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Case Report

Wilkie's syndrome in a patient with Duchenne's muscular dystrophy: A Case Report ☆

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ABSTRACT

Superior mesenteric artery syndrome, or Wilkie's syndrome, is one of the rarest gastrointestinal disorders known to medical science. It is characterized by the vascular clamp of the third portion of the duodenum, between the superior mesenteric artery and the aorta. It presents as an uncommon cause of upper intestinal obstruction. Imaging is required, preferably with a contrast-enhanced CT or an MRI; conservative management is preferred, leaving surgery for the most complex cases.

We present the case of a 34-year-old man with Duchenne's muscular dystrophy and a history of substantial weight loss after hospital admission for aspiration pneumonia. He underwent an abdominal CT scan that showed enlargement of the stomach, the second and third parts of the duodenum; without observing masses, the patient received conservative management with a nasojejunal feeding tube. At the outpatient reevaluation, 1 month postdischarge, he became asymptomatic and had progressive weight gain.

Introduction

Wilkie's syndrome, or superior mesenteric artery syndrome (SMA), is a strange cause of duodenal obstruction due to extrinsic compression between the SMA and the aorta (AMS—Ao) and a morbid entity when diagnosis is delayed. Von Rokitansky described this entity for the first time in his book with a case report in 1842; later, Wilkie described the pathological findings and diagnosis with 75 of his cases in 1927. Thus, this syndrome is also known as Wilkie's syndrome [1].

The most common cause occurs after sudden and important weight loss, but other medical causes can also cause this duodenal compression. This syndrome can become a vicious cycle created by abdominal pain that leads to food aversion, poor intake, and weight loss. It is considered a diagnosis of exclusion based on clinical findings [2].

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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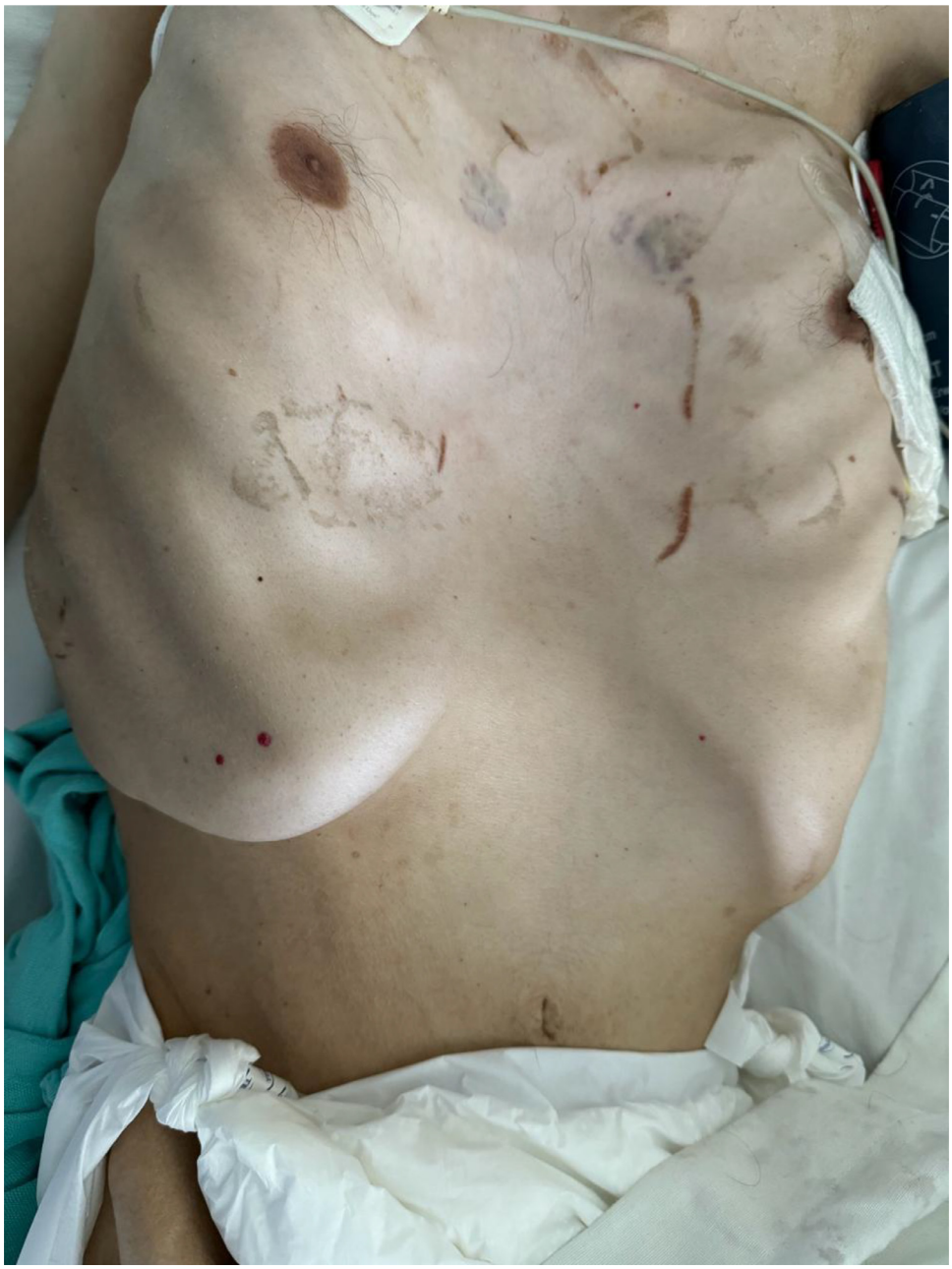

