consistency varying by intervention type (Fig. 1). This observation is crucial for confirming the patency and proper positioning of the stent, indicating a successful connection between the targeted organs.

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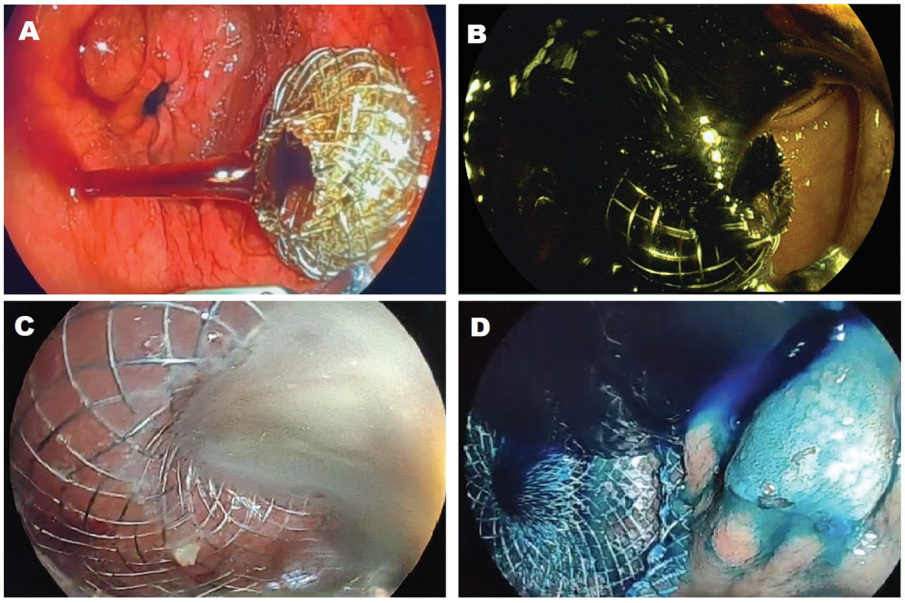

Fig. 1. A) Yellow-orange bile flowing across the stent in EUS-guided gallbladder drainage for acute cholecystitis. B) Cholestatic black bile seen in choledoco-duodenostomy for malignant jaundice. C) Thick, whitish discharge of pus during drainage of an infected peripancreatic collection. D) Dilute methylene blue flowing across the stent in gastro-enterostomy.

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A rare case of endoscopic ultrasound-guided fine needle aspiration biopsy complicated with acute pancreatitis and acute appendicitis

To the Editor,

Endoscopic ultrasound (EUS) is an important tool in modern gastroenterology allowing for diagnostic imaging of the intestinal wall and pancreaticobiliary system. Another clinical use of EUS is tissue acquisition via EUS-guided fine needle aspiration (EUS-FNA) biopsy [1]. The morbidity associated with EUS-FNA is low, and the most common complications of EUS-FNA include infection, bleeding, and acute pancreatitis [2]. Below we present a rare case of EUS-FNA complicated with acute pancreatitis and acute appendicitis. To our knowledge, such a case has not yet been reported.

A 67-year-old male was admitted to our department for EUS-FNA due to a suspected focal lesion of the major duodenal papilla (papilla of Vater). On admission, the patient complained of only intermittent pain in the right upper quadrant of the abdomen. His medical history included acute pancreatitis, ST elevation myocardial infarction, hypertension, coronary artery disease and nicotine addiction. On examination, the patient was in good condition and slightly jaundiced. The patient underwent EUS examination with EUS-FNA biopsy, which revealed a fragment tubular adenoma with low-grade dysplasia. After the procedure, the patient reported severe "tightening" pain in the upper abdomen and nausea. The abdomen was bloated and painful on palpation, with local rigidity; peristalsis was present. His blood pressure (BP) was 152/93 mmHg, heart rate (HR) was 85/min, and the peripheral oxygen saturation (SpO2) was 99%. The patient was administered buprenorphine and diclofenac, which relieved the pain with good effect. A strict diet was ordered. Elevated serum amylase level confirmed the diagnosis of acute pancreatitis. On the following day, the patient's vital signs worsened, BP was 115/73 mmHg, HR was 82/min and SpO2 was 91% on 2 liters of O2 through a nasal cannula. The patient was started on piperacillin with tazobactam. A computed tomography (CT) scan of the abdomen was suggestive of contained perforation of the duodenum or the biliary tract. It also revealed extrahepatic bile duct dilatation, thickening of the duodenal wall in the vicinity of the papilla of Vater and inflammatory infiltration involving the appendix. In the following hours, his condition worsened, diffuse abdominal rigidity and other signs of peritonitis were observed. The patient

