CASE REPORT
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A rare cause of upper gastrointestinal system obstruction: Superior mesenteric artery syndrome (Wilkie's syndrome); two different case reports

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ABSTRACT

Superior mesenteric artery syndrome is defined as a collection of clinical symptoms and findings that result from compression of the third part of the duodenum between the aorta and the superior mesenteric artery. Here, we describe two patients who were diagnosed with superior mesenteric artery syndrome. Two patients, 18 and 38 years old, respectively, presented to our clinic with complaints of nausea, vomiting, and weight loss. Computed tomography scans of both patients supported diagnoses of superior mesenteric artery syndrome. The 18-year-old patient recovered with conservative treatment. However, our 38-year-old patient did not recover with conservative treatment and required two surgeries. In the first operation, duodenal release with Treitz's ligament dissection and pyloroplasty were performed because of concomitant hypertrophic pyloric stenosis. Because the patient exhibited gastroparesis and gastric ptosis after the first operation, subtotal gastrectomy and Roux-n-Y gastrojejunostomy were performed in the second operation. No complications were observed during follow-up after the second operation. Superior mesenteric artery syndrome should be considered in the differential diagnosis of patients with nausea, vomiting, and weight loss of unknown cause. During treatment, weight-gaining conservative approaches should be attempted initially, but surgical treatment should not be excessively delayed in patients who do not respond to medical treatment.

Keywords: Superior mesenteric artery syndrome, Wilkie's syndrome, upper gastrointestinal system obstruction

INTRODUCTION

Superior mesenteric artery (SMA) syndrome is defined as a collection of clinical symptoms and findings that result from compression of the third part of the duodenum between the aorta and SMA. This syndrome is also known as chronic duodenal ileus, cast syndrome, mesenteric root syndrome, intermittent arterio-mesenteric obstruction, and Wilkie syndrome (1). In patients with nonspecific symptoms of proximal intestinal obstruction (e.g., nausea, vomiting, and abdominal pain), SMA syndrome is often not considered during differential diagnosis. In these instances, diagnosis is often delayed; late diagnosis may lead to complications such as electrolyte anomalies, gastric perforation, gastric pneumatosis and portal venous gas, and obstructive gastric bezoar (2). With the widespread use of cross-sectional imaging methods, SMA syndrome has been diagnosed with increasing frequency (3). Herein, we describe two patients who presented to our hospital with signs of gastrointestinal obstruction and were diagnosed with SMA syndrome based on computed tomography (CT) findings.

CASE REPORT

Case 1

An 18-year-old man was admitted to our hospital with long-standing intermittent abdominal pain, nausea, and vomiting. Physical examination revealed that the patient had a cachectic appearance, with sensitivity in the epigastric region. The patient reported no significant disease in his medical history or family history. Haemogram and biochemical parameters were normal. Abdominal CT revealed that the third segment of the duodenum was compressed between the aorta and SMA. The angle between the abdominal aorta and SMA was 12 degrees; the aortomesenteric distance was 5 mm (Figure 1). Dilatation was present in proximal segments

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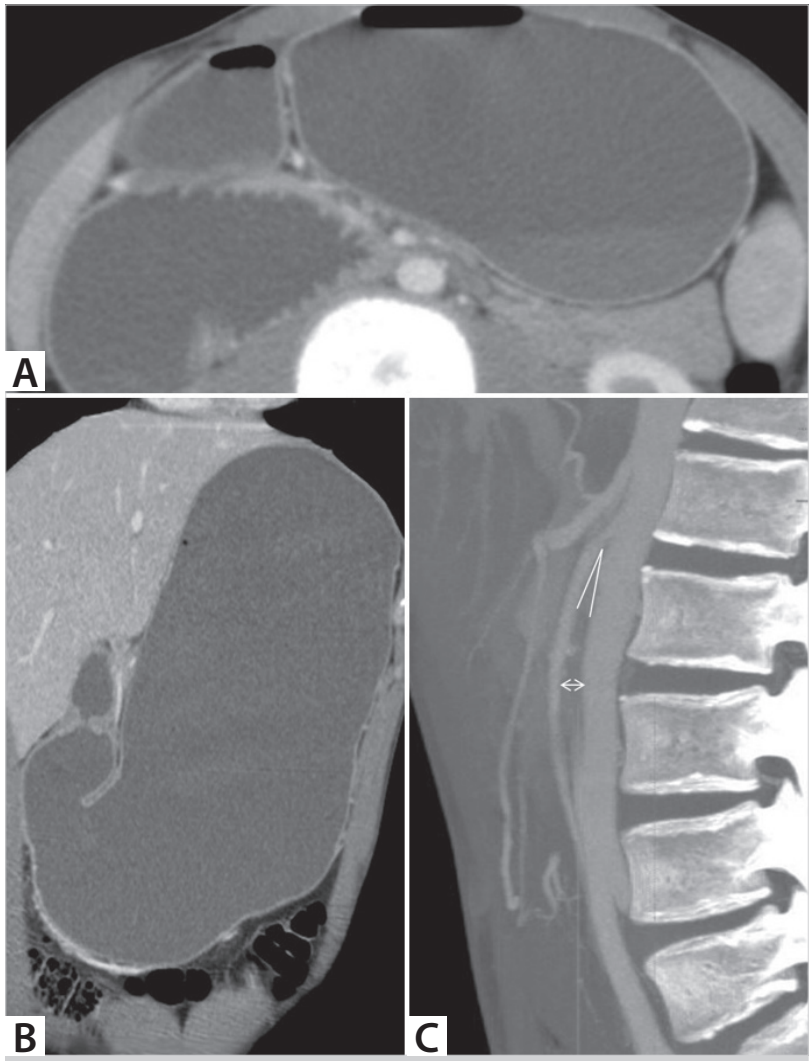

