The Korean Journal of *Helicobacter* and Upper Gastrointestinal Research
Vol. 12, No. 1, 50-53, March 2012

A Case of Primary Aortoenteric Fistula Mimicking Duodenal Subepithelial Tumor

Primary aortoenteric fistula is a fatal condition which poses a considerable diagnostic challenge because of its infrequency and nonspecific presentation. Here, the authors report the case of an 83-year-old man who presented with hematemesis and melena. During endoscopy, a 3 cm sized subepithelial mass with central ulceration was found in the second portion of the duodenum. At first, bleeding from a duodenal subepithelial tumor, such as, gastrointestinal stromal tumor was suspected. An endoscopic hemostasis trial failed and computed tomography (CT) scan was performed prior to possible angiographic embolization. The CT scan revealed the mass lesion observed by endoscopy as an aneurismal dilatation with fistula formation to the distal second portion of the duodenum. The patient succumbed to hypovolemic shock while preparations were being made for emergency surgery. This case provides an example of a primary aortoenteric fistula mimicking a duodenal subepithelial tumor during endoscopy. **(Korean J *Helicobacter* Up Gastrointest Res 2012;12: 50-53)**

Key Words: Intestinal fistula; Hematemesis; Duodenal neoplasms

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Received : December 9, 2011
Accepted : February 2, 2012

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INTRODUCTION

Primary aortoenteric fistula (PAEF) is an extremely rare condition and usually occurs as a complication due to compression of the adjacent bowel by an aortic aneurysm. The reported incidence of PAEF is only 0.04～0.07% in the general population, making this condition highly uncommon entity for inclusion in the differential diagnosis of gastrointestinal bleeding.[1] About 300 cases of PAEF have been published, and these demonstrate that its preoperative diagnosis is extremely difficult. Therefore, it is important that the possibility of an aortoenteric fistula be suspected initially. PAEF presents mainly as a gastrointestinal hemorrhage, but the active bleeding site is difficult to locate during endoscopy.[2] Here, we report a rare case of PAEF mimicking a duodenal subepithelial mass.

CASE REPORT

A 83-year-old man presented at our hospital with an episode of hematemesis. He had been suffered from melena for 5 days before visiting our hospital. He had no specific medical history and was a current smoker, and was conscious but looked pale. Initial vital signs were as follows: temperature 36.6°C, pulse rate 90 beats/min, and blood pressure 140/90 mmHg. Physical examination demonstrated an elderly man in acute distress. Conjunctiva appeared slightly anemic, and the abdomen was soft, nontender, with normal bowel sounds, and without organo-

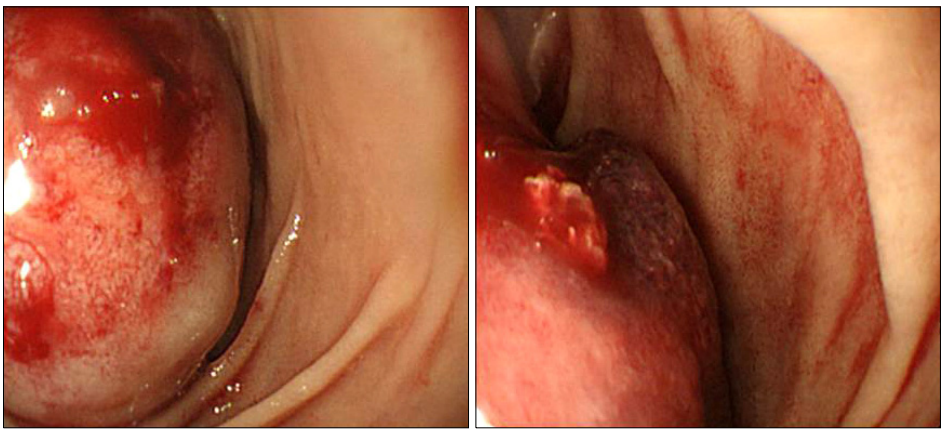

Fig. 1. Endoscopic findings. About 3 cm sized submucosal mass with central exposed vessel was noted.

megaly or a pulsatile mass. A rectal examination revealed dark fresh blood-colored stools.

Initial laboratory findings were as follows: hemoglobin 12.5 g/dL, white blood cell count 21,300/mm^3, platelet 158,000/mm^3, Na/K/Cl/TCO2 141/4.2/103/14 mEq/L, blood urea nitrogen/creatinin 12/1.3 mg/dL, prothrombin time 90% (international normalized ratio 1.06).

