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Case report

Acquired Superior Mesenteric Artery Syndrome: A Case Report with Associated Mortality

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Abstract

Introduction: Superior Mesenteric Artery Syndrome (SMAS) is a rare vascular-origin intestinal obstruction affecting the third portion of the duodenum. It is often underdiagnosed due to its multiple confounding factors, leading to delays in diagnosis and a high morbidity and mortality rate. The primary underlying mechanism is significant weight loss, and it is more commonly seen in young women. Current treatment approaches include either medical management or minimally invasive surgery. **Case Presentation:** We present the case of a 61-year-old woman with a six-month history of insidious abdominal pain and recurrent episodes of vomiting, accompanied by severe weight loss. Imaging studies confirmed a diagnosis of SMAS, which was secondary to tumor recurrence in metastatic breast cancer. Conservative management was initiated; however, the patient ultimately succumbed to infectious complications. **Conclusions:** SMAS is a rare and challenging condition to diagnose. Various underlying factors contribute to significant weight loss, which is central to its pathophysiology. Computed tomography (CT) and magnetic resonance imaging (MRI) are the preferred diagnostic modalities. While conservative management remains the first-line treatment, it is associated with a high recurrence rate. Surgical intervention, though an option, carries substantial morbidity risks due to the complexity of the procedure.

Keywords

Superior Mesenteric Artery Syndrome, intestinal obstruction, weight loss, breast neoplasm, duodenal obstruction.

INTRODUCTION

Superior mesenteric artery syndrome (SMAS) is a rare condition, first described by Rokitansky in 1842. It results from extrinsic intestinal obstruction due to vascular origin, occurring at the third portion of the duodenum. This condition is caused by a reduction in the angle between the superior mesenteric artery and the aorta[1]. The prevalence is unknown, with varying incidences reported in imaging studies, ranging from 0.013%-0.78% up to 2.67% in patients with constipation[1,2]. Currently, it is an underdiagnosed condition due to its nonspecific symptoms. In fact, 10.8% of patients with SMAS were previously treated for dyspepsia[3]. All of this leads to a delay in diagnosing a condition with a reported mortality rate of approximately 33%[4,5].

This report presents the case of a 61-year-old woman with fatal outcomes due to tumor relapse from a breast neoplasm in the context of SMAS.

PRESENTATION OF THE CASE

This is a 61-year-old woman with a history of primary clinical hypothyroidism and right breast cancer diagnosed two years prior, currently in remission (following neoadjuvant therapy and mastectomy). She was admitted due to a six-month clinical course characterized by progressive colicky pain in the epigastrium and mesogastrium, occurring postprandially. In the last 20 days, she developed episodes of vomiting with food content and occasionally bile, leading to oral intolerance and a weight loss of approximately 10 kg. Initial tests revealed hypokalemia (2.8 mEq/L), hypochloremia (94 mEq/L), and metabolic alkalosis, accompanied by signs of dehydration. These abnormalities were adequately corrected. Other tests, including thyroid function tests, were within normal limits.

An abdominal X-ray was performed, showing dilated intestinal loops with abundant fecal material and the presence of gas in the rectal ampulla. After performing an intestinal lavage with a nasogastric tube, an upper gastrointestinal endoscopy was conducted, revealing erosive grade B esophagitis according to the Los Angeles Classification. No pyloric syndrome or obstruction was observed in the examined areas.

A contrast-enhanced abdominal CT scan was performed, showing evidence of a distended gastric chamber and first and second portions, with collapse of the distal third portion at the level of the superior mesenteric artery (SMA). The aortomesenteric angle was 22° and the aortomesenteric distance was 7 mm (**Figures 1** and **2**). Additionally, blastic lesions were noted in the iliac bones and vertebral bodies.

In the context of tumor relapses in the patient with sarcopenia, along with clinical and imaging findings of SMAS, initial management focused on parenteral nutritional recovery and concurrent oncological treatment. The patient developed complications during her hospital stay, including pulmonary-origin sepsis, which led to a fatal outcome.