Figure 1. A. Axial and **B.** coronal CT scans show excessive stomach dilation. **C.** Sagittal CT scan shows narrowed SMA-Aort angle (Patient 1).

of the duodenum and in the stomach (Figure 1). The patient was diagnosed with SMA syndrome based on radiological and clinical findings. The patient's height was 172 cm, weight was 44 kg, and body mass index was 14.9 kg/m^2. Total parenteral nutrition and enteral nutrition solutions and a fluid regimen were initiated. The patient gained two kg in two weeks. After medical treatment, the patient's complaints and radiological findings improved. He was discharged with proper feeding and intermittent clinical follow-up recommendations. Four months after discharge, the patient was contacted by telephone; he reported no complications and a weight of 48 kg.

Case 2

A 38-year-old man was admitted to our outpatient clinic with complaints of nausea, vomiting, and abdominal pain. The patient exhibited cachexia and pale colour. No clinically significant findings were present in the patient's laboratory values; moreover, there was no significant disease in his medical history or family history. The patient was subsequently hospitalised. Abdominal CT revealed that the aortomesenteric angle was reduced, the third part of the duodenum was compressed, and the stomach and proximal duodenum were dilated (Figure 2a, b). Thus, the patient was diagnosed with SMA syndrome. Nasogastric decompression was applied; the patient then began to receive intravenous hydration and total parenteral nutrition. The nasogastric tube was withdrawn when it was no longer needed, and a fluid regimen was initiated. Enteral nutrition solutions were added to the diet. The patient's height was 190 cm, weight was 52 kg, and body mass index was 14.4 kg/m^2; the patient was weighed daily during hospitalisation. After two weeks, his weight had increased by three kg and he was able to tolerate a semi-solid regimen; he was discharged with a prescription for enteral nutrition solutions. One week after discharge, he was

