

Duodenal Emphysema Complicated with Superior Mesenteric Artery Syndrome in a Patient with Cerebral Paralysis: A Case Report

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Superior mesenteric artery syndrome (SMAS) is characterized by an arteriomesenteric duodenal compression commonly resulting from significant weight loss. Vomiting is the most frequent symptom. SMAS can be complicated by massive gastric dilatation. Patients with cerebral palsy have various factors that can predispose them to SMAS. In this paper, we report a rare case of SMAS complicated by duodenal, peritoneal and retroperitoneal emphysema in a patient with cerebral paralysis, referring to the relevant literature. In this case, severe vomiting associated with epilepsy and weight loss may have contributed to the development of duodenal emphysema.

Key words: superior mesenteric artery syndrome, duodenal emphysema, cerebral paralysis

INTRODUCTION

Superior mesenteric artery syndrome (SMAS) is a condition where compression of the third portion of the duodenum occurs between the superior mesenteric artery (SMA) and the spine and aorta [1]. It frequently occurs in patients who have had rapid weight loss. The symptoms of SMAS include vomiting, nausea, early satiety, anorexia, and abdominal pain [1, 2]. SMAS can be complicated by massive gastric dilatation and, in very rare cases, by gastric emphysema or portal venous gas [3-6]. Both gastric and duodenal emphysemas caused by SMAS are extremely rare [3]. In this paper, we report a case of duodenal, peritoneal and retroperitoneal emphysema complicating SMAS in a patient with cerebral paralysis. The patient had marked gastric dilatation without wall pneumatosis, and his condition improved with conservative treatment.

CASE DESCRIPTION

A 28-year-old male, whose mother had an uneventful pregnancy and delivery, developed cerebral paralysis due to subdural effusion at the age of 7 months. He had profound epilepsy, cerebral paralysis, and developmental impairment. An elemental diet tube and intravenous hyperalimentation catheter were inserted on initial admission. Six months later, he was admitted to our hospital because of severe epilepsy. His body weight was 40 kg and body mass index was 14.3 on admission. The patient had several episodes of vomiting. Two days after admission, an abdominal computed tomographic (CT) scan was performed because of marked gastric dilatation and suspected duodenal emphysema detected on radiographs (Fig. 1). He

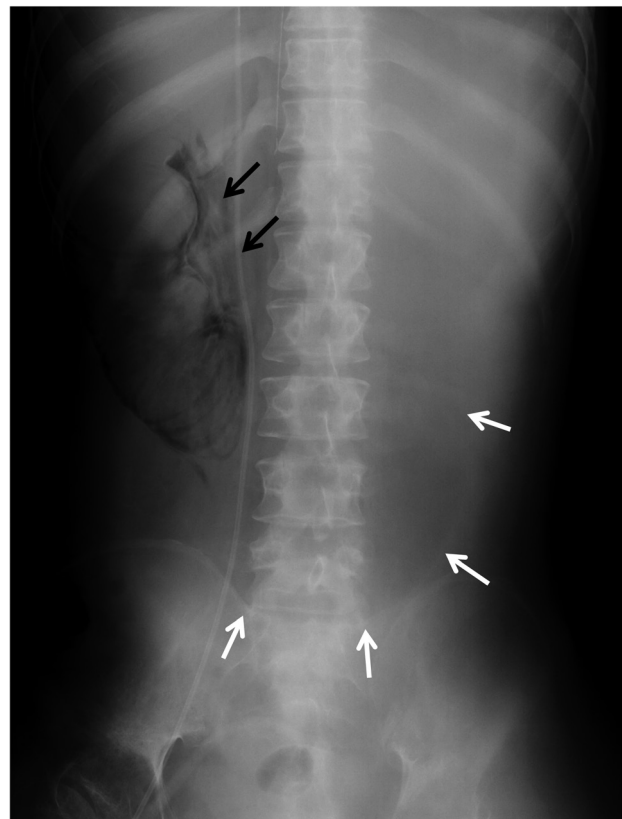


Fig. 1 Abdominal radiograph shows gastric dilatation (white arrows), duodenal wall gas, and abnormal linear gas in the left side of the duodenal second position (black arrows).

