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Case Report

Wilkie's syndrome admitted for acute abdomen: A case presentation

Hasret Ayyıldız Civan, Didem Gülcü, Tülay Erkan, Fügen Çullu Çokuğraş, Tufan Kutlu

Department of Pediatrics, Division of Pediatric Gastroneterology, Hepatology and Nutrition, İstanbul University-Cerrahpaşa, Cerrahpaşa School of Medicine, İstanbul, Turkey

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Abstract

Superior mesenteric artery syndrome, also known as Wilkie's Syndrome, is a life threatening clinical entity which developes as a result of obstructed second or third part of duodenum compressed between aorta and superior mesenteric artery. In this rare syndrome, a rapid weight loss is accompanied by stomach ache, abdominal distension, lack of appetite, nausea and vomiting after meals. In patients admitted for acute abdomen, superior mesenteric artery syndrome should be included in the differential diagnosis in case of a preceeding rapid weight loss. X-ray of barium passage, abdominal ultrasound, gastroscopy, abdominal angio-tomography or abdominal magnetic resonance angiography may be useful for diagnosis. Conservative and surgical approaches are available for the treatment. In this report we aimed to emphasize that superior mesenteric artery syndrome cases may admit for acute abdomen and that superior mesenteric artery syndrome should be included in differential diagnosis.

Keywords: Acute abdomen, duodenal obstruction, superior mesenteric artery

Introduction

Superior mesenteric artery syndrome (SMAS), which is also known as Wilkie's syndrome because it was described by Wilkie as chronic duodenal ileus in 1927 for the first time, is a life-threatening clinical picture that develops as a result of obstruction of the second or third part of the duodenum compressed between the aorta and superior mesenteric artery (1). The first pediatric cases were reported by Prouty and Waskow in 1957 (2). Localization of the SMA below its normal position, presence of malrotation in the duodenum, short Treitz ligament, cancer leading to rapid weight loss, burns, previous surgical operation or psychiatric disorder and associated loss of mesenteric adipose tissue, aneroxia nervosa or presence of scoliosis may be involved in the etiology of this rare syndrome (3, 4). Advances in computed tomography and magnetic resonance imaging provided recognition of openness of the angle between the aorta and SMA and an increased diagnostic rate (5). In treatment, a conservative treatment approach is applied in acutely developing cases. Here, the objective is to provide nutritional support and regain loss of the adipose tissue pad, which is thought to lead to narrowing of the aorto-mesenteric angle by helping weight gain. Surgical treatment should be used in patients whose symptoms continue long term and who do not respond to conservative treatment. The most commonly preferred surgical method is side-to-side duodenojejunostomy (6). In this article, we report a patient who presented with a prediagnosis of acute abdomen and was diagnosed as having SMAS.

Case

A 16-year-old girl who had no known previous disease, presented with symptoms of flatulance, loss of appetite, and vomiting, which continued for the last four days. She was referred to the Pediatric Surgery Clinic of our hospital with a prediagnosis of acute abdomen because