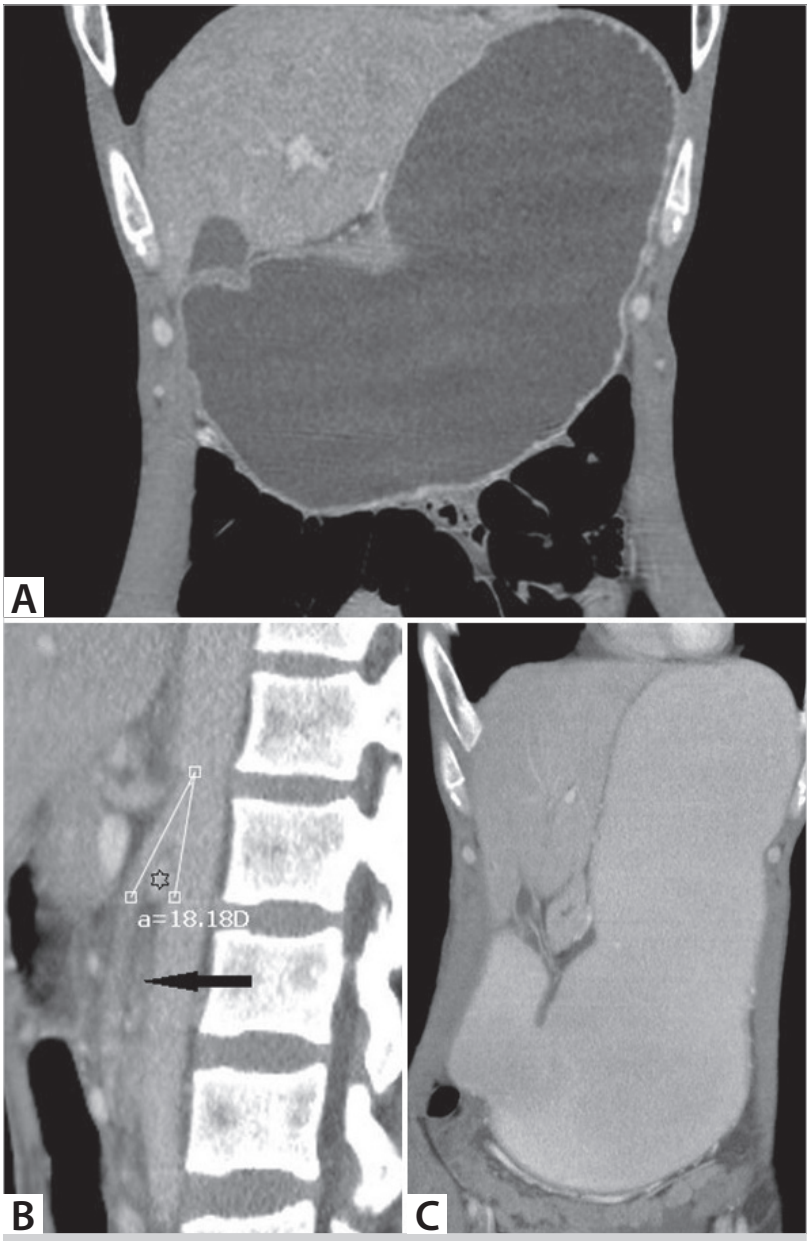

Figure 2. A. Preoperative coronal CT image shows gastric dilatation. **B.** Sagittal CT scan shows narrowing of the aortomesenteric angle and reduction of the aorta-SMA distance. **C.** CT image shows gastric ptosis and gastroparesis after initial surgery (Patient 2).

hospitalised again with the complaint of nausea and vomiting. A nasogastric tube was inserted and total parenteral nutrition was started. Because the patient could not tolerate oral intake, surgery was planned. During laparotomy, the third part of the duodenum was subjected to compression; however, hypertrophic pyloric stenosis was observed. The Treitz ligament was dissected to partially free the duodenum, and pyloroplasty was performed. The postoperative day 4 regimen was started; three days later, a nasogastric tube was inserted again due to excessive vomiting. During subsequent endoscopy, the pylorus was easily passed, the duodenum was easily accessed until the third part, and reflux gastritis and gastric dilatation were observed. Administration of a proton pump inhibitor was initiated, along with domperidone, metoclopramide, and ursodeoxycholic acid. The patient was discharged after treatment and his symptoms returned. Three days after discharge, the patient was again admitted to the ward with excessive abdominal distention and vomiting. CT showed gastric ptosis, gastroparesis, and excessive stomach dilatation (Figure 2c). After routine procedures, reoperation was planned. In the second operation, subtotal gastrec-

