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Superior Mesenteric Artery Syndrome - Process of Diagnosis and Treatment of Problematic Cases

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ABSTRACT

Superior mesenteric artery (SMA) syndrome is an angulation of SMA causes compression of the duodenum between the SMA and the aorta. The symptoms are not unique and can be seen in several other gastrointestinal pathologies like ileus, gastroesophageal reflux, pancreaitis etc. We report a 25-year-old female who presented with intermittent abdominal pain and intractable vomiting. The patient underwent Nissen-fundoplication, laparotomy, small bowel resection, and bridectomy operations. Also the patient received medical theraphies for pancreatitis, ileus and anorexia. This case emphasizes the challenges in the diagnosis of SMA syndrome and the need for increased awareness of this entity. This will improve early recognition in order to reduce irrelevant tests and unnecessary treatments.

Keywords: *diagnosis, ileus, pancreatitis, superior mesenteric artery syndrome*

ÖZ

Superior Mezenter Arter Sendromu - Sorunlu Olguların Tanı ve Tedavi Süreci

Superior mezenter arter (SMA) sendromu, duodenumun SMA'nın açılanması sonucu SMA ve aorta arasında sıkışmasıdır. Semptomlar özgül olmadığı için ileus, gastroözefagial reflü, pankreatit gibi diğer gastrointestinal patolojilerle karışabilmektedir. Aralıklı karın ağrısı ve yoğun kusma yakınması ile başvuran 25 yaşında kadın hastamızı sunduk. Hastamız öncesinde benzer yakınmalar nedeniyle Nissen Fundoplikasyon, laparatomi, ince bağırsak rezeksiyonu ve bridektomi ameliyatları geçirmiş. Hasta pankreatit, ileus ve anoreksia için medikal tedaviler almıştır. Bu olgu SMA sendromu tanısındaki zorlukları vurgulamaktadır ve bu sendormun varlığını konusunda dikkat çekmek için sunulmuştur. Bu konu hakkında farkındalık erken tanı ile birlikte ilişkisiz testleri ve gereksiz tedavileri azaltacaktır.

Anahtar kelimeler: *ileus, pankreatit, superior mesenter arter sendromu, tanı*

INTRODUCTION

Superior mesenteric artery syndrome in other names Cast syndrome, Wilkie syndrome, arteriomesenteric duodenal obstruction and chronic duodenal ileus is the compression of the third portion of the duodenum due to the narrowing of the angle between superior mesenteric artery and the aorta [(1)]. The symptoms of Wilkie syndrome are abdominal pain, nausea, anorexia, weight loss and vomiting. Because of its rarity and overlap with numerous other gastrointestinal pathologies like gastroesophagial reflux disease, pancreatitis, gastritis, ileus; the patients can be misdiagnosed. Delayed diagnosis is generally seen because of the ineffective therapies, lack of suspicion and inappropriate investigations [(2,3)].

Here we want to present a case with Wilkie syndrome who underwent several inappropriate investigations, ineffective therapies and unneccesary operations.

CASE REPORT

A 25-year-old women was referred to our clinic with epigastric pain, postprandial discomfort, bloating bilious vomiting, and inability to gain weight. Her symptoms improved with postural change to knee-chest position. In her medical history, the body mass index (BMI) of the patient was nineteen years old. She has been suffering from abdominal pain and retrosternal burning and bloating. She had undergone laparoscopic Nissen-Fundoplication 2 years ago. However 1 month later, the symptoms of bloating and postprandial discomfort had worsened. She had undergone laparotomy with suspicion of ileus. No pathology had been found but segmenter small bo-

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wel resection and anastomosis had been performed because of iatrogenic injury. Two months later, the patient underwent surgery because of ileus and only bridectomy was performed. Her symptoms did not improve. She was hospitalized for 10 days with the diagnosis of acute pancreatitis and received long term proton pump inhibitor theraphy. The patient was also consulted to psychiatry with the suspicion of anorexia neurosa and was followed-up by the psychiatrist for 6 months. The patient was admitted to emergency department for recurrent abdominal pain and subileus symptoms several times. When she was admitted to our department, she had been under liquid diet for 3 months and her BMI was 15.4. In the endoscopic evaluation, we observed megoduodenum and undigested food residues in the duodenum (Figure 1). With the suspicion of SMAS, we measured the angle between aorta and SMA 15º in the computer tomography (CT) (Figure 2). The patient was treated with Roux-Y duodenojejunostomy and has gained 8 kg weight in the follow up in six months.

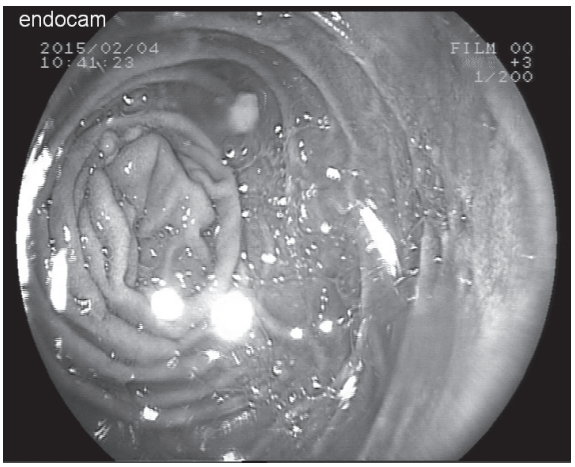

Figure 1. Endoscopic evaluation of the patient. Megaduodenum was seen.

