Case Report

Case report: Wilkie's syndrome presenting as persistent vomiting, severe malnutrition and avoidant/restrictive food intake disorder; a late post-op complication of scoliosis surgery and a clinical challenge

Caso Clínico: Síndrome de Wilkie que se presenta como vómitos persistentes, desnutrición grave y trastorno de evitación/restricción de la ingesta de alimentos; una complicación postoperatoria tardía de la cirugía de escoliosis y un desafío clínico

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Abstract

Sixteen-year-old male with previous surgery for severe scoliosis, 18 months before the onset of symptoms, consults due to five weeks of epigastric pain and vomiting after acute, self limiting diarrhea. Patient lost eight kilograms. Endocrinologic, metabolic causes and celiac disease were ruled out. Barium esophagogram showed a slight dilatation of duodenum bulb. A new upper gastrointestinal endoscopy confirmed these findings and a pulsatile vessel was found, which could correspond to superior mesenteric artery. A clip was installed to mark the site. A computed tomography scan was performed, showing a clear point of lumen stenosis with proximal dilatation. Wilkie's Syndrome was diagnosed. A laparoscopic duodenal-jejunal anastomosis was performed. The patient had complete remission of symptoms, weight gain and was able to go back to school.
Key words: Wilkie's syndrome, Superior Mesenteric Artery Syndrome, bowel obstruction, vomiting, scoliosis surgery.

Resumen

Paciente masculino de 16 años, con cirugía previa por escoliosis severa, 18 meses antes del inicio de los síntomas, consulta por dolor epigástrico de cinco semanas y vómitos, posteriores a diarrea aguda autolimitada. El paciente perdió ocho kilogramos. Se descartaron causas endocrinológicas, metabólicas y enfermedad celíaca. El esofagograma baritado mostró una ligera dilatación del bulbo duodenal. Una nueva endoscopia digestiva alta confirmó estos hallazgos y se encontró un vaso pulsátil, que podría corresponder a la arteria mesentérica superior. Se instaló un clip para marcar el sitio. Se realizó una tomografía computarizada, que mostró un punto claro de estenosis de la luz con dilatación proximal. Se diagnosticó síndrome de Wilkie. Se realizó una anastomosis duodeno-yeyunal laparoscópica. El paciente tuvo remisión completa de los síntomas, aumento de peso y pudo regresar a la escuela.
Palabras clave:

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Abbreviations

ARFID: Avoidant/Restrictive Food Intake Disorder.

CT: Computed tomography.

SMA: superior mesenteric artery.

TBE: Timed Barium Esophagogram.

WS: Wilkie's Syndrome.

UGE: upper gastroesophageal endoscopy.

Introduction

Wilkie's Syndrome (WS), is a rare cause of duodenal obstruction, often intermittent, dynamic, partial or complete, acute or chronic, caused by the compression of the duodenum by the superior mesenteric artery (SMA)[1]. The incidence in the general population is difficult to calculate, and it is estimated to be around 0.1-0.3%[2], with a slight predominance in women (3:2)[1]. It is caused by the compression of the third portion of the duodenum (D3) by the SMA over the aorta, and a reduction of the aortomesenteric angle from 45° (normal range between 38-56°) to about 6-25°[3,4]. The usual clinical presentations occur in patients who complain of postprandial abdominal pain, distress and voluminous postprandial vomiting, leading to weight loss. Contrastenhanced CT-scan can be diagnostic with typical findings of duodenal distension along with narrowing of the aortomesenteric angle and assessment of retroperitoneal fat in a symptomatic patient[5-9]. Scoliosis surgery is a risk factor to the relative lengthening of the spine postoperatively[10,11], decreasing aortomesenteric angle.

Moreover, some cases of WS are easily confused with psychiatric disorders related to food ingestion, such as bulimia, anorexia and Avoidant/Restrictive Food Intake Disorder (ARFID) or functional disorders such as functional dispepsia[1]. ARFID describes a group of patients with avoidant or restrictive eating behaviours unrelated to body image disorder or weight loss desire[12,13].

