Life-threatening Duodenal Ulcer Bleeding from a Ruptured Gastroduodenal Artery Aneurysm in a Patient with Neurofibromatosis Type 1

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Vasculopathy is rarely reported in neurofibromatosis type 1, but when it occurs it primarily involves the aorta and its main branches. Among vasculopathies, aneurysmal dilatation is the most common form. Although several case reports concerning aneurysms or pseudoaneurysms of visceral arteries in neurofibromatosis type 1 patients have been reported, there are no reports describing gastroduodenal artery aneurysms associated with neurofibromatosis type 1. We experienced a case of life-threatening duodenal ulcer bleeding from a ruptured gastroduodenal artery aneurysm associated with neurofibromatosis type 1. We treated our patient by transarterial embolization after initial endoscopic hemostasis. To our knowledge, this is the first reported case of its type. High levels of suspicion and prompt diagnosis are required to select appropriate treatment options for patients with neurofibromatosis type 1 experiencing upper gastrointestinal bleeding. Embolization of the involved arteries should be considered an essential treatment over endoscopic hemostasis alone to achieve complete hemostasis and to prevent rebleeding. (Korean J Gastroenterol 2015;66:164-167)

Key Words: Neurofibromatoses; Duodenal ulcer; Hemorrhage; Aneurysm, ruptured; Embolization, therapeutic

INTRODUCTION

Neurofibromatosis type 1 is an autosomal dominant disorder of variable penetration affecting 1 in 3,000 people that predominantly involves abnormal growth of neuroectodermal tumors in the peripheral nervous system. Other clinical manifestations include café-au-lait spots, Lisch nodules of the iris, meningeval tumors and skin fold freckling. Besides these features, vasculopathy sometimes occurs with neurofibromatosis. The pathogenesis of neurofibromatosis vasculopathy is not completely understood but may be associated with dysfunction of the neurofibromin in vascular smooth muscle and endothelial cells. Neurofibromatosis type 1 is caused by mutations of NF1 gene, which encodes neurofibromin. Aneurysm formation in neurofibromatosis type 1 may be attributable to friable vascular structure secondary to arterial dysplasia or vascular invasion by neurofibroma. Vascular lesions occur in 3.6% of patients with neurofibromatosis.
fibromatosis type 1, and most involve the aorta and its main branches.\textsuperscript{7,8} The renal artery is most frequently affected, usually resulting in secondary hypertension, particularly in young patients.\textsuperscript{8} Complications of vascular involvement include aneurismal dilation, arteriovenous fistula, arteriovenous malformations, vascular stenosis, and rupture.\textsuperscript{9} Among such vasculopathies, vascular aneurysms are most common.\textsuperscript{7} Several cases of aneurysms or pseudoaneurysms of visceral arteries in type 1 neurofibromatosis patients have been reported, with involved arteries including the intercostal, extracranial, internal pudendal and gastrointestinal arteries.\textsuperscript{10-13} However, there are no reports regarding gastroduodenal artery aneurysms associated with neurofibromatosis type 1.

We experienced a case of life-threatening duodenal ulcer bleeding resulting from a ruptured gastroduodenal artery aneurysm associated with neurofibromatosis type 1. We treated our patient by transarterial embolization (TAE) after initial endoscopic hemostasis. To the best of our knowledge, this is the first case of duodenal ulcer bleeding from a ruptured gastroduodenal artery aneurysm in a patient with neurofibromatosis type 1. This case suggests that high levels of suspicion and prompt diagnosis are required to select appropriate treatment options for such patients, and that the embolization of the involved arteries should be an essential treatment over endoscopic hemostasis alone to accomplish primary hemostasis and to prevent rebleeding in such cases.

CASE REPORT

A 43-year-old man visited the emergency department for sudden onset of hematemesis three hours before his visit. He was admitted twice to another hospital for duodenal ulcer bleeding (one month and two years prior) and was managed with endoscopic hemostasis. He had a known history of neurofibromatosis, diagnosed 25 years ago. He denied consumption of alcohol, tobacco, or any medication. He had a number of cutaneous neurofibromas on his neck and trunk (Fig. 1). Physical examination revealed hyperactive bowel sounds, and the patient complained of mild tenderness on the upper abdomen without any rebound tenderness. There were no signs of ulcer perforation or peritonitis. Upon arrival at our hospital, the patient was in a state of shock and had a blood pressure of 70/60 mmHg and a heart rate of 102 beats/min. The patient’s laboratory data on admission included a white blood cell count of 16,400/mm\textsuperscript{3}, a hemoglobin concentration of 8.1 g/dL and a platelet count of 221,000/mm\textsuperscript{3}. Other findings, including a chemical battery, PT, aPTT and electrolytes, were not remarkable. After a large volume of fluid resuscitation, upper gastrointestinal endoscopy was performed to detect the cause of the patient’s gastrointestinal hemorrhage and for hemostasis. Endoscopy revealed a small active ulcer (3×7 mm) with a visible vessel in the stenosed duodenal bulb (Fig. 2). During epinephrine injection, pulsatile bleeding began at the injection site. Argon plasma coagulation, injection of additional epinephrine, and

Fig. 1. Photograph of the patient shows a number of cutaneous neurofibromas on the neck and trunk.