Fig. 1 – Abdominal examination showing severe malnutrition.

Duchenne muscular dystrophy is an important, escalating, muscle-wasting disease leading to complications in mobility and the need for assisted ventilation and premature death [3]. Here, we describe a rare case of a patient who suffered this disease after a common complication of the underlying condition.

Case presentation

A 34-year-old male engineer who was already diagnosed with Duchenne muscular dystrophy suffered from difficulty breathing, coughing, and fever. When he was lethargic, the patient went to our emergency unit for treatment and was diagnosed with aspiration pneumonia that required intubation; he was treated with broad-spectrum antibiotics. In the following month, his weight dropped from 62 kg to 42.5 kg; after solving the infection, he was extubated, and in the first 3 days and with a diet consisting of polymeric-based diet, he presented with severe vomiting and nausea, his vomit was gastro-biliary, and the symptoms worsened after eating, at this point, we opted for an abdominal CT, that showed signs of upper intestinal obstruction and gastromegaly. We started conservative treatment and solved it within the first 48 hours. A thorough physical examination revealed that the patient was

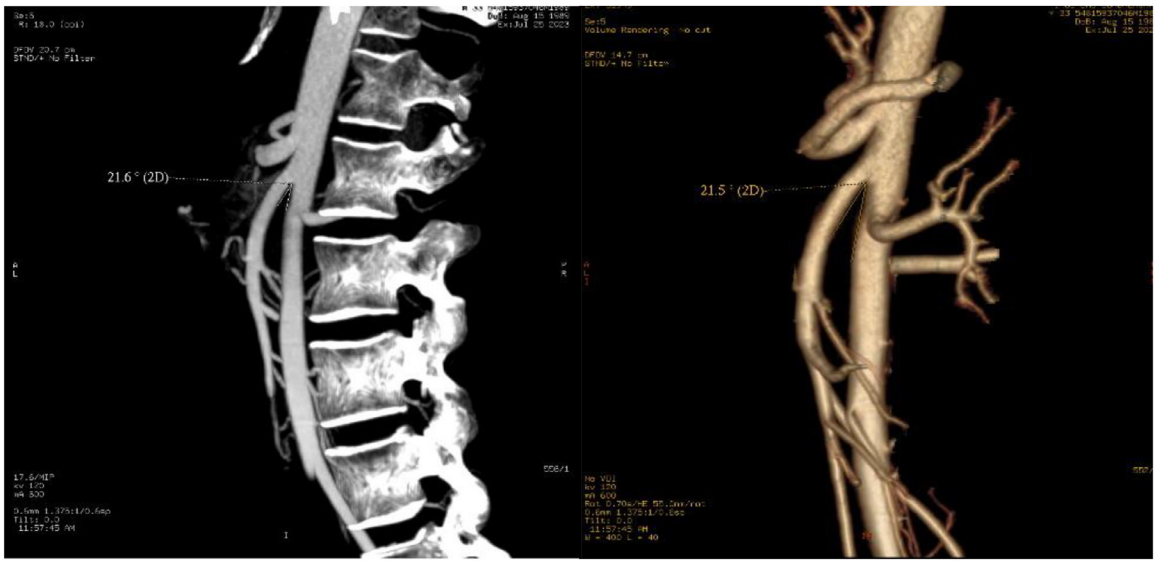

Fig. 2 – Abdominal CT reconstruction scan suggests the reduced angle between the SMA and abdominal aorta.

slender (160 cm in height, 42.5 kg in weight, BMI = 16.6 kg/m^2) with a pectus carinatum, severe depression in the abdomen (Fig. 1); no abdominal tenderness or rebound tenderness was observed.