Case presentation

A 16 year-old male, with corrective surgery due to severe scoliosis (T3-L4 arthrodesis) which increased his height by 5 cm about 1.5 years before current symptoms. He presented to an ambulatory gastroenterology unit with a history that began 5 weeks before, characterized by postprandial epigastric pain and vomiting up to 56 times per day, and in the first week of symptoms he had reported self-limited diarrhea. During this time, the patient lost 8 kilograms of weight. Symptoms were severe as to cause school absenteeism. The main finding of the physical exam was weight loss (clinical sarcopenia and loss of fatty tissue). A first upper endoscopy did not find obstructive causes. Due to systemic involvement, the patient was hospitalized in order to improve his nutrition status.

In the hospital, endocrine, metabolic and celiac disease causes were ruled out. CT enteroclysis and brain MRI were normal. Also, a gastric emptying scintigraphy for solids was within normal limits. Psychiatric team evaluated the patient and they proposed a ARFID in the context of external stressors and personality traits. Patient was discharged with an olanzapine prescription in order to complete monitoring and pending tests.

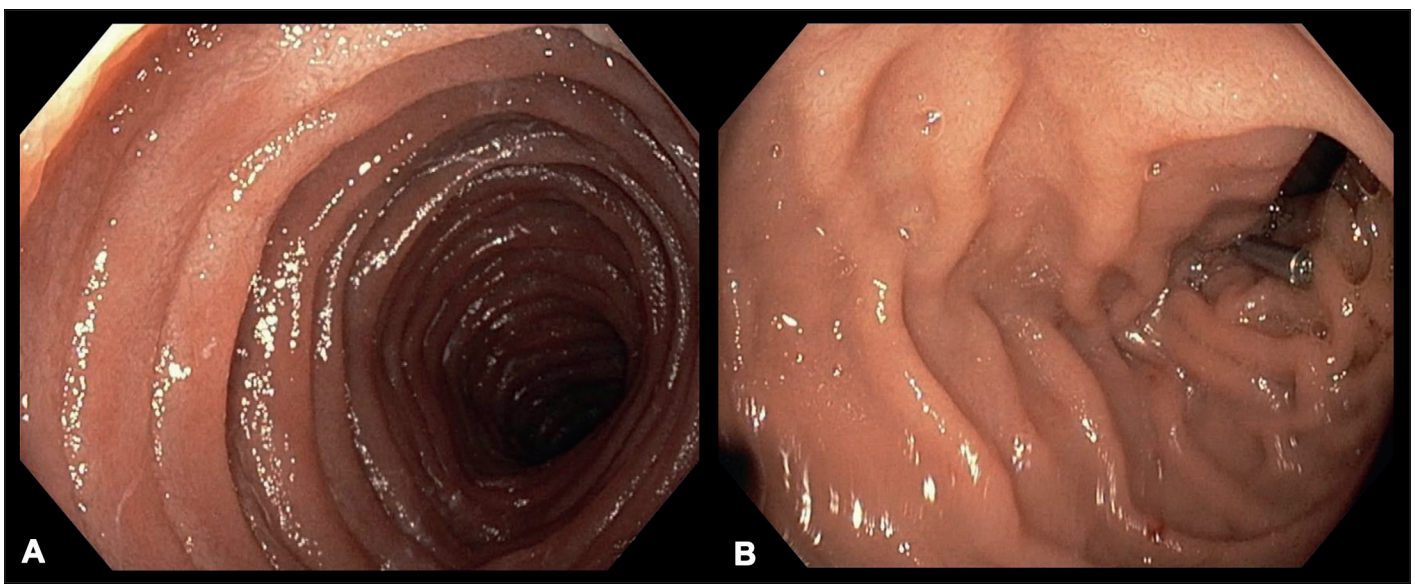

Figure 1. Upper Gastroesophageal Endoscopy. **A:** observed D2 and D3 portion of the duodenum dilated. **B:** Placement of Blip under SMA impression.

