An Unusual Cause of Abdominal Pain: Superior Mesenteric Artery Syndrome: Case Report

Karın Ağrısının Nadir Bir Sebebi: Superior Mezenter Arter Sendromu

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ABSTRACT We present a patient who suffered from bowel obstruction and discuss associated findings of Superior Mesenteric Artery (SMA) Syndrome. A 19-year-old female admitted to emergency service with epigastric pain. On physical examination there was a splash sound from her stomach and the abdomen was tender. An abdominal tomography scan showed a very large stomach. She was referred to the general surgery department. At operation it was seen that the third portion part of the duodenum was compressed between superior mesenteric artery and the aorta. SMA syndrome is a rare disorder and is under-diagnosed in emergency medicine. Patients present with symptoms of intestinal obstruction and treatment includes nasogastric decompression as well as intravenous fluids. Surgery must be planned if the conservative treatment fails. Early diagnosis can save the life. Emergency department physicians also should keep this diagnosis in mind in the differential diagnosis of abdominal pain.

Key Words: Superior mesenteric artery syndrome; abdominal pain; emergency service, hospital

ÖZET Bu olgu sunumunda barsak obstrüksiyonundan kaynaklanan şikayetlerle başvuran bir hasta ve Superior Mezenterik Arter (SMA) Sendromu ile ilişkili bulguları tartışılacaktır. 19 yaşındaki kadın hasta acil servisimize epigastrik ağrı ile başvurmuştur. Fizik muayenede çalkantı (splash) sesi ve hassas karın mevcuttu. Batın tomografisi çok büyük bir mide göstermekteydi. Hasta Genel Cerrahi bölümüne konsülte edildi. Operasyon sırasında duodenum 3. kısmının aort ve superior mezenter arter tarafından baskılandığı görüldü. SMA sendromu, acil tıpta nadir görülen ve tanı konulamayan bir sendromdur. Hastalar barsak obstrüksiyon semptomları ile başvurur ve tedavisi nazogastrik dekompresyon yanısıra intravenöz sıvı uygulamasını içerir. Konservatif tedavi başarısız olursa cerrahi tedavi planlanmalıdır. Erken tanı hayat kurtarıcı olabilir. Acil servis doktorları karın ağrısı ayırıcı tanısında bu antiteyi de akla getirmelidir.

Anahtar Kelimeler: Süperior mezenterik arter sendromu; karın ağrısı; acil servis, hastane

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The superior mesenteric artery syndrome (SMAS, also known as Wilkie's syndrome, aortomesenteric compression, arteriomesenteric duodenal compression or duodenal vascular compression) is a rare condition that results from vascular compression of the third part of the duodenum, in the angle between the aorta and the superior mesenteric artery.[1] The syndrome was first described by Von Rokitansky in 1842 and since then about 400 cases has been reported in the literature but SMAS is not well recognised and often diagnosed late when till the patients are far

advanced with their symptoms.[2] The classic symptoms are occasional epigastric pain, postprandial discomfort, nausea and vomiting of bile-stained material, fullness, and weight loss.[3] Although about 400 cases are described in the English language literature, many researchers have doubted the existence of the SMA syndrome as a real entity; indeed, some investigators have suggested that the SMA syndrome is overdiagnosed, because it is confused with other causes of megaduodenum.Nevertheless, this syndrome still eludes most clinicians, and patients suffer from the disease for a long time before a firm diagnosis is reached. Therefore, any report illustrating the clinical or imaging appearance of this syndrome may be helpful in facilitating earlier diagnosis.[4]

Here we present a patient who suffered from bowel obstruction and discuss associated findings of SMAS. It has been presented as poster presentation in the "EuSEM (European Society for Emergency Medicine) 2012 Congress (3-6 October 2012). The case report was conducted in accordance with the principles of the Declaration of Helsinki. Informed consent was taken from the patient.