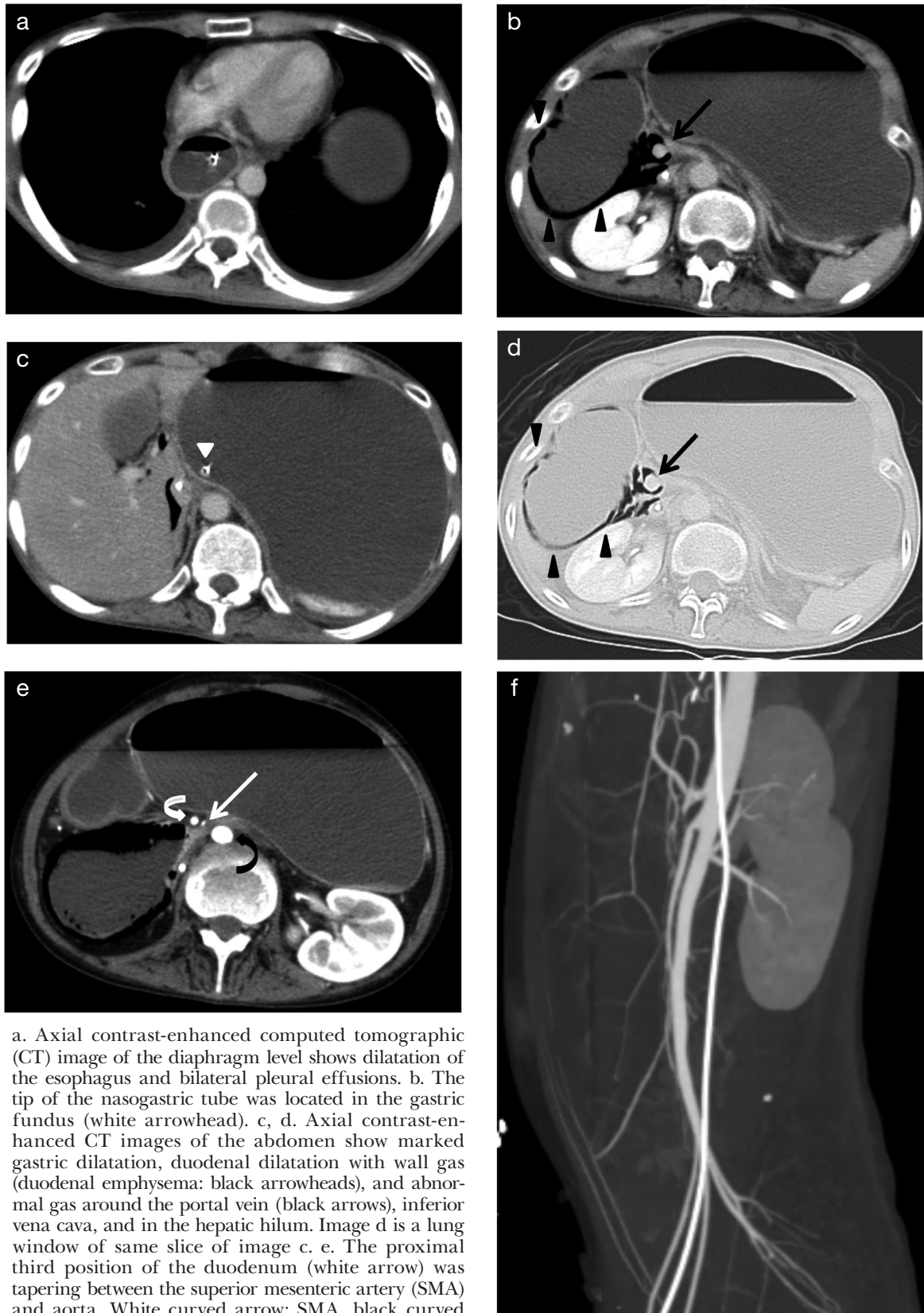


Fig. 2 a. Axial contrast-enhanced computed tomographic (CT) image of the diaphragm level shows dilatation of the esophagus and bilateral pleural effusions. b. The tip of the nasogastric tube was located in the gastric fundus (white arrowhead). c, d. Axial contrast-enhanced CT images of the abdomen show marked gastric dilatation, duodenal dilatation with wall gas (duodenal emphysema: black arrowheads), and abnormal gas around the portal vein (black arrows), inferior vena cava, and in the hepatic hilum. Image d is a lung window of same slice of image c. e. The proximal third position of the duodenum (white arrow) was tapering between the superior mesenteric artery (SMA) and aorta. White curved arrow: SMA, black curved arrow: aorta. f. Maximum intensity projection image of CT angiography shows aorto-SMA angle was 10°.

had no abdominal pain. Laboratory studies were unremarkable, with exception of C-reactive protein (11.5 mg/dl). CT images showed marked dilatation of the stomach, the tapering of the proximal duodenal third portion to the point of obstruction between the SMA and aorta, and emphysema of duodenum, peritoneal

and retroperitoneal cavities (Fig. 2a-e). The gastric wall was also enhanced. The tip of the nasogastric tube was observed in the gastric fundus (Fig. 2b). CT angiography showed a marked decrease in the angle between the SMA and aorta (Fig. 2f). These CT findings were compatible with SMAS. Upper gastrointestinal barium