Three months later, the patient was readmitted to hospital because he continued with postprandial vomiting. He did not attend ambulatory medical appointments and pH-impedance monitoring could not be performed as the patient did not tolerate the pH-impedance probe. At this point, the patient had lost 15 kilograms of weight, and he had clinical signs of dehydration and lipothymia. The patient required parenteral nutrition initially due to severe malnutrition. Remarkably, the patient showed no aversion to weight gain, body dysmorphia or a desire to stop eating, but was reticent to eat due to a fear of vomiting. There was a lack of response with antiemetics and prokinetics. A second upper endoscopy and biopsies were performed, without relevant findings. Time barium esophagogram (TBE) was informed without alteration.

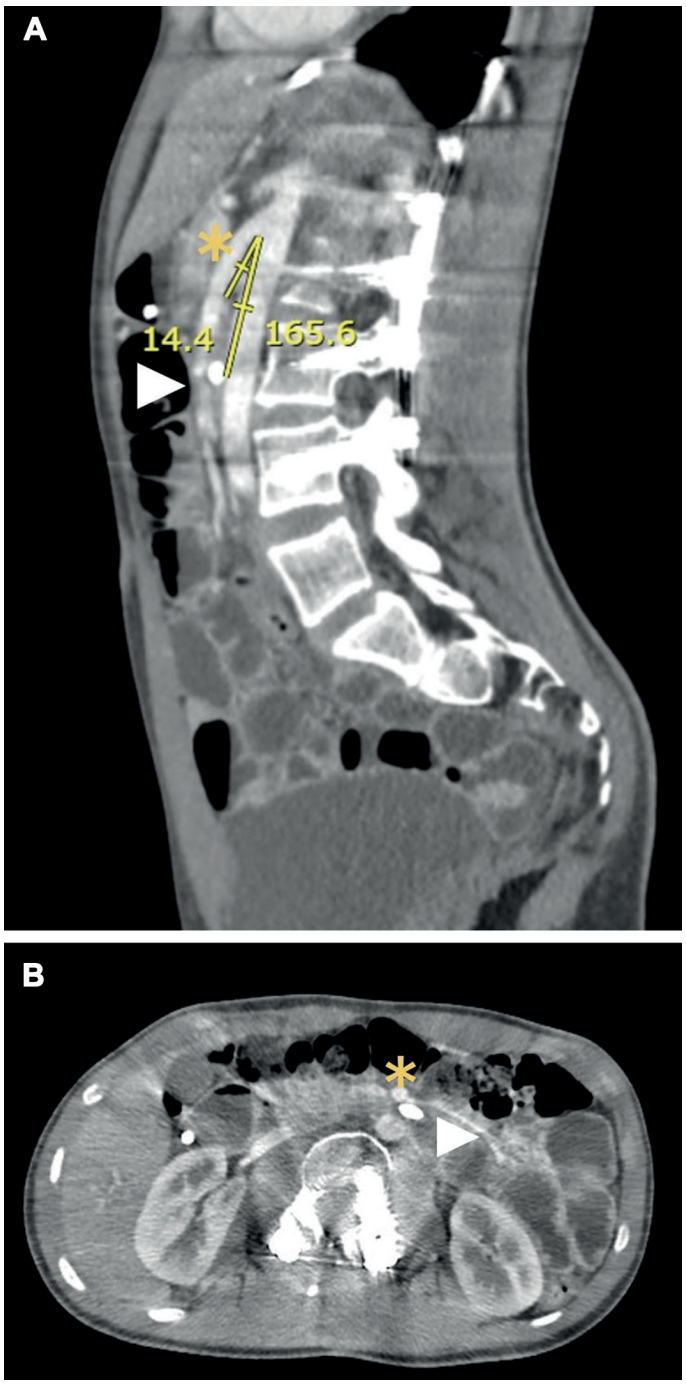

Figure 2. CT with Enteroclysis protocol. **A.** Sagittal view, **B.** Axial view. Yellow asterisk: SMA White triangle: third portion of duodenum with nasoenteric feeding tube in lumen.