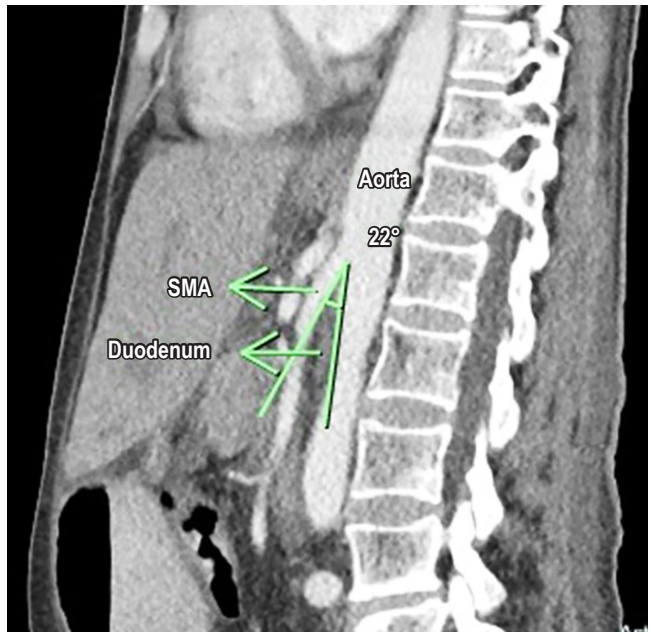

Figure 1. Contrast-enhanced abdominal CT scan in the sagittal section showing an angle of 22° between the aorta and the superior mesenteric artery. The image is the property of the authors.

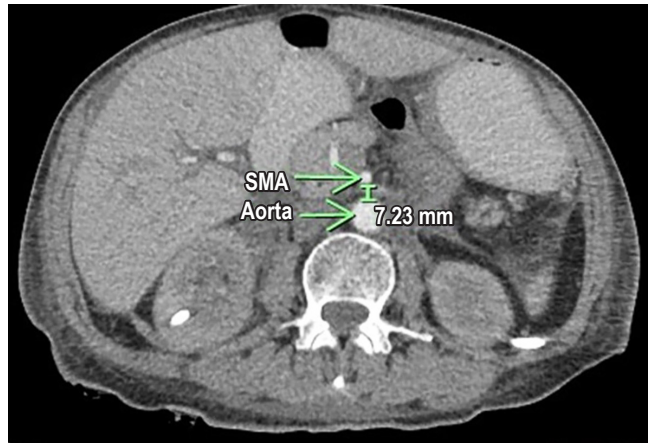

Figure 2. Contrast-enhanced abdominal CT scan in axial section showing an aortomesenteric distance of 7.3 mm. The image is the property of the authors.

DISCUSSION

SMAS, also known as Wilkie's syndrome, cast syndrome, mesenteric duodenal obstruction, or aorto-mesenteric compass syndrome, was first described by Carl Freiherr Von Rokitansky in 1861. However, it was Wilkie in 1927 who published a case series of 75 patients, which is why the syndrome is named after him[6,7]. As of 2022, there are 730 articles with 2400 reported cases[1]. Its incidence is low, estimated to be between 0.1% and 0.3% of the general population. However, in specific conditions such as anorexia nervosa (2.73%)[8] and functional dyspepsia (10.8%)[3], the prevalence remains unknown. SMAS is more commonly seen in women than in men, with a female-to-male ratio of 3:1, and an average age of onset of 23 years (range: 10-40 years). However, there has been a growing number of reports in individuals over the age of 40 (range: 0-91 years)[1,9].

The third part of the duodenum, which is the longest and narrowest, passes between the aorta and the superior mesenteric artery (SMA) from right to left. Both vessels are covered by retroperitoneal fat and lymphatic tissue, which act as cushioning and reinforcement, allowing the duodenum to pass unobstructed between them[10]. The SMA arises from the front section of the abdominal aorta

behind the pancreas at the level of the L1 vertebra, creating an acute angle with the aorta. Typically, the angle between the aorta and the superior mesenteric artery is around 45° (range: 38°-65°), and a normal aortomesenteric distance is described as between 10 and 28 mm[1,7]. When there is a decrease in the available space for the duodenum, intestinal compression occurs due to the vascular structures. An angulation between the abdominal aorta and the superior mesenteric artery is described as ranging from 6° to 25°, with an aortomesenteric distance between 2 and 8 mm[11].