was subsequently transferred to the operating room (OR). During the surgery, no perforation was found. The inflamed appendix was removed, and the post-operative histopathological examination confirmed acute appendicitis.

It is impossible to distinguish whether the appendicitis had a primary or secondary character. Usually, acute appendicitis occurs in the 10-30 age group and approximately 90.7% of cases occur before the age of 65 years [3, 4]. On the one hand, these statistics seem to favor the theory of the secondary nature of the described appendicitis. On the other hand, the possibility of time coincidence between appendicitis and EUS-FNA and pancreatitis cannot be excluded.

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A rare case of small intestine ulcers, bleeding, obstruction, and perforation caused by amyloidosis

To the Editor,

A 57-year-old male presented with abdominal pain, abdominal distention, and cessation of flatus and stool for a duration of 10 days. Before admission, he received conservative treatment for intestinal obstruction at his local hospital, but he found no significant relief. Therefore, an intestinal obstruction catheter was inserted. Physical examination after hospitalization showed slight tenderness in the lower abdomen. Laboratory tests revealed red blood cells 4.16 x 10^12/L, hemoglobin 12 g/dL, and albumin 34.2 g/L. Contrast-enhanced computed tomography (CT) scan showed thickening of the small intestine wall, intestinal dilation, and visible fluid accumulation, with slight thickening of the ascending colon, descending colon, and sigmoid colon walls, and slight thickening of the gastric wall. Colonoscopy revealed scattered congestion in the terminal ileum, ascending colon, and rectal mucosa, multiple superficial ulcerative lesions, and surrounding mucosal congestion and edema, with no diagnostic abnormalities found in pathology (Fig. 1A). Gastroscopy showed chronic superficial gastritis and duodenitis. The patient was treated with fasting, gastrointestinal decompression, suppression of digestive fluid secretion, enemas, oral mesalazine granules, and nutritional support. Follow-up abdominal X-ray showed intestinal obstruction.

On the 10th day of hospitalization, after obtaining the patient‘s written informed consent, a pre-oral double-balloon enteroscopy (DBE) was performed, revealing multiple ulcers and strictures (Fig. 1A), which led to difficulties in endoscope insertion and resulted in intestinal perforation. An emergency surgery was carried out, removing approximately 200 centimeters of the small intestine (Fig. 1B).

Postoperative pathology revealed extensive plasma cells infiltration and stromal hemorrhage within the mucosa, significant homogeneous deposition in the submucosa and perivascular areas, along with obvious submucosal hemorrhage, edema, and degeneration of the muscularis propria (Fig. 1C). Special staining showed Congo red (+), and typical apple-green birefringence observed under polarized light (Fig. 1D), which is the pathological diagnostic criterion for amyloidosis. Ultimately, he was diagnosed with primary localized intestinal amyloidosis, which is a rare cause of hemorrhagic enteritis.

He received glucocorticoids, enteral nutrition, as well as symptomatic treatment, but he still experienced recurrent peptic ulcers and bleeding. The patient refused bone marrow biopsy to clarify the type of amyloidosis. After 121 days of hospitalization, he gave up further treatment and then discharged. Unfortunately, he died six days after discharge.