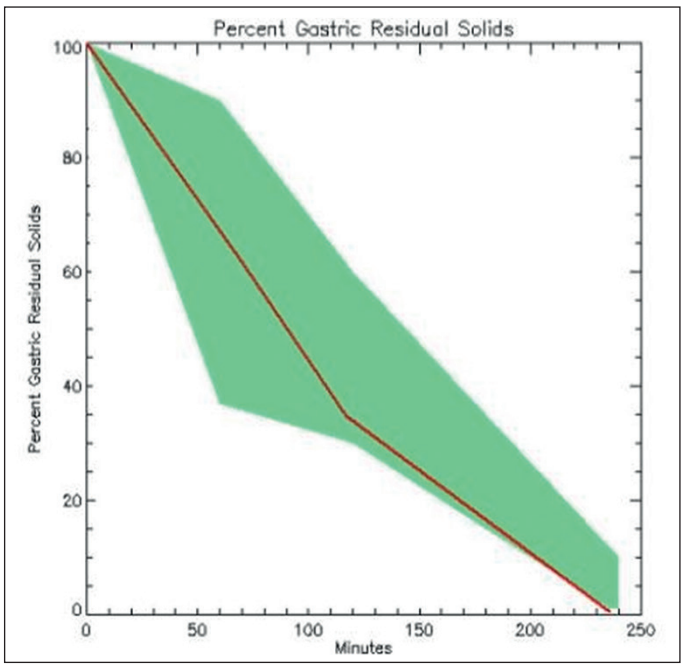

Fig 3.a (first hospitalization). Gastric Emptying Centigram: green demarcated area corresponds to normal values. Red line corresponds to patient results.

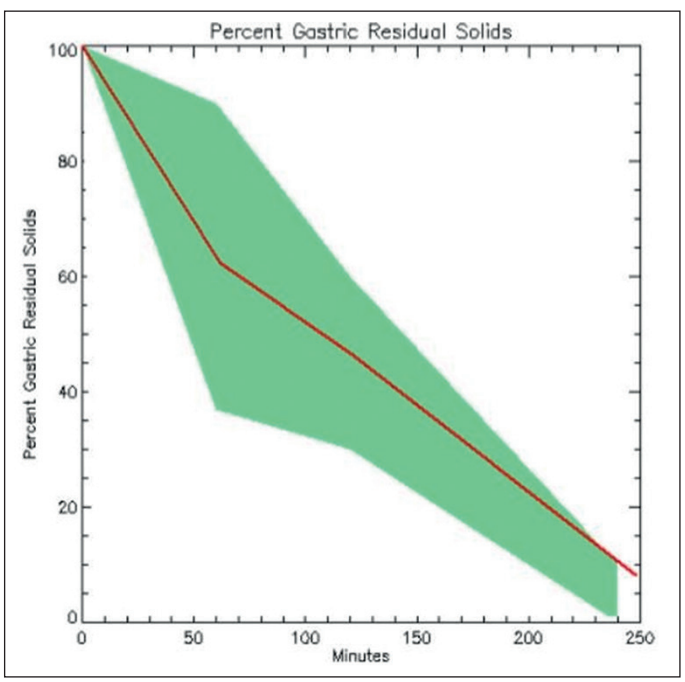

Fig 3.b (second hospitalization). Gastric Emptying Centigram: green demarcated area corresponds to normal values. Red line corresponds to patient results.

