

# Aorto–mesenteric compass syndrome (Wilkie's syndrome) in the differential diagnosis of chronic abdominal pain

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### SUMMARY

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**To cite:** Forte A, Santarpia L, Venetucci P, *et al. BMJ Case Rep* 2023;**16**:e254157. doi:10.1136/bcr-2022-254157 A woman in her 40s was admitted to hospital with weight loss, asthenia, persistent abdominal pain and post-prandial nausea and vomiting. Other comorbidities were anxiety-depressive disorder, gastro-oesophageal reflux disease and fibrocystic mastopathy. On admission her body mass index (BMI) was 15.57 kg/m<sup>2</sup> with a reported weight loss of 6 kg during the last 3 months. The patient underwent a double contrast abdominal CT scan, which showed that the third portion of the duodenum appeared to be compressed between the superior mesenteric artery and the abdominal aorta. After a multidisciplinary evaluation, a conservative approach and nutritional supplementation was decided upon and administered. At the 1-year follow-up the symptoms had greatly improved; the epigastric pain, although persistent, was reduced, also due to the weight gain to 50 kg (BMI 19.5 kg/m<sup>2</sup>). Wilkie's syndrome, in its acquired form, predominantly affects young women after rapid weight loss. In the diagnostic work-up, case history, physical examination and radiological findings play a key role.

# BACKGROUND

Superior mesenteric artery syndrome (SMAS), or Wilkie's syndrome, is an infrequent cause of chronic obstruction of the third portion of the duodenum, resulting in its compression between the superior mesenteric artery and the aorta.<sup>1</sup> According to the literature, its prevalence in the general population ranges between 0.0024% and 0.34%, with women being more often affected than men.<sup>1</sup> The main clinical characteristics are epigastric abdominal pain, post-prandial nausea and vomiting and a recent history of rapid weight loss.<sup>2</sup> The onset of the symptoms is due to weight loss, which causes intermediate mesenteric adipose pad reduction.

An accurate account of the medical history and physical examination are essential for diagnosis. An abdominal CT scan with contrast is given to confirm the diagnosis by determining a reduction in the angle between the aorta and the superior mesenteric artery, with compression of the third duodenal portion. Physiologically, the width of this angle ranges between  $25^{\circ}$  and  $60^{\circ}$  and the distance between the two blood vessels is  $10-20 \text{ mm.}^3$ 

The therapeutic approach is often conservative and is based on nutritional support, which aims to increase the mesenteric fat pad and the aortomesenteric angle. If this treatment approach fails, surgery is indicated.<sup>45</sup>

# CASE PRESENTATION

A woman in her 40s was admitted to our department with weight loss, asthenia, post-prandial nausea and vomiting and persistent abdominal pain, mainly located in the epigastrium. The patient reported that the pain increased at the end of meals and she described it as colic. She also had anxietydepressive disorder, gastro-oesophageal reflux disease and fibrocystic mastopathy. She reported that in adolescence she had suffered from eating disorders with anorexia and bulimia.

Deep palpation showed that the patient's abdomen was tense with an upper part guarding and marked epigastric regional pain; no abdominal organ enlargement was found. A tympanitic sound to percussion was found in all quadrants together with normal bowel peristalsis.

Blood chemistry did not show any changes other than a mild increase in indirect bilirubin values related to Gilbert's syndrome. A urine test showed a urinary tract infection which was treated with specific antibiotic therapy.

At admission, the patient's weight was 45 kg with a body mass index (BMI) of  $14.69 \text{ kg/m}^2$ . The patient reported a weight loss of 6 kg in the previous 3 months linked to the severe post-prandial epigastric pain.

Abdominal ultrasonography, influenced by considerable gastrointestinal meteorism, showed two cystic formations in the right ovary, which was previously known. Transabdominal and transvaginal ultrasonography showed the presence of a small infiltrating endometriotic outbreak isolated at the right sacral uterine ligament level. An x-ray of the digestive tract with Gastrografin showed significant gastric dilation compatible with Wilkie's syndrome (figure 1).