Fig. 2. Endoscopy upon admission reveals a small ulcer (3×7 mm) in the duodenal bulb with visible vessel.
endoscopic hemoclipping were tried and hemostasis was successful. Intravenous pantoprazole (Pacific Pharm., Seoul, Korea) was administered with high-dose continuous infusions (i.e., pantoprazole 80 mg bolus followed by 8 mg per hour). On the fourth hospital day, the patient complained of hematemesis and went into hemorrhagic shock that was not responsive to aggressive fluid therapy. Therefore, considering the active re-bleeding from duodenal ulcer, emergency mesenteric angiography was performed to identify and embolize the culprit vessel. Before angiography, abdominal computed tomography was performed to outline the vascular anatomy, revealing an aneurysm emerging from the gastroduodenal artery and another aneurysm in the right distal renal artery. Subsequent selective angiography of the common hepatic artery showed an aneurysm in the mid-portion of the gastroduodenal artery and contrast extravasation into the duodenal lumen before embolization (Fig. 3). After selectively probing the gastroduodenal artery and aneurysm with a microcatheter, embolization was performed at the distal and proximal segments from the aneurysm using two coils (diameter 6 mm, length 14 cm, Nester® Embolization Micro-coil; Cook Medical, Bloomington, IN, USA) (Fig. 4). These consecutive procedures produced immediate cessation of radio-contrast extravasation and there were no immediate complications. Upper endoscopy five days after TAE revealed the active duodenal ulcer with the metallic coil at the ulcer base (Fig. 5A), and 11 days after TAE a follow-up endoscopic view showed the ulcer with regenerating epithelium, and the coil material was not visible (Fig. 5B). There were no stigmata of recent bleeding observed on these sequential endoscopic examinations. During first follow-up endoscopy, gastric mu-
cosal biopsies were performed for rapid urease tests and Giemsa staining for the identification of *Helicobacter pylori*.

The patient received antibiotics therapy for *H. pylori* eradication due to positive results of the Giemsa staining and the intravenous proton pump inhibitor was changed to an oral agent. He was discharged in good condition on the 18th day of hospitalization. The patient remained asymptomatic and showed no recurrent duodenal ulcer bleeding for nine months after the TAE.

**DISCUSSION**

Technical advances in endoscopic hemostasis enable effective gastrointestinal bleeding control in most patients. However, endovascular management and surgery may prove necessary after failed endoscopic management for intractable gastrointestinal bleeding, as in our case. Upper gastrointestinal bleeding (UGIB) can be caused by a number of pathologies. Aneurysms of the gastroduodenal artery are rare causes of UGIB. If UGIB associated with a gastroduodenal artery aneurysm occurs, the massive bleeding can be catastrophic. Therefore, early recognition and prompt treatment are crucial in UGIB patients suspected to have gastroduodenal artery aneurysms. A literature review of visceral aneurysm cases performed by Moore et al. found that 35% of gastroduodenal artery aneurysms are UGIB. If UGIB associated with a gastroduodenal artery aneurysm occurs, the massive bleeding can be catastrophic. Therefore, early recognition and prompt treatment are crucial in UGIB patients suspected to have gastroduodenal artery aneurysms. A literature review of visceral aneurysm cases performed by Moore et al. found that 35% of gastroduodenal artery aneurysms are already ruptured at presentation, with an associated mortality rate of 21%. The most common clinical event associated with ruptured gastroduodenal artery aneurysms is UGIB, which occurs in 50% of patients, with retroperitoneal and intraperitoneal bleeding occurring less frequently.

To the best of our knowledge, this is the first case of duodenal ulcer bleeding resulting from a ruptured gastroduodenal artery aneurysm in a patient with neurofibromatosis type 1. This case suggests that a high level of suspicion for aneurysmal rupture bleeding is needed to select appropriate treatment options for such patients, and that embolization of the involved arteries should be considered an essential treatment over endoscopic hemostasis alone to prevent recurrent bleeding in such cases.

**REFERENCES**