CASE REPORT

A 19-year-old female presented to the emergency department of Marmara University Medical Faculty Pendik Research and Training Hospital complaining of epigastric pain, nausea and vomiting. Symptoms had started 2 weeks earlier and persisted despite supportive treatment in another hospital with a diagnosis of peptic ulcers. She was hospitilized in the internal medicine ward in another hospital for one day and treated for dehydration for 5 days. During hospitalization, no pathological findings could be identified and the patient was discharged. Symptoms persisted and she was eventually admitted to us. Her past medical history included intermittent epigastric abdominal pains and intractable intermittent vomitting. The vomitus usually consisted of mostly undigested food eaten over the past days. She had rapid loss of weight (7 kg in one month). There was no history of an eating disorder, nor medication taken and no remarkable family history. On physical examination there was a splash sound from her stomach and the abdomen was tender. With the prediagnosis of pylorus stenosis or ileus, abdominal computed tomography was required. However due to intractable vomiting oral contrast was not given. The abdominal computed tomography scan (Figure 1, 2) showed a massively distended stomach and proximal duodenum. The third portion of the duodenum was compressed between the SMA and the aorta. The superior mesenteric vein was also compressed and there was a very large stomach, a dilatation of the first and second parts of the duodenum. This was reported as a superior mesenteric artery syndrome. For gastrointestinal decompression, a nasogastric tube was inserted. She was given nothing orally, and intravenous fluids were given. From the nasogastric tube we drained approximately 2500 mL initially, then the drainage continued. She was referred to the general surgery deparment and after failure of conservative treatment for 2 days in the ward, she was subjected to a duodenojejunostomy. During the operation it was seen that the third portion of duodenum was compressed between superior mesenteric artery and aorta. No complication occured. Enteral feeding started in the second post-operative day and she was sent home in the 5th post-operative day with well-beeing.

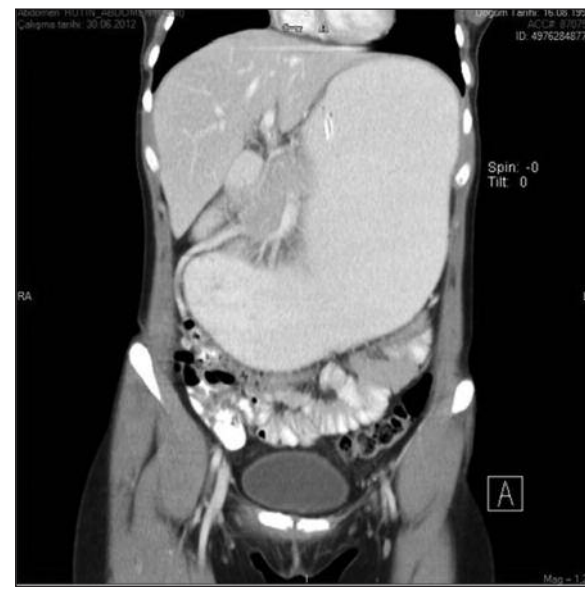

FIGURE 1: Over distension of the stomach.

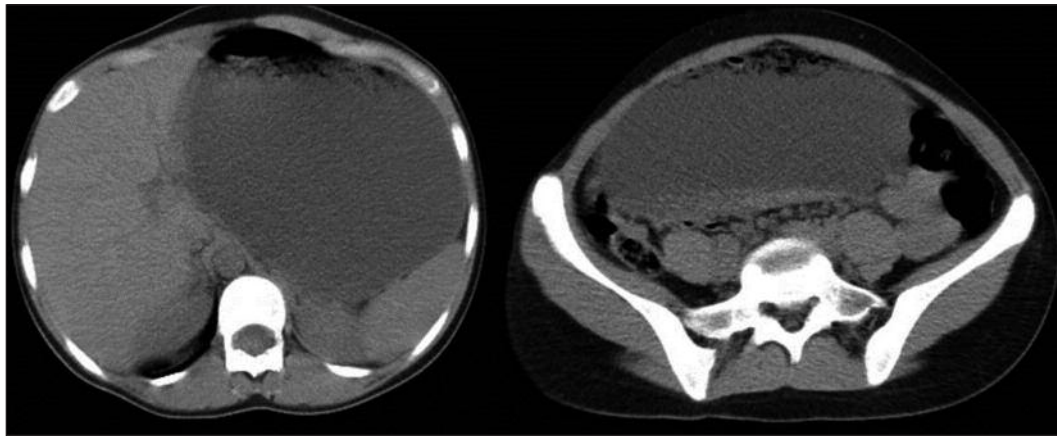

FIGURE 2: The distension of the intestinal system up to third part of the duedenum.

