INTRODUCTION

A primary aortoenteric fistula (PAEF) is a rare but often life-threatening cause of massive gastrointestinal bleeding [1-4]. PAEFs have a mortality rate of nearly 100% in the absence of surgical intervention, and diagnosis is not established preoperatively [2-5]. Diagnostic procedures are often non-confirmatory and can sometimes impede urgently needed surgical intervention [3,4,6]. Although it is infrequent, gastrointestinal endoscopy may be a double-edged sword when PAEFs coexist with multiple bleeding sites [7]. Here, we report such a case in which the cause of death was massive gastrointestinal bleeding due to a PAEF after leaving the hospital.

CASE REPORT

A 68-year-old man went to a hospital after suffering from melena for several days. He was conscious but looked pale. He had a blood pressure of 120/60 mmHg. His hemoglobin level was 5.7 g/dL and his hematocrit was 19.8%. An abdominal examination appeared normal, and no abdominal mass was observed. An emergency gastrointestinal endoscopy revealed a bleeding ulcer in the second part of the duodenum (Figure 1A). Other potential sources of acute bleeding were excluded. The patient was diagnosed with a probable duodenal ulcer, which was the presumed source of his bleeding. Endoscopic hemoclippling was performed (Figure 1B), and the patient received a blood transfusion and hemostatic agents. Following these procedures, the patient had no signs of hemorrhaging and recovered from anemia.

After a follow-up endoscopy on the 13th day (Figure 1C), the patient was discharged from the hospital. However, the following day, the patient complained of lumbago and abdominal distension. In the middle of the night, he suddenly entered into a state of shock, experienced a cardiac arrest, and was taken to the hospital. In the emergency room, a nasogastric tube was inserted and
nasogastric aspiration was performed producing 880 mL of blood.

**Autopsy findings**

The patient measured 162 cm in height and weighed 58 kg. Postmortem hypostasis on the back was very slight. No petechiae were observed in the palpebral conjunctivae. External examination revealed no injuries except for those caused by clinical procedures performed in the emergency room. Internal examination revealed that the gastrointestinal tract was dilated and contained 2200 mL of clotted blood between the stomach and jejunum. No abdominal organ injury and blood were found in the abdominal cavity. An abdominal aortic aneurysm (AAA) was located above the bifurcation of the aorta and resembled a fusiform swelling, 4 cm in diameter and 5 cm in length. An AAA with atherosclerosis thrust forward, adhering firmly to the digestive duodenal wall (Figure 2). The inferior mesenteric artery was hardly identified due to fibrous adhesion. The internal surface of the AAA had highly-calcified atheromatous ulcers covered with mural thrombus (Figure 3). A pinhole rupture was located on the third part of the duodenal mucosa (Figure 4) and fistulized into the adjacent AAA. A scarred ulcer with a hemoclip was observed on the second part of duodenal mucosa (Figure 5). No other origin of bleeding was found in the gastrointestinal tract.

**Microscopic investigations**

Histological examination revealed a fibrous adhesion between the aorta and duodenal serosa. The aneurysm contained atheromatous degeneration covered by a mural thrombus with a layer of fibrin. The internal and external elastic membranes were destroyed and the media of the artery was thinned under the most advanced plaque. Fibroblast proliferation and granulation tissue were revealed in the adventitia of the aorta and perivascular tissue. Inflammation of the duodenum was more serious at the serosa, but there were no signs of inflammation of the duodenal mucosa (Figure 6). Because repair reaction of the aorta was broader than that of
DISCUSSION

A PAEF, defined as a communication between the native aorta and gastrointestinal tract, is a rare cause of gastrointestinal bleeding [2,3,8,9]. A large autopsy series reported an incidence of PAEF of 0.04%-0.07% [1,2,7]. A fistula most commonly originates from an AAA, of which 85% are atherosclerotic. AAA is a common disease among middle-aged and older subjects, and its clinical consequences depend on its location and size. The major complications of AAA include rupture into the peritoneal cavity, occlusion of a branch vessel and embolism from atheroma or mural thrombus. PAEF is the rarest complication of AAA. The most frequent site of a fistula is the third part of the duodenum, as in the current case [1-4,10]. The literature reports that PAEFs are often fatal, with a total mortality rate of 80%-100% and a perioperative mortality rate of 18%-63% [1,3,8,11]. However, the actual incidence and mortality rates are not known because many patients die of PAEFs before they have been correctly diagnosed [9,10].

The classical triad of symptoms, i.e. gastrointestinal bleeding, abdominal pain, and a pulsating abdominal mass is overemphasized [1,3,4], as it occurs in less than 25% of PAEF cases [8]. The diagnosis of a PAEF is difficult because of its nonspecific and subtle clinical presentation [5,4]. However, PAEFs usually present with a herald bleeding prior to exsanguination [1-4,9]. Herald bleeding is usually minor and self-limiting, and it is probably due to a spasm of the intestinal wall musculature in response to sudden distention [1,3]. Herald bleeding is usually minor and self-limiting, and it is probably due to a spasm of the intestinal wall musculature in response to sudden distention [1,3]. Bleeding can be further limited by hypotension and thrombus formation [1-3]. Consequently, excessive volume therapy and endoscopy may promote fatal exsanguination [1,12]. The time interval between a herald bleeding and exsanguination is known to range from hours to months [1-4,9]. The interval was about 2 wk in the current patient. When the patient dies of fatal exsanguination after going to the hospital due to a herald bleeding, his family may suspect an error in medical treatment.