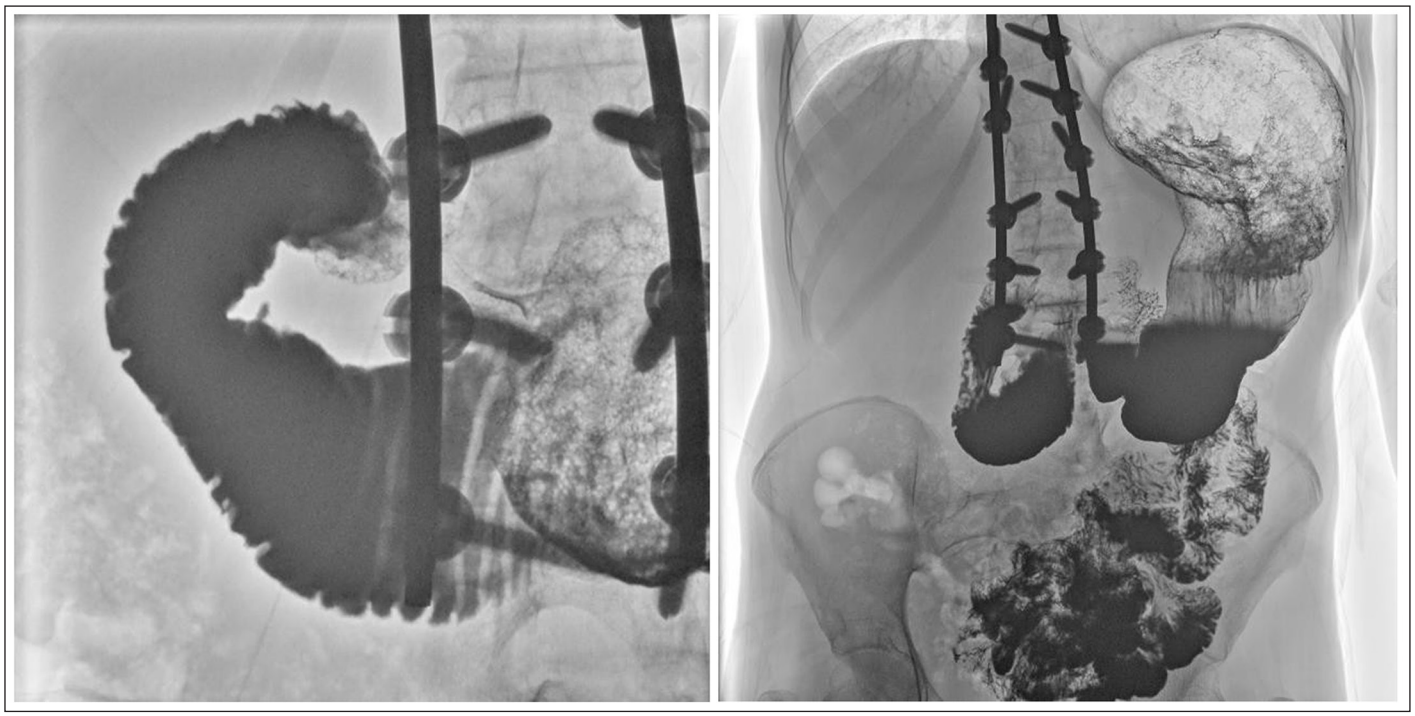

Fig 4. Timed Barium Esophagogram, of note D2 and D3 of the duodenum are dilated with tapering in and cut-off of contrast.