DISCUSSION

Rokitansky is generally credited with the earliest description of the compression of the third part of the duodenum by the superior mesenteric artery (SMA). Surprisingly, there was little interest in the subject until 1927, when Wilkie reported seven instances of superior mesenteric artery syndrome (SMAS).[5] Since then, this entity has remained a subject of controversy. The signs and symptoms characterising the syndrome were not regarded as unique. Many observers considered them to be mere manifestations of another abdominal process and remained unconvinced of the reality of the syndrome. They regarded the role of the SMA in producing duodenal obstruction as circumstantial, especially as in many of the reported cases, the obstruction was later found to be due to neoplastic or inflammatory disease, or electrolyte imbalance.

In Turkey, the precise incidence of this entity is unknown. Females are more affected by SMA syndrome. The SMAS usually occurs in older children and adolescents. The acute type is less common and may present as a surgical emergency. In its acute or chronic forms, the syndrome becomes clinically manifest with predominantly postprandial epigastric pain, a sense of repletion, and vomiting. The precursor symptom of the syndrome is postprandial abdominal pain with associated nausea, vomiting and anorexia. The pain is characteristically relieved by a prone, knee-chest or left lateral decubitus position. Patients may complain of chronic abdominal pain, the severity of which depends on the cause of obstruction and may run for months or even years.[1,2] In rare conditions the syndrome may present acutely, mimicking upper intestinal ileus.

It is caused by mechanical compression of one of the most fixed parts of the duodenum (third), between the aorta and the origin of superior mesenteric artery. Normally this angle is between 25-60 degrees with an opening of 10-20 mm, which, in the case of this syndrome, gets reduced to between 6-15 degrees for the angle and 2-8 mm for aortomesenteric aperture.[6] The width of the aortomesenteric angle is related to the body mass index. Lack of retroperitoneal and periduodenal fat pads can lead to a more acute angle resulting in duodenal "clamping." SMAS is thus triggered by any condition compromising the normal fat cushions and the mesenteric angle. Fat cushion loss can be seen in catabolic and postoperative states, in the presence of congenital anomalies or trauma.[3]

With the advent of modern radiological techniques in the 2000s, computed tomography (CT) has proved successful in providing diagnostic insight into cases of SMAS. Then CT or MR angiography became additional radiological diagnostic methods. CT was able to demonstrate the characteristic duodenal distension and the close proximity of superior mesenteric vessels and aorta

simultaneously. Its advantages include that it is a safe, rapid and a relatively noninvasive technique.[6,7]

The clinical diagnosis can be confirmed by radiologic studies in 95% of cases. The following radiologic criteria have been established for the diagnosis of SMAS:[8]

- dilatation of the first and second portions of the duodenum, with or without gastric dilatation;
- abrupt vertical and oblique compression of the mucosal folds;
- antiperistaltic flow of barium proximal to the obstruction, producing a to-and-fro movement;
- delay of 4 to 6 hours in transit through the gastroduodenal region;
- relief of obstruction when the patient is placed in a position (prone or knee-chest) that diminishes the drag of the small-bowel mesentery.

CT is also useful in demonstrating both the duodenal distention, the anatomy and the relationships of the superior mesenteric vessels and also for excluding other pathology.[9]

Treatment is initially conservative. Surgical treatment is indicated if conservative treatment fails or if there is severe progressive weight loss, pronounced duodenal dilatation with stasis and complicating peptic ulcer disease.[3,4]

In conclusion, SMAS is a rare disorder and is under-diagnosed in emergency medicine. Appropriate history and physical examination will facilitate the diagnosis. SMAS should be ruled out in patients with postprandial abdominal pain, vomiting and weight loss. Nowadays, noninvasive techniques such as CT or MRI scans can establish the diagnosis.

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