Corresponding Author: Hasret Ayyıldız Civan E-mail: hasretayyildiz@yahoo.com
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of the presence of suspicious air-fluid levels on direct erect abdominal radiography. The patient was referred from the Division of Pediatric Surgery to the Division of Pediatric Gastroeneterology with a prediagnosis of inflammatory bowel disease because she had poor general status and had lost weight in the last few months. In her history, it was learned that she had no known previous disease, she had constipation which lasted for two weeks four months ago, and bloody stool twice; loss of appetite started in this period, she lost weight because of the loss of appetite, she preferred not to eat because she liked losing weight and thus she lost 15 kg in a short period. The patient started to visit a dietician to gain weight for the last three weeks and managed to gain 3 kg. She presented to hospital, when flatulence developed for the last one week and vomiting after meals started. In her familial history, there was no pathology except for the suspicion inflammatory bowel disease and related fistule operation in her father. On physical examination, she looked cachectic and pale. Body weight 39 kg (<3 percentile), height 163 cm (60 percentile). Her abdomen was relaxed, she had no tenderness or organomegaly. An examination of the other systems was found to be normal. In the laboratory tests, hemogram amd routine biochemical tests were found to be within the normal limits. Plain erect abdominal radiography did not show air-fluid level. Elective colonoscopy was performed to exclude inflammatory bowel disease because the patient was referred with a suspicion this diagnosis. During the procedure, abdominal guarding developed and plain erect abdominal radiography and abdominal ultrasonography were performed immediately. Air-fluid level or intraabdominal or subdiaphragmatic free air was not present on a plain erect abdominal radiograph. On abdominal ultrasonography, massive free fluid starting from the periumbilical level and filling all quadrants with a size of 10 cm at the widest part was observed in the abdomen and it was reported that it could be compatible with perforation. A naso-gastric catheter was placed and 4500 cc biliary fluid was drained in a few minutes. Following drainage, the patient's abdomen was relaxed and soft. Thus, it was thought that the intra-abdominal fluid observed on abdominal ultrasonography was gastric content and esophago-gastroduodenal radiography was obtained with a suspicion of SMAS. On the radiograph, it was found that the stomach and the 1st and 2nd part of the duodenum were markedly enlarged, the gastric outlet was not obstructed, the horizontal part of the duodenum had an appearance compatible with SMA compression, and passage in this part was delayed. On abdominal CT angiography, it was observed that the stomach and the first two segments of the duodenum were markedly enlarged, the 3rd part of the duodenum (horizontal segment) was compressed between the aorta and SMA, and the angle between the aorta and SMA was reduced to 5° (normal angle: 38-56°) (Figure 1) and a diagnosis of SMAS was made. A nasojejunal tube was placed and enteral nutrition was initiated in the patient. The patient tolerated enteral nutrition and she gained weight in the follow-up. Written informed consent was obtained from the patient.

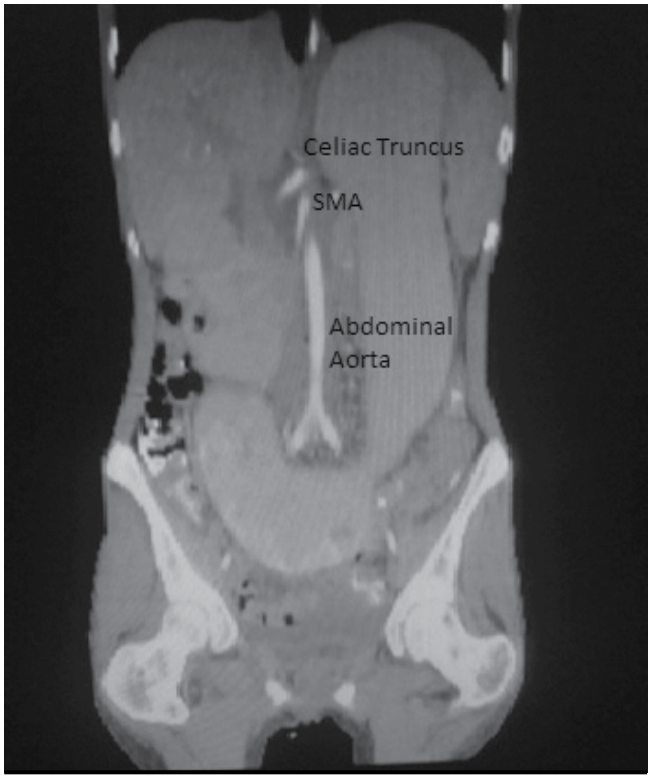

Figure 1. **Appearance of the duodenum compressed below the enlarged stomach and superior mesenteric artery on abdominal angio-tomography**

Discussion

Superior mesenteric artery syndrome is an acute or chronic picture that occurs as a result of compression of the second or third part of the duodenum between the aorta and the upper part of SMA (7). In our patient, abdominal CT showed that the 3rd part of the duodenum was compressed between the aorta and SMA.

The causes of superior mesenteric artery syndrome include anatomic conditions influencing vascular angle and various psychiatric and physiologic disorders leading to rapid weight loss (4, 8). Our patient lost 15 kg in the last four months and she voluntarily lost this

much weight. When evaluated by child psychiatry, it was thought that she might have anorexia nervosa.

Although there is no definite information about its frequency, findings supporting this diagnosis were found with a rate of 0.013-0.3% on barium radiographs in one study. It generally occurs in women aged between 10 and 30 years, as in our patient (7).

