Case Report

A Rare Cause of Abdominal Pain: Wilkie's Syndrome: Case Report

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ABSTRACT

Wilkie's syndrome is a rare acquired disease that occurs when acute dilatation occurs between the superior mesenteric artery and the aorta, compressing the duodenum between these two arteries. A loss of fat pads around the SMA due to reasons such as anorexia nervosa, weight loss, burns, trauma or prolonged bed rest is thought to be involved in the etiology of the disease. While the disease has an acute and chronic form, the chronic form is more common. Initially, conservative medical therapy is recommended for the treatment of Wilkie syndrome. In cases where conservative treatment fails, surgical treatment is indicated. In cases of extreme dilation of the stomach and duodenum being observed in the CT of patients presenting with complaints such as abdominal pain, nausea, and vomiting, a differential diagnosis of Wilkie's syndrome must be considered. A 22-year-old female patient was presented to our hospital with the complaints of epigastric pain, abdominal distention and nausea continuing for three days. On general examination, it was determined that her abdomen was distended and her epigastric region was sensitive. The diagnosis and treatment of this patient was examined with considering literature.

Key words: *Wilkie's syndrome, superior mesenteric artery, abdominal pain*

Wilkie's syndrome (superior mesenteric artery syndrome) is a rare, acquired disease caused by the acute angulation between the aorta and the superior mesenteric artery (SMA) and resulting compression of the duodenum between these two arteries [1]. This syndrome is also known as arterio-mesenteric artery syndrome, intermittantarterio-mesenteric obstruction, chronic duodenal ileus, mesenteric root syndrome, and cast syndrome. The incidence in the general population has been reported to be between 0.0024% with 0.34% [2]. It is seen in cases where the angle between the abdominal aorta and the SMA is 6° -25° (normal value: 38° -56°). Loss of fat tissue, which is a different consequence of weight loss, and acute angulation of the SMA are accepted as etiologic factors [3]. Wilkie syndrome is a rare clinical condition that causes symptoms such as epigastric pain, bloating and weight loss. It has an acute and chronic form [4]. With the help of this case report, we present the clinical progression and treatment process of a 22-year-old female patient with Wilkie's syndrome. The condition is rarely seen and difficult to diagnose, and in which surgical treatment is applied as a definitive treatment of the disease, together with literature data.

CASE REPORT

A 22-year-old female patient presenting with epigastric pain, abdominal distention and nausea continuing for three days was treated intravenously and medical treatment. She was discharged following improvement of the condition after decompression with a nasogastric tube. One day later, the patient presented to the general surgery outpatient clinic with renewed symptoms. Her esophagogastrodued-enoscopy revealed an external compression image of the posterior aspect of the gastric corpus, which did not impair

the mucosal integrity. In the patient's history, there were similar complaints that required intermittent hospitalization for a period of 1 year approximately 10 years prior.

In her physical examination, it was determined that her abdomen was distended and her epigastric region was sensitive. Her body mass index was 23.6 kg / m². There were no abnormalities in hemogram and biochemical values. Direct standing abdominal x-ray showed an increase in gastric fundus gas (Figure 1). An abdominal tomography showed marked distention of the stomach and duodenum up to the 3rd portion. Air-fluid leveling and stenosis in the 3rd portion of the duodenum were found (Figure 2). The angle between the SMA and the aorta was 10 degrees in the sagittal sections of tomography (Figure 3).

The patient underwent nasogastric decompression and drainage of 5000-6000 cc with bile occurrence. The patient's oral intake was discontinued; parenteral nutrition treatment, proton pump inhibitor, and antiemetic drugs were started. The patient was operated for SMA syndrome in the absence of a decrease in clinical complaints following medical treatment for about 1 week. It was found that the 3rd portion of the duodenum was stuck between the aorta and the SMA, and the segments of the stomach and duodenum remaining proximal to this region was dilated to a high degree. She underwent duodenojejunostomy. The patient was sent on the 7th day after the operation and was followed up after 6 month. She had no complaints and has gained 10 kg in controls carried out.

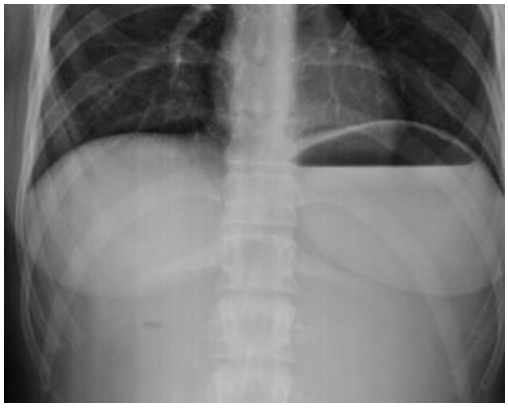

Figure 1: Stomach fundus gas on standing empty abdominal radiograph

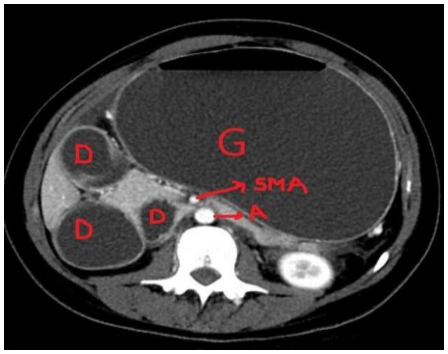

Figure 2: Axial contrast-enhanced CT scan
*SMA: Superior Mesenteric Artery, A: Aorta, G: Gaster, D: Duodenum