Laboratory examination shows the following: white blood cells: 15.6 × 109 /L (3.5–9.5), lymphocytes 11% (17-45); hemoglobin: 8 g/dL (115–150); K+: 2.8 mmol/L (3.5–5.3); Na +: 136 mmol/L (137–147); Cl−: 96.8 mmol/L (99–110), amylase: 113 U/L (35–135); lipase: 128.6 U/L, (0–60); C-reactive protein (CRP)<3 mg/L (0–3), albumin 1.8 g/dL (3.4-5.4).

Based on the clinical presentation and the acute weight loss, CT reconstruction was performed, and we performed measurements, obtaining an aorto-mesenteric angle of 21.6 (Fig. 2), and the distance between the Aorta and the SMA was 2.6 mm (Fig. 3).

We made the diagnosis of SMA syndrome. Considering the patient's age and condition, we decided to intervene with nasojejunal tube placement and enteral nutrition. After the nasojejunal tube was placed, the patient no longer vomited and was discharged from the hospital 5 days later. Follow-up revealed that he did not experience vomiting after his discharge. After 4 weeks, the patient's weight increased to 49 kg and BMI to 19 kg/m^2.

After 2 months postdischarge and adding some liquids via mouth, we decided to take out the nasojejunal tube and continue enteral nutrition, without any discomfort or issues.

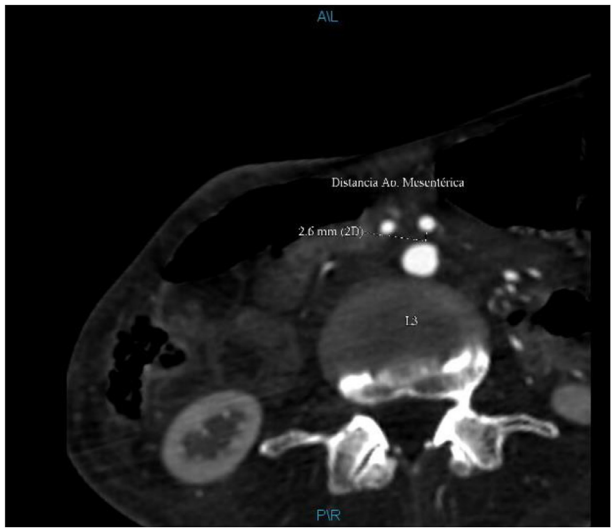

Fig. 3 – Contrast-enhanced abdominal CT shows diminished distance between the abdominal aorta and SMA.

Discussion

This syndrome affects young female adults frequently. A revision found that the mean age of diagnosis is 23 years. The affected age appears to be related to the underlying conditions of the patients (congenital scoliosis or chemotherapy-induced weight loss). The female: male ratio is 3:2 [4].