Superior mesenteric artery syndrome may present acutely or chronically. Its chronic form is observed more frequently. In the acute form of superior mesenteric artery syndrome, abdominal distension, persistent vomiting and epigastric pain are the most common symptoms. In the chronic form, epigastric pain, which generally starts postprandially, nausea, vomiting, loss of appetite and weight loss are observed (7). In our patient, loss of appetite and weight loss that were compatible with the chronic form of SMAS were found, but acute abdomen was considered when vomiting and swelling developed acutely and she was referred to the Division of Pediatric Surgery. She was then referred to the Division of Pediatric Gastroenterology with a prediagnosis of inflammatory bowel disease because air-fluid level was not found on plain erect abdominal radiography and there was a history of weight loss.

Investigations including CT angiography, color Doppler ultrasonography, MRI, MR angiography, endoscopic ultrasonography, gastric emptying scintigraphy, and esophago-gastroduodenal radiography may be used for the diagnosis. Observation of reduced aorto-mesenteric angle or gap on abdominal CT angiography and/or enlarged stomach and upper duodenal segment on barium passage radiography are significant for the disease (8). In our patient, it was stated that free abdominal fluid was observed on abdominal ultrasonography, but it was observed that this fluid was gastric content, which was extremely extended and reduced up to the pelvis on abdominal CT angiography. The aorto-mesenteric angle was reduced to 5° (normal angle: 38-56°). The stomach and 2nd and 3rd part of the duodenum were found to be extremely enlarged on esophaga-gastroduodenal radiography.

The essential difficulty in the diagnosis is the physicians' lack of consideration of the disease primarily. Our patient was referred to the Division of Pediatric Surgery with a prediagnosis of acute abdomen and then referred to us with a prediagnosis of inflammatory bowel disease.

In treatment, the first step is conservative approach in both acute and chronic cases. In conservative treatment, the objective is to provide nutritional support and to regain loss of the adipose tissue pad, which is thought to lead to narrowing of the aorto-mesenteric angle by helping weight gain (4). In this treatment, nasogastric decompression, intravenous fluid therapy, feeding with liquid foods and nasojejunal tube, switching to knee-chest position or maneuvers including lying on the right side first and then on the left side after meals and metoclopramide treatment are used (10). We initiated enteral nutrition in our patient by placing a nasojejunal tube with a conservative approach, in accordance with the literature. Our patient started to gain weight with enteral and parenteral nutrition. It was decided that a surgical approach was not needed because weight gain was good in the first month in the follow-up.

If weight loss that develops in a short time and flatulence after meals are present in the history in patients presenting with vomiting, Wilkie syndrome should definitely be considered in the differential diagnosis, and patients should be investigated in this regard.

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References

1. Wilkie DPD. Chronic duodenal ileus. Am J Med Sci 1927; 173: 643-9. [CrossRef]
2. Prouty M, Waskow, WL. Duodenal compression by the mesenteric artery. J Pediat 1957.p.734. [CrossRef]
4. Elbadaway MH. Chronic superior mesenteric artery syndrome in anorexia nervosa. Br J Psychiatry 1992; 160: 552-4. [CrossRef]

5. Unal B, Aktaş A, Kemal G, et al. Superior mesenteric artery syndrome: CT and ultrasonography findings. Diagn Interv Radiol 2005; 11: 90-5.
6. Zaraket V, Deeb L. Wilkie's syndrome or superior mesenteric artery syndrome: fact or fantasy? Case Rep Gastroenterol 2015; 9: 194-9. [CrossRef]
7. Welsch T, Büchler MW, Kienle P. Recalling superior mesenteric artery syndrome. Dig Surg 2007; 24: 149-56. [CrossRef]
8. Stheneur C, Rey C, Pariente D, Alvin P. Acute gastric dilatation with superior mesenteric artery syndrome in a young girl with anorexia nervosa. Arch Pediatr 1995; 2: 973-6. [CrossRef]
9. Lippl F, Hannig C, Weiss W, Allescher HD, Classen M, Kurjak M. Superior mesenteric artery syndrome: diagnosis and treatment from the gastroenterologist's view. J Gastroenterol 2002; 37: 640-3. [CrossRef]
10. Dietz UA, Debus ES, Heuko-Valiati L, et al. Aorto-mesenteric artery compression syndrome. Chirurg 2000; 71: 1345-51. [CrossRef]