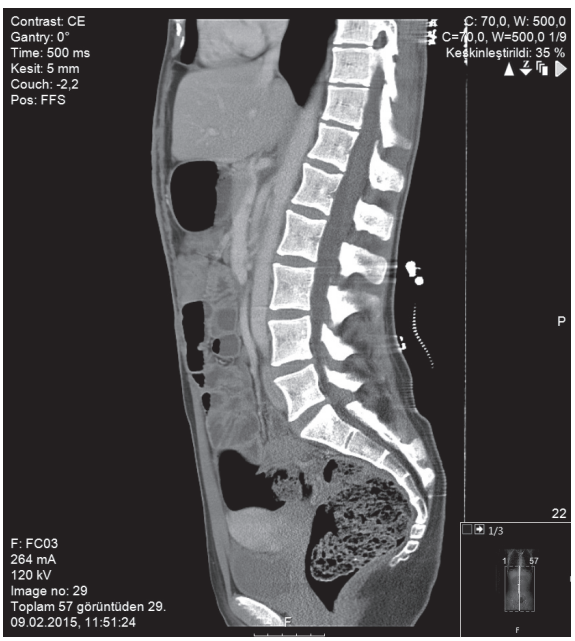

Figure 2. The computed tomography image show the narrowed angle between the superior mesenteric artery and the aorta.

DISCUSSION

Wilkie syndrome is caused by the decreased aortomesenteric angle. The controversy of this entity is due to the fact that its signs and symptoms were not regarded as unique, because they could be found in other circumstances. Several coexisting diseases had been published in the literature, but more than one misdiagnosis like in our case is rare in the literature. Surgery and severe weight loss are the blamed factors. Compression after corrective spinal surgery is the most described surgical cause of Wilkie syndrome. Wasting conditions auch as AIDS, malabsorbtion, cancer, cerebral palsy, cachexia, severe burns, anorexia nervosa, drug abuse, bariatric surgery, and abdominal trauma are blamed due to decreased fat pad between aortomesenteric angle [5]. Age and sex distribution may reflect the predisposing cause of condition. Although not knowing exactly, low BMI might be the predisposing factor.

The symptoms of Wilkie syndrome are confusing [4]. Epigastric pain, vomiting, hearthburn might be signs of other GI pathologies like gastroesophageal reflux as presented in the literature [5]. Endoscopy might show severe esophagitis and 24h pH monometer might support this diagnosis as in our patient. Careful evaluation of duodenum for dilatation or upper gastrointestinal series with delay in passage can show the diagnosis. The first operation of the patient had been fundoplication due to gastroesophageal reflux. However obstruction of third portion of duodenum and fundoplication exacarbated the symptoms. The next operation was laparotomy because of ileus. However it's hard to define Wilkie syndrome intraoperatively unless there's a suspicion or known pathology. After unfortunate operations, the symptoms of epigastric pain and relieving of symptoms after lying prone, the patient was treated with suspicion of pancreatitis. We

believed that the precipitating factor was her initial antireflux disease. The mechanism of Nissen fundoplication with coexisting Wilkie syndrome in our patient resulted in a closed loop obstruction which led to ileus and pancreatitis. This kind of situation was seen in the study of Petroysan et al. [6]. Pancreatitis in the absense of gallstone and alcohol, there might be several reasons. Although pancreatitis in eating disorders is a rare entiity, it can be seen as a results of pancreatic injury in malnutrition [7]. Pancreatitis in our patient was most likely secondary to abnormal pancreatico-duodenal reflux within the closed loop of the intestine. After unsuccessful treatments, the patient was suspected to have anorexia nervosa.

The diagnosis of duodenal obstructionis are made with X-ray studies or CT imaging. Failure of contrast passage beyond the third part of duodenum and the aortomesenteric angle between 9º and 22º are the diagnostic factors. Endoscopic evaluations shows megaduodenum in Wilkie syndrome [8]. In adult patients non-operative therapy is often prolonged the hospitalization peridon with low success rate varied between 14 and 71% [9]. Strong's operation, gastrojejunostomy, and duodenujejunostomy are the treatment options for Wilkie syndrome [10]. The roux-en-Y duodenojejunal bypass left no blind loop, with free drainage of not only the duodenum proximal but also distal to the compression site at the SMA. Lapaorscopic treatment is also a popular approach in Wilkie syndrome. In our case, becasue of the risk of intrabdominal adhesion due to previous operations, we performed laparotomy.

The diagnosis of Wilkie syndrome frequently relies on a high index of suspicion and is often made by a process of exclusion, resulting in ineffective symptomatic therapies and inappropiate investigations [4]. As in our patient, the treatment was not only delayed but also resulted in unneccesary operations.

In the evauation of the patients with epigastric pain bilious vomiting, pain relieving with lying prone or left lateral decubitis position, weight loss and nausea, SMAS should be kept in mind and megaduodenum or megabulbus should be searched during endoscopy. For this reason, detailed history should be taken and after careful endoscopic evaluation, CT images should be analysed.

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