Wilkie's syndrome is characterized by a narrowing of the aortomesenteric angle. In a physiologic state and due to the erect position, the aorta-SMA angle ranges from 45 to 60°. The presence of perivascular fat tissue maintains the average AOM distance between 10 and 20 mm [5]. In Wilkie's syndrome, the angle acutely decreases, ranging from 22 to 28 degrees with an AOM distance between 2 and 8 mm strongly suggestive of SMA syndrome in the correct patient setting, leading to extraluminal compression of the duodenum [5].

The causes of SMA syndrome are diverse. The known causes are anatomical variation, mesenteric fat reduction, anatomical deformation caused by surgery or trauma, and

consumptive disease or trauma. The key point in its pathogenesis is the small angle between the SMA and the aorta [6].

SMA syndrome is caused by compression of a duodenal horizontal segment. The increased intestinal pressure after obstruction may affect duodenal papillary dysfunction, leading to bile reflux into the common bile duct and pancreatic duct. Whether or not bile reflux can cause pancreatitis and gallbladder distention (or cholecystitis) remains a controversial issue [6].

Regardless of the associated factors, a depletion of the fatty cushion around SMA occurs, leading to the anatomical alterations mentioned above. In our case, the patient presented with prolonged intubation that required prolonged starvation and severe weight loss.

Patients with SMA syndrome may have different clinical presentations: chronic subtle symptoms or an acute exacerbation of symptoms. Signs and symptoms of duodenal obstruction characterize the latter; on the other hand, chronic cases present with perpetuated vague abdominal symptoms or repetitive episodes of abdominal pain associated with vomiting. Other less common symptoms are esophageal reflux, early satiety secondary to increased transit time, and gastric distension [7].

The diagnosis of SMA syndrome is challenging and often delayed due to its variable symptomatology. Clinical conjecture is needed, and the diagnosis is based on clinical evidence supported by radiological findings. Starting with a plain abdominal x-ray, followed by barium radiography, demonstrates dilatation of the first and second part of the duodenum with or without gastric dilatation, the anti-peristaltic flow of the contrast proximal to the obstruction and a delay of 4 to 6 hours in gastro-duodenojejunal transit time, with symptom improval when the patient is placed in different positions: prone, knee-chest or left lateral position. Contrast-enhanced CT or magnetic resonance angiography allows visualization of the vascular clamp of the duodenum and require measurement of the aorto-mesenteric angle and distance. Endoscopic examination may visualize an extrinsic compression, leading to suspicion of this condition [7]. In this case, we observed a dilated stomach suggestive of upper GI obstruction, and we decided to take CT scans and 3D reconstruction.

Treatment consists of conservative measures, such as gastric decompression, parenteral nutrition, and postpyloric feeding when possible, followed by an oral diet as tolerated. No time limit has been specified for the medical treatment [1].

Surgery is considered therapy if conservative treatment fails. Duodenojejunostomy is the surgical choice to solve the obstruction, with a success rate of up to 90%. Some other surgical procedures, less invasive, are plausible, like Strong's procedure, which involves remotion of the ligament of Treitz with mobilization of the duodenum; the perk of this operation is its failure rate of 25% [7]. In this case, we offered our patient conservative management, with symptom improvement when weight gain was achieved.

Conclusion

Although Wilkie's syndrome is a rare disease, it's a life-threatening disease that we believe is underdiagnosed; it can be associated with other variable symptoms in addition to vomiting and can mimic a variable amount of gastrointestinal problems. Clinical awareness is of utmost importance, and having a range of therapeutic strategies is beneficial for the patient, leaving surgery for the most complex cases due to its high complication rate. With our patient, due to his comorbidities, he is prone to losing weight quickly as hospital stay is suspected to increase. Surgical treatment would be the next step in management.

Patient consent

We obtained approval from the patient for his case to be reported. He also agreed to the pictures shown in the case report.

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