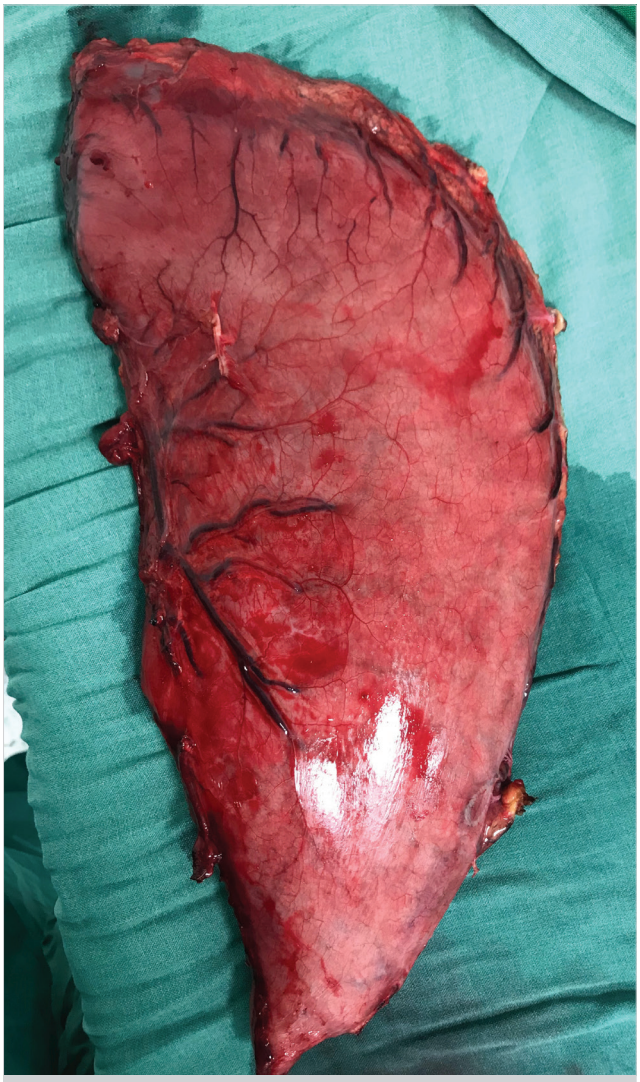

Figure 3. Subtotal gastrectomy and Roux-n-Y gastrojejunostomy specimen obtained during the second operation (Patient 2).

tomy and Roux-n-Y gastrojejunostomy were performed (Figure 3). Intraoperative arterial pressure was monitored by placement of an intraarterial cannula from the radial artery before each induction. Because the patient's body mass index was low, he was given low doses of drugs. After surgery, the patient was safely awakened without any anaesthesia complications. Oral intake was started on the fourth postoperative day, and the patient was discharged on the seventh postoperative day. There were no complications in the second postoperative month, and the patient's weight was 57 kg.

DISCUSSION

SMA syndrome was first mentioned in an anatomy book published by Austrian professor Card von Rokitansky in 1842 (4). A number of patients have been described in the literature. In 1927, Wilkie published a series of 75 cases. Therefore, SMA syndrome is also known as Wilkie's syndrome (1). According to radiological studies, the incidence of SMA syndrome is between 0.2% and 0.78% (5). SMA syndrome is reportedly twofold more common in women than in men; two-thirds of affected patients are between 10 and 39 years of age (1). Our patients were 18 and 38 years old, and both were men.

According to the literature, the factors leading to SMA syndrome may be congenital or acquired. The congenital presence of a short mesentery, presence of an abnormally located SMA or its branches, excessive mobility of the right colon, intestinal malrotation, and abnormal fixation of the duodenum by the Treitz ligament are congenital causes that may lead to the development of SMA syndrome (6). Normally, retroperitoneal adipose and lymphoid tissues serve as a cushion under the SMA, separating it from the vertebral column; this prevents the duodenum from being pinched between the aorta and the SMA. Excess weight loss creates a risk of SMA syndrome by reducing retroperitoneal fat and lymph tissues. Conditions such as anorexia nervosa, burns, obesity surgery, pulmonary tuberculosis, and cardiac cachexia reduce the retroperitoneal fat tissue and cause SMA syndrome. Symptoms may be acute (often after surgical procedures) or progressive. The most common symptoms include anorexia, postprandial epigastric pain, weight loss, nausea, and biliary vomiting (1). Similar symptoms were present in our patients.

The diagnosis should be confirmed by performance of specific radiological examinations in patients with clinical symptoms suggestive of SMA syndrome. Upper abdominal X-ray, upper gastrointestinal system barium radiography, CT, CT angiography, magnetic resonance angiography, ultrasonography, and endoscopy are useful in the diagnosis of SMA syndrome (1). Our patients were diagnosed by CT examination. On plain abdominal X-ray, the presence of gas in the duodenum and stomach is an important finding that supports a diagnosis of SMA syndrome (6). Barium radiography plays an important role in diagnosis. With the expansion of the proximal duodenum, abrupt interruption of barium in the third part is a classical, but non-specific, finding in patients with SMA syndrome. The following findings in barium radiography are important criteria for the diagnosis of SMA syndrome: enlargement in the first and second parts of the duodenum with or without expansion in the stomach; sudden external pressure obstruction in the third part of the duodenum in oblique or vertical style; antiperistaltic current proximal to the obstruction; 4-6 hours' delayed barium passage in the stomach and duodenum; and reduction of obstruction by lying on the left side, in the prone position, and in the knee-elbow position (7).