This syndrome can be either congenital or acquired. The congenital type is associated with malrotation, low take-off of the superior mesenteric artery, Ladd's bands, idiopathic peritoneal adhesions, a shortened and thickened mesenteric root, and a more cephalic or hypertrophied position of the Treitz ligament[1,6]. The acquired type results from weight loss (due to cancer, bariatric surgery, chronic infections, burns, eating disorders, intestinal surgeries, malabsorption, rheumatoid arthritis, among others), which leads to a reduction in the perivascular fat surrounding the abdominal aorta and the SMA[12].

The clinical symptoms are nonspecific and can be either acute or chronic, sometimes starting insidiously. Symptoms include postprandial epigastric pain (59%), nausea (40%), vomiting (50%), early satiety (32%), weight loss, and anorexia (32%). Obstruction can progress rapidly, leading to distal ischemia, with consequences such as lactic acidosis and even death[12]. A key diagnostic point is that abdominal pain worsens in the supine position and improves in the lateral decubitus position (knees-to-chest position) or with the Hayes maneuver (applying pressure below the umbilicus in a cephalad direction, which relieves pain by reducing mesenteric tension)[1,12].

Abdominal X-ray is typically the first radiological study, showing findings consistent with high intestinal obstruction, such as a dilated gastric and gastroduodenal chamber. Other studies that may help include, on the one hand, an upper barium contrast study showing gastroduodenal obstruction with improvement upon position changes, and on the other hand, abdominal ultrasound, which could be helpful in measuring the aortomesenteric angle. Currently, contrast-enhanced tomography and magnetic resonance imaging are the preferred studies for measuring the aortomesenteric angle and distance, offering detailed views of adjacent structures[11]. An endoscopic study is necessary to rule out any mechanical cause of obstruction[7].

Among the described complications are hypovolemic shock, aspiration pneumonia, severe electrolyte imbalance, gastrointestinal mucosal damage (peptic and biliary acid reflux), emphysema, necrosis, gas in the portal vein, pneumoperitoneum, pancreatitis, and even sudden death (secondary to electrolyte imbalance and acidemia)[1].

The treatment is based on two main pillars: symptom relief and nutritional replenishment. Conservative medical management includes gastrointestinal decompression with the passage of a nasoenteric tube, the use of prokinetics (e.g., metoclopramide), feeding in small amounts, positional repositioning or the Hayes maneuver, and total parenteral nutrition to improve nutritional status and increase the aortomesenteric angle by increasing perivascular adipose tissue. The duration of conservative management is variable; however, after 4-6 weeks without improvement, surgical treatment should be considered[1,10,13,14]. Surgical management is the treatment of choice if there is no response to conservative management and in patients with multiple abdominal interventions, immobility, and atherosclerosis of the SMA[1]. There are no clinical trials comparing medical versus surgical management, but a cohort study has shown recurrence rates of 71.3% versus 15.8%, respectively[15]. The three available surgical options are gastrojejunostomy, duodenojejunostomy, and the Strong procedure. Duodenojejunostomy is the most common surgical procedure and appears to have the best outcomes, with a success rate of 80%[13,16]. Open duodenojejunostomy was traditionally the preferred approach; however, the current standard of care favors minimally invasive techniques, including ultrasound-guided endoscopic gastroenterostomy[17].

CONCLUSION

SMA syndrome is an underdiagnosed condition associated with mortality, and in adults, it is linked to any situation that leads to overall weight loss. Its diagnostic approach relies on a combination of clinical evaluation and imaging studies (CT or MRI). Management should be individualized, as conservative treatment has a high recurrence rate, while surgical management carries significant morbidity.

Conflicts of interest

The authors declare no conflict of interest.

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Síndrome de la arteria mesentérica superior adquirido: presentación de un caso con mortalidad asociada

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