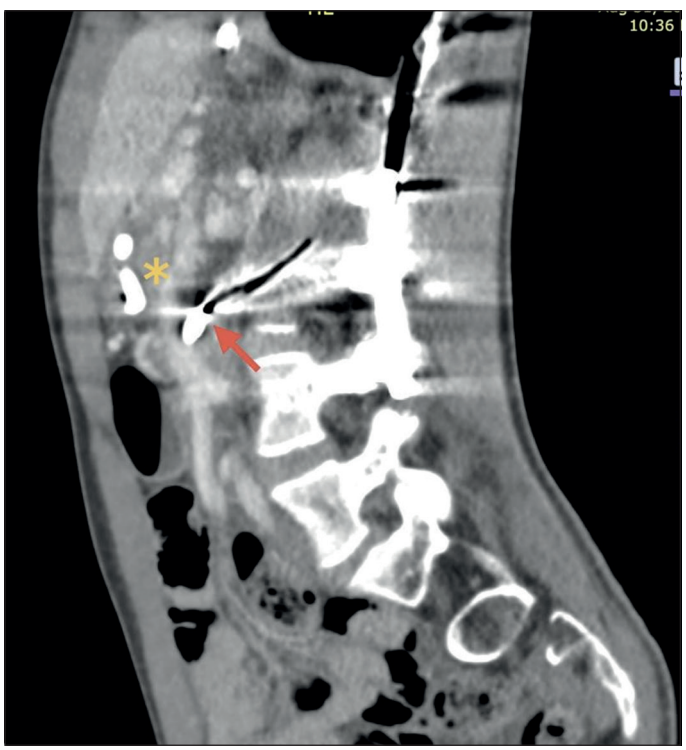

Fig 5.a. CT post placement of clips in D3. A. Sagital view: Yellow asterisk: Superior mesenteric artery. Red Arrow: endoscopic clip. SMA is seen over duodenum.

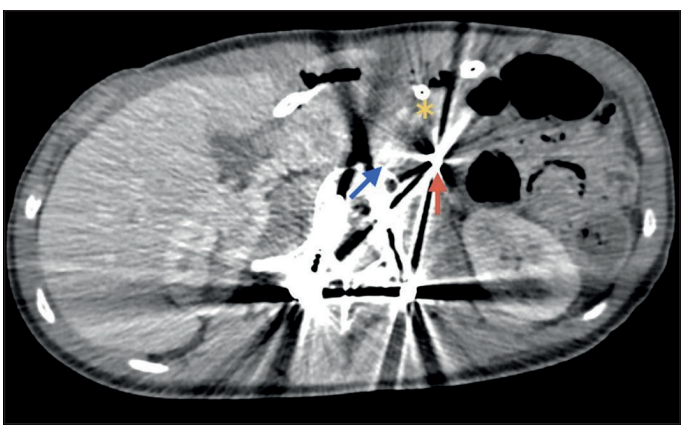

Fig 5.b. CT post placement of clips in D3. Axial view Asterix: SMA, Red arrow: endoscopic clip Blue Arrow: aorta.

Enteral nutrition was prescripted with good tolerance with nasojejunal feeding tube without vomiting. When the tube was repositioned in the gastric chamber, the patient again began vomiting. Due to this observation, TBE was reevaluated and showed a slight dilation of the duodenum. Multidisciplinary team proposed SMA syndrome. A new upper gastrointestinal endoscopy showed a slight dilation of the duodenum bulb and distal to the second portion of duodenum.

Table 1.