Frequently, the choice of a diagnostic procedure is based on the clinical condition [1,3,5]. Diagnostic imaging techniques, such as contrast-enhanced computed tomography (CT) and angiography, are useful investigation modalities for identifying PAEFs [1-4]. Even if there is limited bleeding at the examination, CT might reveal the size, location, and degree of calcification of an AAA. Fibrous adhesion between the aorta and duodenum might facilitate PAEF identification. In a hemodynamically stable patient with gastrointestinal bleeding, endoscopy is the preferred primary procedure which provides valuable information [1-3,5]. However, endoscopy rarely reveals confirmatory evidence of a PAEF because stable patients do not often have an active bleeding [2,3,11,13]. Additionally, endoscopic visualization of a fistula that is present...
lower than the third part of the duodenum, which is the most common site of PAEF occurrence, is extremely difficult\cite{2,4,14}. In the clinical setting, the absence of identifiable bleeding lesions with initial gastrointestinal endoscopy is regarded by some as a strong indicator for laparotomy\cite{3,13}. Recently, since the introduction of capsule endoscopy for clinical use, small bowel bleeding from the ampulla of Vater into the terminal ileum can be easily visualized and defined\cite{15}. Capsule endoscopy, which non-invasively captures images of the gastrointestinal mucosa without any pressure load, might be the best strategy for identifying a PAEF. However, if upper endoscopy or colonoscopy detects other coexisting bleeding sites in PAEF patients, the findings may be misleading\cite{3,4,6,7,13}. Furthermore, concomitant gastrointestinal lesions are occasionally found in PAEF patients, and this incidence is 25% as indicated in previous series\cite{5,7,13,16}. In the current patient, a bleeding ulcer was identified as the source of gastrointestinal bleeding. Hence, the possibility of a PAEF might have been overlooked. Ultimately, the key to early diagnosis of PAEF is an endoscopist’s heightened index of suspicion\cite{4,5,13}. Endoscopists need to recognize PAEFs as a potential cause of gastrointestinal bleeding.

REFERENCES

1. Saers SJ, Scheltinga MR. Primary aortoenteric fistula. Br J Surg 2005; 92: 143-152
2. Lemos DW, Raffetto JD, Moore TC, Menzoian JO. Primary aortoduodenal fistula: a case report and review of the literature. J Vasc Surg 2003; 37: 686-689
3. Tareen AH, Schroeder TV. Primary aortoenteric fistula: two new case reports and a review of 44 previously reported cases. Eur J Vasc Endovasc Surg 1996; 12: 5-10
4. Sweeney MS, Gadacz TR. Primary aortoduodenal fistula: manifestation, diagnosis, and treatment. Surgery 1984; 96: 492-497
5. Gelister JS, Fov JA. Primary aortoenteric fistula. J R Soc Med 1987; 80: 459-460
6. Delgado J, Jotkowitz AB, Delgado B, Makarov V, Mizrahi S, Szendro G. Primary aortoduodenal fistula: Pitfalls and success in the endoscopic diagnosis. Eur J Intern Med 2005; 16: 363-365
7. Peck JJ, Eidemiller LR. Aortoenteric fistulas. Arch Surg 1992; 127: 1191-1193; discussion 1193-1194
8. Voorhoeve R, Moll FL, Bast TJ. The primary aortoenteric fistula in The Netherlands--the unpublished cases. Eur J Vasc Endovasc Surg 1996; 11: 429-431
9. Finch L, Heathcock RB, Quigley T, Jiranek G, Robinson D. Emergent treatment of a primary aortoenteric fistula with N-butyl 2-cyanoacrylate and endovascular stent. J Vasc Interv Radiol 2002; 13: 841-843
10. Mirarchi FL, Schatzle MD, Mitre RJ. Primary aortoenteric fistula in the Emergency Department. J Emerg Med 2001; 20: 29-27
11. Dossa CD, Pipinos II, Shepard AD, Ernst CB. Primary aortoenteric fistula: Part I. Ann Vasc Surg 1994; 8: 113-120
12. Tozzi FL, da Silva ES, Campos F, Fagundes Neto HO, Lucon M, Lupinacci RM. Primary aortoenteric fistula related to septic aortitis. Sao Paulo Med J 2001; 119: 150-153
13. Duncan JR, Renwick AA, Mackenzie I, Gilmour DG. Primary aortoenteric fistula: pitfalls in the diagnosis of a rare condition. Ann Vasc Surg 2002; 16: 242-245
14. Korkut AK, Arpinar E, Yasar T, Guney D. Primary aortoduodenal fistula complicated by abdominal aortic aneurysm. J Cardiovasc Surg (Torino) 2000; 41: 113-115
15. Nakamura T, Terano A. Capsule endoscopy: past, present, and future. J Gastroenterol 2008; 43: 93-99
16. Jones AW, Kirk RS, Bloor K. The association between aneurysm of the abdominal aorta and peptic ulceration. Gut 1970; 11: 679-684