Following initial management involving transfusion and hydration, the patient underwent emergency endoscopy, which revealed a 3 cm sized subepithelial mass in the second portion of the duodenum. At the top of the mass, a shallow ulceration and an exposed vessel were seen. Blood gushed out of the vessel (Fig. 1). The first endoscopic impression was of active bleeding from an exposed vessel at ulceration on a subepithelial tumor in the second portion of the duodenum. An endoscopic trial for injection sclerotherapy failed to achieve bleeding control, and thus, an abdominal computed tomograpy (CT) scan was performed prior to possible angiographic embolization. However, CT scan revealed an abdominal aortic aneurysm connected to the second portion of duodenum with fistula formation (Fig. 2), confirmed by the extravasations of a large amount of contrast dye into the second portion of the duodenum.

On returning from the CT scan, the patient suddenly entered a state of shock. A massive blood transfusion did not recover his blood pressure and 1 hour later, he experienced cardiac arrest, and died while preparations were being made for emergency surgery to treat the abdominal aortic aneurysm.

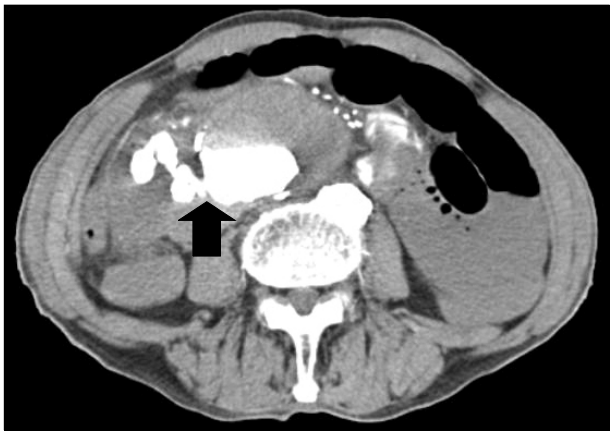

Fig. 2. Abdomen CT findings. Huge abdominal aortic aneurysm was seen and the aneurysm was connected to 2nd portion of duodenum with fistula formation (arrow).

DISCUSSION

PAEF is defined as a fistula between the native aorta and gastrointestinal tract, and is a extremely rare cause of gastrointestinal bleeding.[1] A large autopsy series reported an incidence of PAEF of 0.04～0.07%.[1,2] PAEF arises from several underlying diseases though atherosclerotic abdominal aortic aneurysm is the most common cause.[3] Other less common conditions include infective aortitis due to syphilis or tuberculosis, carcinoma, ulcers, foreign bodies (needle, fish bone, or chicken bone),[4] and complications of radiotherapy.[5] Fistulas can form between the aorta and any organ from the esophagus to the sigmoid, but about 80% of PAEFs affect the esophagus and duodenum.[1] The most frequent site is in the third portion of the duodenum. Overall, 54% of PAEFs occur at the fixed retroperitoneal portion of the distal duodenum, anterior to the aorta and two-thirds of these occur in the lowest third part of the duodenum.[1,2,6] PAEFs are often fatal, and have a total mortality rate of 80～100% and a perioperative mortality rate of 18～63%.[1,7]

The clinical manifestations of PAEFs vary, and the classical triad of abdominal pain, upper gastrointestinal bleeding, and an abdominal pulsating mass is overemphasized, because this triad is present in less than 25% of cases.[1,8] Gastrointestinal bleeding is the most common symptom, and irrespective of its location, spontaneous bleeding followed by massive, and sometimes fatal bleeding, is typical of a PAEF.[8] Spontaneous bleeding is usually minor and self-limiting because thrombus formation plugs the fistula as a result of hypotension and low local blood flow. Thrombus may plug the fistula and temporarily stop overt bleeding; however, in this condition, excessive volume therapy and endoscopy may promote fatal exsanguinations.[1,9] In 70% of patients the time between initial spontaneous bleeding to exsanguinations is more than 6 hours, in 50% over 24 hours, and in 29% more than 1 week.[10] In our patient, this took about 5 days, and he presented at our hospital at the time of exsanguinations. However, it is possible that he experienced recurrent episodes of spontaneous bleeding before the fatal incident.