A contrast-enhanced abdominal CT scan (oral and intravenous) was performed for additional diagnostic investigation, which showed significant dilation of the stomach with progression of the contrast slowing down in the first and second portion of the duodenum and an accentuated reduction in the third portion. Extrinsic compression of the duodenum between the superior mesenteric artery and the aorta was also seen (figures 2–4). The radiological images also showed a reduction in the retroperitoneal adipose pad which normally interposes between the upper margin of the duodenum and the lower surface of the superior mesenteric artery. The angle between the aorta and the upper mesenteric artery was less than 20° (normal value 25-60°), confirming compression of the third



**Figure 1** X-ray examination with Gastrografin showing slowdown of venous contrast and significant dilation of the stomach.

duodenal portion between the aorta and the superior mesenteric artery.

Management of the patient was discussed in a multidisciplinary meeting involving an internist, a radiologist, a gastroenterologist and a surgeon and, after close evaluation, a conservative option was chosen with nutritional rehabilitation support.

A high-calorie semi-liquid oral diet consisting of frequent meals of liquid or blenderised food was prescribed. In addition, liquid oral nutritional supplements (total volume 600 mL, energy density 1.5 kcal/mL, protein/carbohydrate/lipid ratio 25%/55%/30%) were prescribed.

At the 1-year follow-up the patient's weight had increased to 50 kg (BMI 19.5 kg/m<sup>2</sup>) with a consequent relief of abdominal symptoms. The epigastric pain, although persistent, had reduced and no post-prandial nausea and vomiting episodes were reported. In addition, a CT scan showed that the dilation of the stomach was reduced (figure 5).

### **OUTCOME AND FOLLOW-UP**

The enteral nutrition treatment chosen for the patient resulted in an early reduction in symptoms. At the 1-year follow-up, as a result of the weight gain and psychological support, the patient showed a significant reduction in her symptoms.



Figure 2 CT sagittal scan showing a reduction in the aortomesenteric angle.



**Figure 3** CT axial scan showing compression of the third portion of the duodenum.

### DISCUSSION

Chronic recurrent abdominal pain without signs of inflammation or organ impairment can be difficult to treat. This case shows a rare cause of abdominal pain, particularly affecting women who undergo rapid weight loss, triggering a vicious circle leading to the onset of SMAS or Wilkie's syndrome.<sup>1</sup>

An accurate medical history and subsequent radiological investigation are important to obtain an early diagnosis and to reduce inappropriate treatments or invasive tests. A medical approach is usually the first therapeutic option.

In this case, the patient's anxiety-depressive syndrome and her past history of anorexia nervosa<sup>6</sup> misled the diagnostic process, with the weight loss initially being attributed to these disorders.

The diagnosis of SMAS could be suggested by abdominal ultrasonography, taking into consideration the possible negative influence that bloating and abdominal pain may have on the measurement of the aorto-mesenteric angle. Therefore, the gold standard for the measurement of this angle is a contrast-enhanced CT scan.

At the 1-year follow-up, our approach with only oral nutritional supplements and psychological support resulted in pain reduction and increased weight.

Wan *et al*<sup>7</sup> have recently reported the use of enteral nutrition in 26 patients with SMAS as an effective option in the relief of symptoms, leading to an increase in weight.

It has been reported that conservative treatment is successful only in patients with a short history of SMAS and a minor degree of duodenal stenosis. In our case, despite the significant stenosis, the



**Figure 4** CT sagittal scan before treatment showing marked gastric overdistension, with the lower edge of the stomach caudally exceeding the umbilical scar by a few centimetres.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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gastric overdistension, with the lower edge of the stomach reaching the umbilical scar.

Figure 5 CT sagittal scan following treatment showed a reduction in

nutritional support restored part of the retroperitoneal fat, widening the angle between the two vessels, which resolved the compression, reduced the stomach dilation and relieved the symptoms.<sup>8</sup>

# Learning points

- ► The signs and symptoms of SMAS are usually non-specific and can easily lead to misdiagnosis.
- SMAS should be suspected in patients with rapid and significant weight loss and chronic abdominal pain not explained by other causes.
- This clinical case is intended to be an example of a condition to consider in the differential diagnosis of abdominal pain.

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