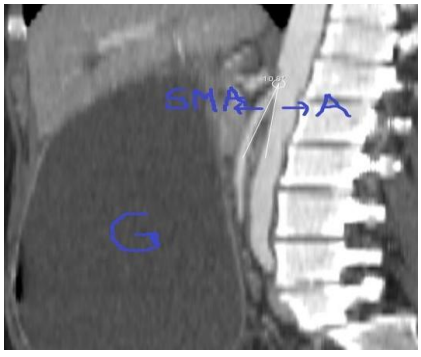

Figure 3: Sagittal contrast-enhanced CT image
*SMA: Superior Mesenteric Artery, A: Aorta, G: Gaster

DISCUSSION

First defined by Rokitanski in 1861 and known as Wilkie's syndrome, SMA syndrome is a condition which occurs due to the contraction of the angle between the two arteries (6°-25°) due to the loss of the fat pedicles between the aorta and the SMA. It results in the 3rd portion of the duodenum to get stuck [1]. Normally, the SMA leaves the aorta at an angle of 45 degrees (38-56°) and passes through the aortomesenteric vascular angle of the third portion of the duodenum. The fat and lymphatic tissue around the SMA normally provides a pillow function that prevents eccentric duodenal compression [1,5]. A loss of these pads due to reasons such as anorexia nervosa, weight loss, burns, trauma or prolonged bed rest is thought to be involved in the etiology of SMA [6]. According to radiological studies, the incidence of Wilkie's syndrome is between 0.2% and 0.78% [7]. It is reported that SMA syndrome is more common in women and two-thirds of

the cases occur between the ages of 10 and 39 years [8]. In this respect, the features of our patient were found to be compatible with literature.

While the disease has an acute and chronic form, the chronic form is more common [4]. In acute cases, there are symptoms of upper gastrointestinal tract obstruction, such as nausea, vomiting and abdominal pain, in addition to dilation in the stomach and duodenum [9]. Chronic SMA syndrome manifests itself with long-lasting, intermittent abdominal pain and abdominal discomfort, weight loss, nausea, and bilious vomiting. SMA syndrome, especially in cases with extreme weight loss, manifests itself mostly with chronic symptoms. In our patient, the disease developed acutely and upper gastrointestinal system obstruction findings such as abdominal pain, nausea were present.

Upper gastrointestinal system X-Ray, abdominal x-ray, CT, magnetic resonance (MR) angiography, CT angiography, ultrasonography, and endoscopy are used in the diagnosis of SMA syndrome [8]. The presence of gas ın the duodenum and stomach on an abdominal X-ray is an important finding supporting SMA syndrome [10]. Contrasted CT angiography is considered to be the most important diagnostic method used in the diagnosis of Wilkie's syndrome. Since, it shows the aortomesenteric angle, the distance between the aorta and the SMA, the amount of fat tissue, the location of the obstruction in the duodenum, and the dilation proximal to the obstruction [11]. SMAs diagnostic criteria are; aortomesenteric angle reduction <20 ° (normal: 28-65 °), aortomesenteric distance reduction <8 mm (normal: 10-28 mm) and gastric and proximal duodenal dilatation [12].

In the treatment of Wilkie's Syndrome, in the absence of aneurysm, mass-induced external compression or other clinical conditions requiring urgent surgical intervention, conservative medical treatment is initially recommended [2,8]. The main aim of medical treatment is to increase the mesenteric fat tissue and to remove the compression in the 3rd portion of the duodenum. In the presence of acute onset SMA syndrome, providing fluid and electrolyte balance, gastric and duodenal decompression with a nasogastric tube, positioning the patient in a face-down position or left side position is an effective treatment approach [13]. In cases where conservative treatment fails, surgical treatment is indicated [8]. Although various surgical techniques have been described in the literature, the technique of duodenojejunostomy, first described by Staveley in 1910, is now accepted [14]. Our patient underwent a duodenojejunostomy procedure.

CONCLUSION

As a result, in cases of extreme dilation of the stomach and duodenum being observed in the CT of patients presenting with complaints such as abdominal pain, nausea, and vomiting, a differential diagnosis of Wilkie's syndrome must be considered.

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