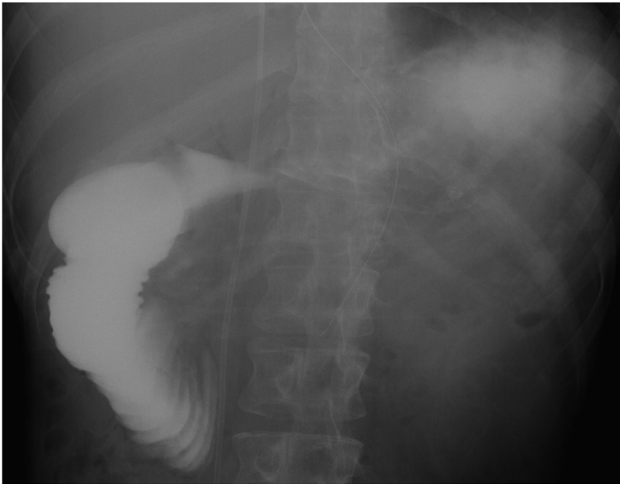


Fig. 3 Upper gastrointestinal barium study through deep insertion of nasogastric tube shows an abrupt cut off of the proximal third portion of the duodenum with a largely dilated proximal duodenum and stomach.



Fig. 4 Abdominal radiograph 3 days after CT scanning showed improvement of duodenal dilatation and emphysema.

study showed abrupt cut out of the proximal third portion of the duodenum (Fig. 3). A nasogastric tube was deeply placed and 2800 cc of gastric juice was aspirated. An abdominal radiograph 3 days after the first CT scan showed marked improvement of the gastric dilatation and duodenal emphysema (Fig. 4). However, the patient underwent gastrostomy for weight gain and recovered uneventfully.

DISCUSSION

SMAS is a rare form of upper gastrointestinal obstruction with a reported incidence of 0.013%-0.3% [2]. The clinical presentation of both acute and chronic forms of SMAS includes nonspecific symptoms of high intestinal obstruction, such as postprandial epigastric pain, early satiety, upper abdominal distension, and vomiting [1-4]. Any factor that sharply narrows the aortomesenteric angle can cause an entrapment and a compression of the third portion of the duodenum. Predisposing factors believed to be involved include a narrow aortomesenteric angle, abnormally high fixation of the ligament of Treitz, lumbar lordosis, anorexia, spinal conditions, and rapid growth with weight variations [1, 3]. Many factors occurring in children who have cerebral palsy may predispose them to SMAS [3]. In the present case, the predisposing factors of SMAS were low body weight and vomiting associated with epilepsy. The patient was unable to eat adequately on his own because of dysphagia caused by cerebral paralysis, and was therefore reduced to a skeleton. Gastric dilatation grew worse because of the shallow position of the nasogastric tube in his stomach as result of frequent vomiting.

Gastroduodenal dilatation associated with SMAS is common; however, gastroduodenal emphysema and

pneumoperitoneum are rare. Gastric emphysema with portal venous gas associated with SMAS is very rare, and only three cases have been previously reported [3-5]. The first case occurred spontaneously in an elderly male without anorexia [5]. The second case was associated with anorexia and bulimia and retrocolic duodenojejunoscopy was performed [4]. The most severe case of gastroduodenal emphysema with portal venous gas was associated with spastic quadriplegia and underwent gastrostomy [3]; portal venous thrombosis occurred due to sepsis, and SMAS was likely due to stenosis of the gastric fundus which caused fundoplication. Only two cases of duodenal emphysema complicating SMAS, including the present case, have been described [3]. A case of marked gastroduodenal dilatation without wall gas, pneumoperitoneum and pneumomediastinum has been reported, but the reason of those coexistences was unclear [7]. Pneumoperitoneum is not always associated with gastroduodenal emphysema. Two major theories exist on the pathogenesis of pneumatosis intestinalis, i.e., the bacterial and mechanical theories [8]. The mechanical theory of mucosal disruption suggests that the loss of intestinal mucosal integrity allows normal gas to enter the intestinal wall, particularly under increased intraluminal pressure, as in the present case. This was the likely cause in addition to gas coming out of the hepatoduodenal ligament and retroperitoneal cavity additionally. Extension of air into the peritoneal and retroperitoneal spaces after duodenal perforation due to an ulcer is a common CT finding [9]. In the present case, duodenal emphysema was observed, but gastric emphysema did not occur. Gastric emphysema with gastric dilation is more common than duodenal emphysema. In contrast to the previous cases, the present case was mild. In the present

case, duodenal emphysema was not caused by ischemic or inflammatory change but by the mechanical distention of the duodenum. Peritoneal and retroperitoneal emphysema might be occurred before gastric emphysema. Therefore, we considered that the condition of duodenal, peritoneal and retroperitoneal emphysema in the present case may have been “benign”.