CT contrast angiography is considered the most important diagnostic method in the diagnosis of SMA syndrome because it shows the aortomesenteric angle, the distance between

the aorta and the SMA, the amount of adipose tissue, the location of the obstruction in the duodenum, and the enlargement proximal to the obstruction. CT can also show tumours and aneurysms, which may cause congestion, and provides a substantial advantage over other diagnostic methods. CT angiography and magnetic resonance angiography are considered equivalent in terms of measuring the aortomesenteric angle and the distance between the aorta and SMA (8). Important diagnostic criteria for SMA syndrome include reduction of the aortomesenteric angle to <20° (normal, 28-65°; in our patients, the aortomesenteric angles were 12 and 18 degrees, respectively), aortomesenteric distance reduction to <8 mm (normal, 10-28 mm; in our patients, the aortomesenteric distances were 5 and 7 mm, respectively) (Figures 1c and 2b), and gastric and proximal duodenal dilatation (5,8).

Upper gastrointestinal endoscopy should be performed in patients with SMA syndrome to exclude obstruction due to intraluminal pathological conditions in the duodenum. In endoscopy, dilation of the first and second parts of the duodenum and stomach, duodenal and gastric ulcers, and alkaline reflux gastritis may be findings that support the diagnosis of SMA syndrome (1).

The purpose of conservative treatment is to provide nutritional support to the patient, to help the patient gain weight, and to recover the loss of the fatty tissue paddle that is presumed to cause narrowing of the aortomesenteric angle. Surgical treatment should be performed in patients with long-lasting complaints who do not respond to conservative treatment (9). Here, we initially treated both patients conservatively and discharged them when they exhibited recovery. However, the development of concomitant hypertrophic pyloric stenosis in our second patient negatively influenced the clinical course and caused the patient to return for further assessment. Surgical treatment options include duodenojejunostomy, gastrojejunostomy, and Strong's procedure (mobilisation of the duodenum by division of Treitz's ligament); currently, duodenojejunostomy is most commonly performed. As a less invasive method, laparoscopic duodenojejunostomy is becoming increasingly common (10). In our second patient, hypertrophic pyloric stenosis had been more prominent in the first operation; thus, we presumed that we could provide palliation by allowing the release of the duodenum via Strong's procedure accompanied by pyloroplasty. However, during postoperative follow-up, gastroparesis and gastric dilatation (Figure 2c) led to persistent vomiting that did not respond to medical treatment; we were compelled to perform subtotal gastrectomy and Roux-n-Y gastrojejunostomy in a second operation (Figure 3).

SMA syndrome should be considered in the differential diagnosis of patients with nausea, vomiting, and weight loss of unknown cause, as well as those who are presumed to exhibit an obstruction at an upper level of the gastrointestinal tract. In the treatment of patients with SMA syndrome, weight-gaining conservative approaches should be tried initially, but surgical treatment should not be excessively delayed in patients who do not respond to medical treatment.

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OLGU SUNUMU-ÖZET
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Üst gastrointestinal sistem tıkanıklığının nadir bir nedeni: Superior mezenterik arter sendromu (Wilkie sendromu); iki farklı olgu sunumu

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ÖZET

Superior mezenterik arter sendromu, duodenumun üçüncü bölümünün aort ve superior mezenterik arter arasında sıkışmasından kaynaklanan klinik semptom ve bulguların bir derlemesi olarak tanımlanmaktadır. Burada superior mezenterik arter sendromu tanısı konan iki farklı olgu sunmak istedik. 18 ve 38 yaşlarında iki hasta kliniğimize bulantı, kusma ve kilo kaybı şikayetleri ile başvurdu. Her iki hastaya bilgisayarlı tomografi yapıldı ve superior mezenterik arter sendromu tanısı kondu. 18 yaşındaki hasta konservatif tedavi ile düzeldi. Ancak 38 yaşındaki hastamız konservatif tedavi ile düzelmedi ve iki kez ameliyat olmak zorunda kaldı. İlk ameliyatta, eşlik eden hipertrofik pilor stenozu nedeniyle Treitz ligamenti diseksiyonu ve piloroplasti ile duodenal geçişi sağladık. İlk ameliyattan sonra gelişen gastroparezi ve gastropitoz nedeniyle ikinci operasyonda subtotal gastrektomi ve Roux-en-Y gastrojejunostomi yapıldı. İkinci ameliyattan sonra hastanın takibinde herhangi bir problem gözlenmedi. Sebebi bilinmeyen bulantı, kusma ve kilo kaybı olan hastalarda ayırıcı tanıda superior mezenterik arter sendromu düşünülmelidir. Tedavide ilk önce kilo aldırıcı konservatif yaklaşımlar denenmeli, ancak tıbbi tedaviye yanıt vermeyen hastalarda cerrahi tedavi çok fazla gecikmemelidir.

Anahtar Kelimeler: Superior mezenterik arter sendromu, Wilkie sendromu, üst gastrointestinal sistem tıkanıklığı

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