Test	Result	Normal Values
Hemoglobin	12.2 mg/dL	13.0 - 16.0 mg/dL
WBC	5.100 /mm^3	4.500 - 11.000 / mm^3
Platelets	265.000/mm^3	140.000 - 400.000/mm^3
Serum Creatinine	0.75 mg/dL	0.7 - 1.2 mg/dL
Sodium	140 mEq/L	135 - 145 mEq/L
Potassium	4.1 mEq/L	3.5 - 5.9 mEq/L
Calcium	9.2 mg/dL	8.5 - 10.5 mg/dL
Total Bilirubin	1.11 mg/dL	< 1.0 mg/dL
AST	18 U/L	< 39 U/L
ALT	24 U/L	< 30 U/L
Alkaline phosphatase	96 U/L	52 - 171 U/L
GGT	10 U/L	< 60 U/L
Albumin	4.0 g/dL	3.5 - 5.0 g/dL
Lipase	32 U/L	13 - 60 U/L
Amilase	43 U/L	28 - 100 U/L
TSH	1.57 uUI/mL	0.7 - 5.7 uUI/mL
Vitamin D*	14 ng/mL	20 - 50 ng/mL
Vitamin B12*	731 pg/mL	271 - 1136 pg/mL
Ferritin*	268 ng/mL	30 - 400 ng/mL
TSAT*	69%	16 - 45%
anti-Trypanosoma cruzi*	Non reactive	< 0.8 (non reactive)
HIV ELISA*	Negative	< 1.0 (negative)
HBV SAg*	Non reactive	< 10 mUI/mL (non reactive)
HBA IgM and IgG*	Non reactive	< 0.8 (non reactive) / < 1.0 (non reactive)
IgA	156 mg/dL	61 - 348 mg/dL
Anti-Transglutaminase Antibodies	Negative	< 12 U/mL (negative)
Anti neutrophil antibodies	1/160 nuclear granular pattern	> 1/80 (negative)
Extractable Nuclear Antigen Antibodies (ENA) Panel	Negative	< 20 U (negative)
Prolactine	36.9 ng/mL	4.04 - 15.2 ng/mL
Follicle-stimulating hormone	1.9 mUI/mL	1.5 - 12.4 mUI/mL
Basal Cortisol	5.2 µg/dL	6.02 - 18.4 ug/dL
Post ACTH Cortisol	21 µg/dL	> 20 ug/dL
ACTH	10 pg/mL	10 - 60 pg/mL
Thyroxine	1.39 ng/dL	0.8 - 2.0 ng/dL
DHEA-SO4	0.74 ug/mL	0.8 - 5.6 ug/mL
Androstenediona	1.3 ng/mL	1.1 - 3.1 ng/mL
Testosterona total	810 ng/dL	180 - 763 ng/dL
SHBG	51 nmol/L	16.5 - 55.9 nmol/L
Somatomedina C	225 ng/mL	173 - 414 ng/mL
Upper Endoscopy Biopsies results*	Esophagus: nonspecific findings compatible with acid reflux Corpus and Fundus: no pathological findings Antrum: crónica superficial gastritis	Normal

*Studies performed during second hospitalization.

A duodenal fold with a pulsatile blood vessel was observed, which appeared to correspond to the transmission of the SMA pulse. A clip was installed at this point to mark the site. Then, a new CT scan was performed using the clip as a guide, showing a clear point of lumen stenosis with proximal dilation (Fig 5). The case was evaluated in a multidisciplinary meeting, where the clinical, serological and radiological findings were deemed sufficient to diagnose SMA syndrome and a surgical approach was offered to the patient and his parents, due to the lack of sufficient response to enteral feeding. A laparoscopic duodenal-yeyunal anastomosis was successfully performed with a preventive jejunostomy. During surgery the first and second portion of the duodenum was dilated and the ligament of Treitz was observed to be high. The postoperative period was uneventful, the patient successfully reached his caloric goals with oral nutrition progressively, and was discharged about two weeks after surgery. Two weeks later the patient went to their post discharge medical appointment and was asymptomatic, eating normally per oral, and had gained 6 kilograms. About 6 weeks after discharge, the patient had recovered 12 kilograms and jejunostomy was removed. Patient lost aversion to eating and he came back to school after several months.

Discussion

Our case is of particular interest for two main reasons. First, symptoms began near the beginning of the school year, making the possibility of a psychiatric disorder higher. It also occurred about 18 months after scoliosis surgery (rates after corrective spinal surgery reported between 0.5% and 2.4%). Secondly it was triggered by an acute gastroenteritis that caused enough weight loss to initiate the symptoms. In this case, the barium contrast image of the duodenum was a helpful tool to find the diagnosis, and a multidisciplinary team was very important to reach the diagnosis. The principal key to getting the diagnosis was the clinical sign of vomiting when the feeding tube was localized in the stomach in opposition to when it was localized in the jejunum. There are scarce medical options for this entity, and changes in diet with small, low consistency foods greatly affect the patients quality of life. Therefore, surgical options in these cases are extremely effective and should always be considered if there is no response to nutritional therapy. Symptomatic response is quick and post-operative care is short.