Confirmation of the diagnosis of SMAS requires precise imaging. Barium studies revealed the obstruction of barium flow at the site where the SMA crosses the third portion of the duodenum and dilatation of the first and second positions of the duodenum [3]. Ultrasound examination is useful for evaluating the aorto-SMA distance by showing decrease in the angle between the SMA and aorta and can also be used to detect SMAS complications [3]. The diagnosis of SMAS is best confirmed using CT angiography, which can not only determine the measurement of the aorto-SMA distance, but also the extent of duodenal distension and the amount of intra-abdominal and retroperitoneal fat [10]. The mean aorto-SMA angle was 9° and 44° in patients with SMAS and controls, respectively, using CT angiography [10]. The mean aorto-SMA distance was 8.1 and 19.6 mm in patients with SMAS and controls, respectively, with measurements taken at the level of the duodenum [10]. In the present case, the aorto-SMA angle was 10° and the aorto-SMA distance was 5.2 mm.

The management of a patient with SMAS requires both the identification and reduction of risk factors [1-6]. In the acute setting, manual decompression of the duodenum and stomach with a nasogastric tube is often preferred for treating symptoms [1-4, 11]. Surgery is indicated for patients who do not improve by conservative management [11]. Similarly to the present case, one of the three cases with gastric emphysema and portal venous gas was treated conservatively [3]. The other two cases underwent operations; in one case, retrocolic duodenojejunostomy was performed and in the other case, manual compression was performed at laparotomy [4, 5]. Complications of gastric emphysema with portal venous gas and thrombosis are usually associated with a high morbidity and mortality rate in adults [11]. Patients with SMAS and gastric emphysema with portal venous gas should be carefully managed. Depending on its cause, SMAS can be potentially life-threatening condition that requires prompt diagnosis

and management.

CONCLUSION

We reported a rare case of duodenal, peritoneal and retroperitoneal emphysema and marked gastric dilatation complicating SMAS in a patient with cerebral paralysis. The patient had some predisposing factors of SMAS, including low body weight and vomiting associated with epilepsy, and the conservative treatment of the condition was successful.

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REFERENCES

- 1) Gustafsson L, Falk A, Lukes PJ, Gamkhou R. Diagnosis and treatment of superior mesenteric artery syndrome. *Br. J Surg* 1985; 71: 499-501.
- 2) Biank V, Werlin S. Superior mesenteric artery syndrome in children: A 20-year experience. *J Pediatr Gastroenterol Nutr* 2006; 42: 522-525.
- 3) Rod J, Sarnacki S, Petit T, Ravasse P. Portal venous gas and thrombosis complicating superior mesenteric artery syndrome (Wilkie's syndrome) in a child. *J Pediatr Surg* 2010; 45: 826-829.
- 4) Lim JE, Duke GL, Eachempati SR. Superior mesenteric artery syndrome presenting with acute massive gastric dilation, gastric wall pneumatosis, and portal venous gas. *Surgery* 2003; 134: 840-843.
- 5) Sakamoto Y, Mashiro K, Matsumoto H, Hara Y, Kutsukata N, Yamamoto Y. Gastric pneumatosis and portal venous gas in superior mesenteric artery syndrome. *Indian J Gastroenterol* 2006; 25: 265-266.
- 6) Kim T, Kim HU, Song HJ. A case of gastric emphysema in anorexia nervosa presenting as acute gastric distension. *K J Gastroenterol* 2012; 60: 315-319 (in Korean).
- 7) Kensinger CD, Mukherjee K, Neelson WH, Solorzano CC. Superior mesenteric artery syndrome presenting with pneumoperitoneum and pneumomediastinum. *The American surgeon* 2013; 79: E240-242.
- 8) Lee KS, Hwang S, Hurtado Rúa SM, Janjigian YY, Gollub MJ. Distinguishing benign and life-threatening pneumatosis intestinalis in patients with cancer by CT imaging features. *AJR Am J Roentgenol* 2013; 200: 1042-1047.
- 9) Yegan N, Auh YH, Fisher A. Extension of air into the right peritoneal space after duodenal perforation: CT findings. *Radiology*. 2009; 250: 740-748.
- 10) Mathenge N, Osiro S, Rodriguez II, Salib C, Tubbs RS, Loukas M. superior mesenteric artery syndrome and its associated gastrointestinal implications. *Clinical anatomy* 2013.
- 11) Faberman RS, Mayo-Smith EE. Outcome of 17 patients with portal venous gas detected by CT. *AJR Am J Roentgenol* 1997; 169: 1535-1538.