A Case of Hide and Seek: An Obstructive Duodenal Tumor Resembling Superior Mesenteric Artery Syndrome

Muzaffer Ece Hakan Sahin*, Esma Gulda Altnunoglu, Fusun Erdenen and Ibrahim Taskin Rakici
Istanbul Training and Research Hospital, Istanbul, Turkey

*Corresponding author: Muzaffer Ece Hakan Sahin, Istanbul Training and Research Hospital, Istanbul, Turkey. Tel: 009053684444608; E-mail: m.ecehakan@gmail.com

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Abstract

Proximal intestinal obstruction caused by compression of the third part of duodenum between aorta and Superior Mesenteric Artery (SMA) is referred as Superior Mesenteric Artery Syndrome (SMAS). Being a rare and hardly diagnosed entity, we present a 69 year old woman referring with intractable nausea and vomiting, sense of fullness, and weight loss. In upper endoscopy, stomach was distended. Computerized tomography showed a decrease in the angle between aorta and SMA, creating a compression of SMA on duodenum misleading us to SMAS. The patient didn't ease with conservative treatment. Endoscopy is performed with a longer probe for further evaluation and a tumoral mass is found in 3rd and 4th parts of duodenum. It appeared that duodenum tumor blocked the passage, caused weight loss, created SMAS clinic. Positron Emission Tomography-Computed Tomography (PET-CT) exhibited metastasis. The case was inoperable therefore General Surgery Department decided on performing gastrojejunostomy as palliative treatment.

Keywords: Superior mesenteric artery syndrome; Duodenum tumor; Vomiting; Gastrojejunostomy

Abbreviations:

SMA: Superior Mesenteric Artery; SMAS: Superior Mesenteric Artery Syndrome; PET-CT: Positron Emission Tomography-Computed Tomography; NG: Nasogastric.

Introduction

Superior Mesenteric Artery Syndrome (SMAS) is a rare cause of proximal bowel obstruction and is linked to morbidity and mortality when diagnosis is delayed. The clinical entity caused by compression of the third portion of duodenum between abdominal aorta and superior mesenteric artery. It causes early satiety and vomiting, leads to anorexia. The symptoms are nonspecific, therefore it is hardly diagnosed. Etiology varies from anorexia nervosa, traumas to cancers. Malignant duodenum tumors are also extremely uncommon tumors of the body. The diagnosis is overlooked if it is not particularly searched for. SMAS should be considered as a syndrome of variable symptoms, yet it can also be the result of another hidden diagnosis.

Case Report

A 69 year old female patient presented to our emergency department with the complaint of nausea, projectile vomiting and weight loss. On admission, the patient's condition was in a moderate status and she was conscious. The alimentary vomiting was present, in projectile motion, precipitating by food intake. The vomit appeared brown in colour, coffee ground appearance, misleading for gastrointestinal bleeding. Abdominal pain in epigastric region occurred occasionally, relieved by vomit discharge. She didn't have fever, night sweats but have 6 kg of weight loss in two months. She had a medical history of type 2 diabetes mellitus, essential hypertension. She had a mitral valve replacement (mechanical valve) sixteen years ago, and using warfarin since. She had no other operation history. Her family history was unremarkable.

Her vital signs were stable; her blood pressure was 119/79 mmHg, 80/min heart rate, and body temperature 36.8°C. The physical examination revealed normal systems except that of signs of dehydration as a consequence of vomit and not enough intake of liquid. A Nasogastric (NG) tube was replaced for gastric decompression and bag was full of discharge, therefore there were no abdominal distention. Epigastrium was tender in palpation, there was no defense or rebound. There were no signs of infection, no signs of increased cranial pressure related projectile vomit.

The results were as followed. The initial complete blood count suggested anemia with hemoglobin of 11.8 g/dL, white blood cells of 8.120 × 10^9/L. Liver function tests were normal. Urea was 87 mg/dL and creatinine was 1.95 mg/dL, because of dehydration. The urine examination was positive for red blood cells, leukocytes, ketone, and protein. The NG tube was inserted, accumulation was measured and symptomatic treatment has begun. She had taken the NG tube out for 3 times, every time followed by inevitable courses of vomiting, and an ease after replacement of NG tube. Bilious gastric fluid was drained every day.

Direct abdominal graph revealed abdominal distention. Esophagus stomach graphy is performed, found normal. Upper endoscopy was performed which showed stomach distention only. The short scope proceeded till the first part of duodenum. Abdominal computed tomography screening exhibited dilated stomach, dilatation at proximal part and stenosis near the third part of the duodenum. The 3rd part of duodenum was compressed between SMA and abdominal aorta, best observed in sagittal sections. This compression narrowed the angle between aorta and SMA (aortomesenteric angle) to 13.6° and aortomesenteric distance to 7 mm (Figure 1). Based on these findings and the symptoms, the case was inferred as SMAS. Supportive
treatment as total parenteral nutrition and electrolyte replacements were implemented. She was consulted to the general surgery for consideration of surgical options, but preliminary supportive treatment was recommended.

In the 4th day of her hospitalization, she complained about a feeling of deterioration. The electrocardiography revealed new T segment negativity at DII, DIII, V3, V4, V5, and V6 along with multiple ventricular extra systoles. Sequential troponin values were normal, but having DM and valve replacement history, she was considered as acute coronary syndrome and therefore coronary angiography was performed. Having 50% ejection fraction, left atrial dilatation and left ventricle segmental contraction defect on echocardiography, her angiography showed 30-40% blockage, she was evaluated as noncritical coronary artery disease.

Upper abdominal ultrasonography demonstrated sub-capsular solid nodular lesions in left lateral lobe and neighbour of left portal vein.