The diagnostic procedure is chosen based on clinical condition.[1,11] Recent CT improvements have resulted in it becoming the preferred initial diagnostic test. CT is less invasive than endoscopy or angiography, is easily performed, and as compared with endoscopy or angiography, poses no danger of thrombus dislodgement. Furthermore, a CT scan with iodinated contrast is the most suitable diagnostic test when an aortoduodenal fistula is suspected.[12] Pathognomonic signs are air residing within the aortic wall and the presence of contrast within the gastrointestinal tract.[1] Occasionally, CT scans show contrast in the duodenum, and thus, confirm the diagnosis, whereas subtle signs, such as, gas within the calcified wall of an aneurysm with an adherent bowel loop, extraluminal gas in the periaortic region, bowel wall thickening over the aorta, or the disruption of the aortic fat cover could also suggest PAEF.[1,13] In described case, we suspected an aorto-duodenal fistula because contrast dye was extravasated into the duodenal lumen. Angiography is another possible diagnostic modality, but its detection rate is only 20%.[1] Furthermore, angiography cannot visualize a fistula during an episode of active bleeding.

In hemodynamically stable patients with gastrointestinal bleeding, endoscopy is the preferred primary procedure and provides valuable information.[1,11,14] However, the detection rate of aorto-enteric fistula is only 25% because stable patients do not often exhibit active bleeding.[1] Furthermore, endoscopic visualization of a fistula in the lower third of the duodenum, which is the most common site for PAEF, is extremely difficult.[7,10] Endoscopic findings suggesting PAEFs are variable. Active bleeding site and adherent blood clots are the most common findings, but few PAEF cases have been diagnosed due to the endoscopic visualization of active bleeding site from a fistula because hemodynamically stable patients rarely show active bleeding. Mucosal deficit made by fistula is often misdiagnosed as erosion or ulcer.[14] Less than 5% of reviewed cases presented with erosion and an eccentric pulsating mass protruding through the gastrointestinal tract as in our case,[15-17] and of these three cases, two were aorto-esophageal fistulas. During endoscopy, esophageal compression, a pulsating subepithelial mass, and an ulcer breaking the mucosa membrane, with adherent clots have been observed at the esophagus.[15,17] Ikeda et al.[16] reported a primary aorto-duodenal fistula associated with an inflammatory abdominal aortic aneurysm, and during endoscopy, in this case, a subepithelial pulsating mass was observed at the duodenum, and PAEF was also suspected after a CT scan. In the present case, PAEF was initially misdiagnosed endoscopically as ulceration associated with a subepithelial tumor, such as, a gastrointestinal stromal tumor (GIST). Duodenal subepithelial tumors include leiomyoma, GIST, lipoma, carcinoid tumor, and others. Often ulcerations are accompanied with a duodenal subepithelial mass caused by mucosal irritation.[18] A mass-like presentation is not a common finding of PAEF, and thus, it is important that PAEF be suspected in patients with a history of abdominal aortic aneurysm presenting with recurrent gastrointestinal bleeding. During endoscopy, a bluish distended pulsating mass in the duodenum, as was observed in the present case, could suggest fistula formation between an aneurysm and the duodenum.

Endoscopy is also especially useful to exclude other causes of acute upper gastrointestinal bleeding, such as, gastroduodenal ulcers, esophageal pathology, or varices. However, endoscopy could delay surgical intervention and lead to misdiagnosis when PEAF coexists with other bleeding sites, such as, ulcers or erosions.[14] Upper gastrointestinal endoscopy with pediatric colonoscopy[19] and capsule endoscopy[8] could also be used to visualize the full length of the distal duodenum.

Due to the 100% mortality rate of aortoesophageal fistulas, treatment is absolutely essential. PAEF can be treated by open surgical correction or endovascular stent placement. However, although the open surgical correction of aortoenteric fistulas is

conventional, this procedure is associated with high morbidity and mortality.[20] We stress that early diagnosis and surgical intervention are crucial for patient survival.

To our knowledge, there were about 8 cases of aortoenteric fistula reported in Korea. Only 4 of 8 cases were PAEF.[21-23] Similar to our case, the majority of cases suffered severe hematemesis, melena. Most cases were diagnosed by abdominal CT scan, but, in 2 cases, upper endoscopy was performed. In both 2 cases, adherent blood clots and blood oozing were noted in 2nd portion of duodenum. None of these cases presented as mass like lesion in duodenum.

A delay in its diagnosis, which is all too common, may partly explain the high morbidity and mortality of PAEF. Therefore, PAEF must be considered a possible etiology of massive gastrointestinal bleeding in elderly patients with a known history of abdominal aortic aneurysm, unless another pathologic source is indicted by endoscopy or CT.

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