Informed Consent: the patient and his legal guardians (his mother) were appropriately informed and consented to the presentation of this case report, with considerations regarding identity and private or personal information. (Supplement 1).

References

1. Oka, A. *et al.* Superior mesenteric artery syndrome: Diagnosis and management. *World J Clin Cases* 11, 3369–3384 (2023).
2. Shiu, J.-R. *et al.* Clinical and nutritional outcomes in children with idiopathic superior mesenteric artery syndrome. *J. Pediatr. Gastroenterol. Nutr.* 51, 177–182 (2010).
3. Zachariah, S. K. Wilkie's syndrome: a rare cause of intestinal obstruction. *BMJ Case Rep.* 2014, (2014).
4. Hadi, S. S., Kareem, T. F. & Kamal, A. M. Normal values of angle and distance between the superior mesenteric artery and aorta in Iraqi population: A single centre study. *J Med Radiat Sci* 69, 191–197 (2022).
5. Kerdiles, T. & Gras, J. Superior Mesenteric Artery Syndrome. *N. Engl. J. Med.* 389, 359 (2023).
6. Lundell, L. & Thulin, A. [Wilkie's syndrome--a rarity?]. *Lakartidningen* 77, 4431–4433 (1980).
7. Laffont, I. *et al.* Late superior mesenteric artery syndrome in paraplegia: case report and review. *Spinal Cord* 40, 88–91 (2002).
8. Warncke, E. S., Gursahaney, D. L., Mascolo, M. & Dee, E. Superior mesenteric artery syndrome: a radiographic review. *Abdom Radiol (NY)* 44, 3188–3194 (2019).
9. Moramarco, M., Borysov, M., Ng, S. Y. & Weiss, H.-R. *Schroth's Textbook of Scoliosis and Other Spinal Deformities*. (Cambridge Scholars Publishing, 2020).
10. Mathenge, N. *et al.* Superior mesenteric artery syndrome and its associated gastrointestinal implications. *Clin. Anat.* 27, 1244–1252 (2014).
11. Welsch, T., Büchler, M. W. & Kienle, P. Recalling superior mesenteric artery syndrome. *Dig. Surg.* 24, 149–156 (2007).
12. Brigham, K. S., Manzo, L. D., Eddy, K. T. & Thomas, J. J. Evaluation and Treatment of Avoidant/Restrictive Food Intake Disorder (ARFID) in Adolescents. *Curr. Pediatr. Rep.* 6, 107–113 (2018).
13. Zimmerman, J. & Fisher, M. Avoidant/Restrictive Food Intake Disorder (ARFID). *Curr. Probl. Pediatr. Adolesc. Health Care* 47, 95–103 (2017).
14. Nakagawa, K. *et al.* Persistent Postprandial Regurgitation vs Rumination in Patients With Refractory Gastroesophageal Reflux Disease Symptoms: Identification of a Distinct Rumination Pattern Using Ambulatory Impedance-pH Monitoring. *Am. J. Gastroenterol.* 114, 1248–1255 (2019).
15. Matheus, C. de O., Waisberg, J., Zewer, M. H. de T. & Godoy, A. C. de. Syndrome of duodenal compression by the superior mesenteric artery following restorative proctocolectomy: a case report and review of literature. *Sao Paulo Med. J.* 123, 151–153 (2005).
16. Feinberg, A. N. Case 1. Duodenal compression and obstruction--superior mesenteric artery syndrome. *Pediatr. Rev.* 19, 281–282 (1998).
17. Doski, J. J., Priebe, C. J., Jr, Smith, T. & Chumas, J. C. Duodenal trichobezoar caused by compression of the superior mesenteric artery. *J. Pediatr. Surg.* 30, 1598–1599 (1995).
18. Traore, M. M. *et al.* [Early form of Wilkie's syndrome: a rare complication of scoliosis surgery, about a case and review of the literature]. *Pan Afr. Med. J.* 25, 90 (2016).