Endoscopy was repeated with a longer colonoscopy probe which reached till 3rd and 4th parts of duodenum presenting a tumoral mass that blocked the passage to further. This time abdominal computed tomography scan was repeated with oral and intravenous contrast administration. In CT scan, at the level of distal to the 3rd part of duodenum, near the Trietz ligament, as dilated duodenum was suddenly narrowed, there revealed a soft tissue consistency mass in retroperitoneal region invasive throughout paraduodenal pancreatic tail (Figure 2). This mass was consistent with duodenal tumor (Figure 3). The duodenal lumen was occluded near totally.

In the light of these findings, PET-CT was planned and the patient was transferred to General Surgery Clinic.

In the surgery it was observed that proximal duodenum was distended. In exploration, liver left and right love upper surface reflected tumoral implantations. In the area of the Trietz ligament, at the pancreas tail level in an invasive 7 × 10 cm sized tumoral formation was present, completely obstructing the lumen. The neighbour meso of small intestine has metastatic implant. No other implant was observed.
on peritoneal surfaces or omentum. Nodule on left lobe inferior surface with PET-CT involvement excised and sent to frozen section examination which was interpreted as metastasis. The patient was evaluated as inoperable and gastrojejunosutomy was performed. Surgery confirmed the point of obstruction was at the point of crossing by the SMA.

In the histopathological examination, the tumoral cells of liver biopsy were detected as CEA diffuse (+), CK7 (+), TAG72 (+), CK20 (-), CDX2 (-), AMACR (-), P53 (+). Ki-67 proliferation index was high. The findings were matching with adenocarcinoma infiltration.

Following the operation, the patient was followed in intensive care unit for 2 days, and discharged from the hospital postoperative day 10. She died 2 months later in intensive care unit with multiple metastasis and multiorgan failure.

Discussion

Proximal intestinal obstruction caused by compression of the third part of duodenum between aorta and SMA is referred as SMAS (Cast syndrome, Wilkie syndrome, arteriomesenteric duodenal obstruction or chronic duodenal ileus) [1]. This compression leads to an obstruction in duodenum, creating a life threatening condition for the patient. The symptoms can vary between postprandial epigastric pain, early satiety, abdominal discomfort, dyspepsia or reflux to severe nausea, unstoppable bilious emesis and inevitable weight loss [2]. These nonspecific symptoms often lead to a delay in diagnosis. Furthermore, SMAS can coexist with pancreatitis, peptic ulcer, intra-abdominal inflammation and tumors presenting with these symptoms [3]. Investigation is done through gastroscopy with biopsy and contrast-enhanced CT essentially. The angle between aorta and SMA is normally 25–60 degrees. It corresponds to the aortomesenteric distance of 10–28 mm. In SMAS, these values range between 6–15° and 2–8 mm respectively. There is a correlation between the clinical symptoms and distance reduction [4]. There are many reasons researched as causatives. Weight loss, causing diminished mesenteric and retroperitoneal fat tissue attributed to malabsorption, anorexia nervosa, traumas, burns or cancers are the major ones [5,6]. Our patient has weight loss due to catabolic effect of duodenal tumor leading to the fatty tissue loss. Patients generally become comfortable with left lateral decubitus, prone or knee-chest position by decreasing the pressure applied on the duodenum by the aorta [6].

Treatment of SMAS can be either conservative or surgical. Supporting by gastric compression, feeding in small amounts multiple times a day in left lateral or prone position or aggressive feeding with nasojejunal feeding bypassing the obstruction by tube [7]. Surgery is indicated in conditions such as long standing disease with progressive weight loss and failed conservative treatment [8]. In our case, before the surgery total parenteral nutrition becomes the only choice till the surgery. In a study involving a group of eight SMAS performed by Merrett and colleagues the prognosis of a patient receiving preoperative TPN shows a better prognosis than other patients [5], suggesting that feeding with TPN is beneficial as it was in our case. Duodenoejunosutomy is the preferred choice of surgery, and laparoscopic approaches are also practiced, gradually replacing the conventional one [8].

Primary adenocarcinoma of duodenum is a rare disease with nonspecific symptoms, corresponding to leading to a diagnosis at advanced stage. Being seen mostly in the second part of duodenum [9]. It accounts for <2% of gastrointestinal tract tumors and up to 42% of small bowel malignancies. Along with the contrast-enhanced CT and enteroclysis; capsule endoscopy, double-balloon enteroscopy and computed-tomography enterography are new modalities that are started to be used in its diagnosis. The optimal treatment is surgery and decreased survival correlates with advanced stage and incomplete resection [10].

The described case manifests a duodenal primary adenocarcinoma initially diagnosed as SMAS. First endoscopy with short probe couldn't show the pathology so CT was performed which reflected the narrowed aortomesenteric angle driving us to SMAS. Supportive treatment was given according to the first diagnosis, but no relief was observed in 10 days. Operation was decided. In preoperative preparations, endoscopy was repeated with colonoscopy probe to reach further and an obstructive mass was encountered. The new diagnosis was proved by contrast enhanced CT followed by PET-CT. Biopsy was taken during gastrojejunosutomy operation. The result was adenocarcinoma of duodenum.

Conclusion

In our case, SMAS appeared as a complication of adenocarcinoma of the duodenum rather than a primary case. Although its pathophysiology isn't known exactly, it is thought that the loss of angle between aorta and SMA may occur due to increased catabolism of adenocarcinoma and consequent weight loss. Adenocarcinoma of duodenum may go unnoticed because of the difficulties in its diagnosis and sometimes it may manifest itself as SMAS. For this reason, when SMAS diagnosis is considered, it should be investigated together with other SMAS causatives, and surgical treatment should be considered as the next option if conservative treatment is unsuccessful.

References