Small intestinal amyloidosis is a rare condition characterized by the accumulation of misfolded proteins in the intestinal tissue, leading to both structural and functional damage [1]. Classification of amyloidosis is based on the chemical analysis of the protein deposited, the most common types affecting humans being immunoglobulin-light-chain related amyloidosis or primary (AL), amyloid A or reactive/secondary amyloidosis (AA) [2]. While amyloidosis generally affects multiple organs, especially heart and kidneys, its involvement in the intestines is less common [3]. Its related symptoms can vary, and often include weight loss, chronic diarrhea, abdominal pain, and dyspepsia [4]. In cases of small intestinal involvement, abdominal pain and obstruction are the most prominent symptoms, with bleeding also being a notable concern.

Diagnosis and treatment of small intestinal amyloidosis pose significant challenges, because its symptoms often resemble those of other gastrointestinal diseases, like inflammatory bowel disease or malignant tumors [5]. This similarity increases the risk of misdiagnosis. A definitive diagnosis usually requires an endoscopic examination and biopsy, which can confirm the presence of amyloid material using Congo red staining [6]. However, neither endoscopic examination nor Congo red staining can effectively differentiate the subtype of amyloidosis. The most reliable approaches for identifying the types of amyloid proteins include immunohistochemistry, immunoelectron microscopy, or mass spectrometry [3]. Until now, there is no specific therapy for small intestinal amyloidosis.

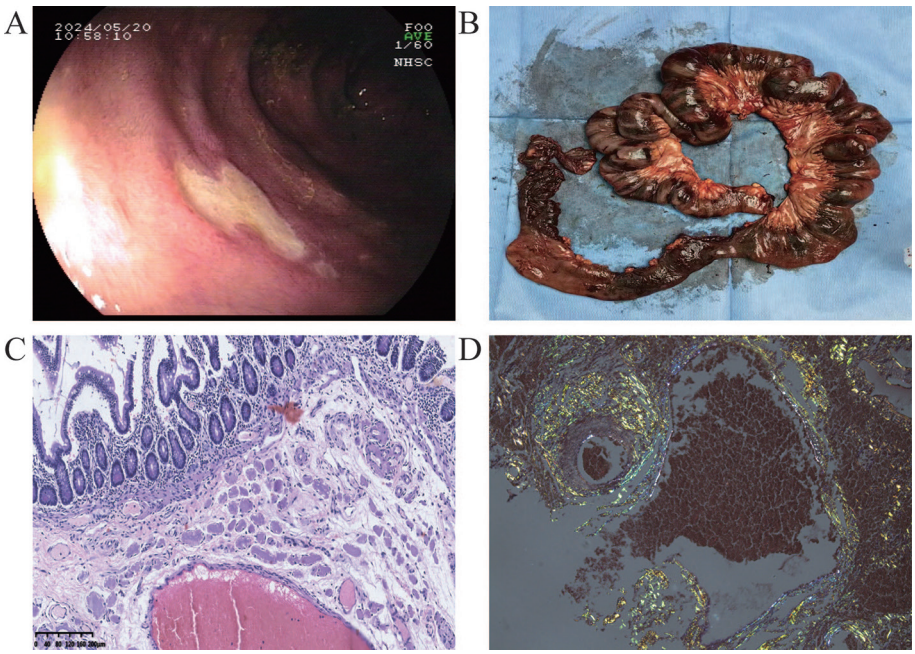

Fig.1. A) Pre-oral DBE revealed multiple ulcers with strictures in the duodenum and jejunum. B) Image of the resected intestinal segment with a length of about 200 centimeters. C) Pathological H&E staining. D) Special staining showed Congo red (+), and typical apple-green birefringence was observed under polarized light.

In summary, small intestinal amyloidosis is complex, but often overlooked. Effective diagnosis and treatment require the collaboration of a multidisciplinary team. Early identification and proper management should be potentially critical for improving the patients' quality of life.

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