Yonsei Medical Journal
Vol. 38, No. 4, pp. 240~244, 1997

A Case of Bleeding from the Dieulafoy Lesion of the Jejunum

Kwi Soon Lee[1], Yoon Jae Moon[1], Sang In Lee[1], In Suh Park[1], Seung Kook Sohn[2], Jeong-Sik Yu[3], and Jeong Hae Kie[4]

Dieulafoy lesion is an uncommon cause of gastrointestinal bleeding, reported to be only 2% of acute or chronic upper gastrointestinal bleeding episodes. Bleeding occurs from a small mucosal erosion involving an unusually large submucosal artery in an otherwise normal mucosa. It is associated with massive, life threatening hemorrhage and is difficult to diagnosis. In most cases the lesion is encountered in the proximal stomach, antrum, duodenum, colon and rectum. In particular, extragastric Dieulafoy lesion is an extremely rare source of intestinal bleeding. In Korea, no case of bleeding from a Dieulafoy lesion of the small intestine has been previously reported. We experienced one case of bleeding from a jejunal Dieulafoy lesion, which was confirmed by the pathologic examination of the resected specimen, and report here.

Key Words: Dieulafoy lesion, gastrointestinal bleeding

Dieulafoy lesion, sometimes referred to as "caliber-persistent artery" or submucosal arterial malformation, is an uncommon cause of gastrointestinal bleeding, reported to be in only 2% of acute or chronic upper gastrointestinal bleeding episodes (Pointer *et al.* 1988a; Eidus *et al.* 1990; Stark *et al.* 1992). This lesion is characterized by the following: a negative past medical history, independence from peptic ulcer disease, sudden onset, increasing bouts of hematemesis, a tiny mucosal lesion, an open submucosal artery of seemingly large caliber, failure of conservative treatment, and 60.5% overall lethality (Miko and Thomazy. 1988). Bleeding occurs from a small mucosal erosion involving an unusually large submucosal artery in an otherwise normal mucosa. In most cases the lesion is encountered in the proximal stomach within 6 cm of the gastroesophageal junction. Similar lesions have been described in the antrum, duodenum, jejunum, colon and rectum (Matuchansky *et al.* 1985; Vetto *et al.* 1989; Jaspersen *et al.* 1994; Dy *et al.* 1995). In particular, extragastric Dieulafoy lesion is an extremely rare source of intestinal bleeding (Jaspersen *et al.* 1994; Dy *et al.* 1995).

It is associated with massive, life-threatening hemorrhage and may be underdiagnosed as a bleeding source due to the difficulty in diagnosis. In Korea, no case of bleeding from the Dieulafoy lesion of the small intestine has been previously reported. We experienced one case of bleeding from a jejunal Dieulafoy lesion, which was confirmed by the pathologic examination of the resected specimen, and report here.

Received June 16, 1997
Accepted August 23, 1997
Department of Internal Medicine[1], Surgery[2], Radiology[3] and Pathology[4], Yonsei University College of Medicine, Seoul, Korea
Address reprint request to Dr. K.S. Lee, Department of Internal Medicine, Yongdong Severance Hospital, Yonsei University College of Medicine, Yongdong P.O. Box 1217, Seoul 135-270, Korea

CASE REPORT

A healthy 20-year-old woman with no history of

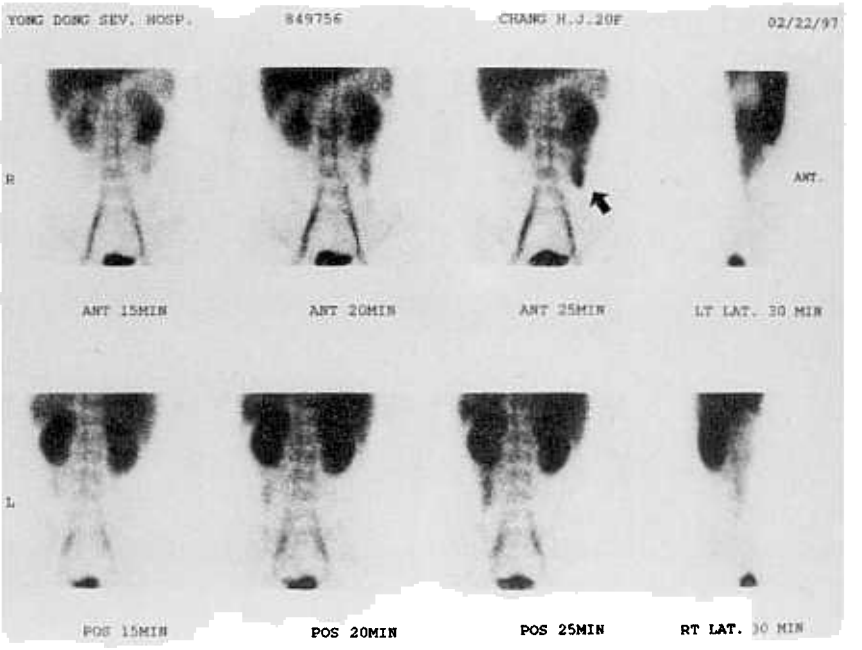

Fig. 1. A technetium-99m-labelled red blood cell scan indicated blood appearing in the jejunal area.

gastric complaints presented with hematochezia. She was not using nonsteroidal anti-inflammatory drugs and had neither a history of smoking nor of excessive alcohol consumption. On admission, her blood pressure was 90/50 mmHg, pulse rate 90/min. On examination, she revealed cool and moist extremities and her conjunctivae were pale. Rectal examination showed hematochezia. The WBC count was 9810/mm^3, hemoglobin 8.1 g/dl, hematocrit 25.0%, platelet 211,000/mm^3. Blood chemistry was normal. Prothrombin time was 12sec (100% of normal) and partial thromboplastin time was 38sec. Initial esophagogastroduodenoscope and colonoscope did not reveal the bleeding sources. A technetium-99m-labelled red blood cell scan indicated bleeding appearing in the jejunum (Fig. 1). Two units of packed red blood cells were infused for hemodynamic stabilization. On the evening of admission, the patient presented a large amount of hematochezia, presyncope and profound orthostatic hypotension. At this time her blood pressure was 60/40 mmHg and pulse rate 130/min. The WBC count was 4500/mm^3, hemoglobin 6.0 g/dl, hematocrit 17.1%, platelet 200,000/

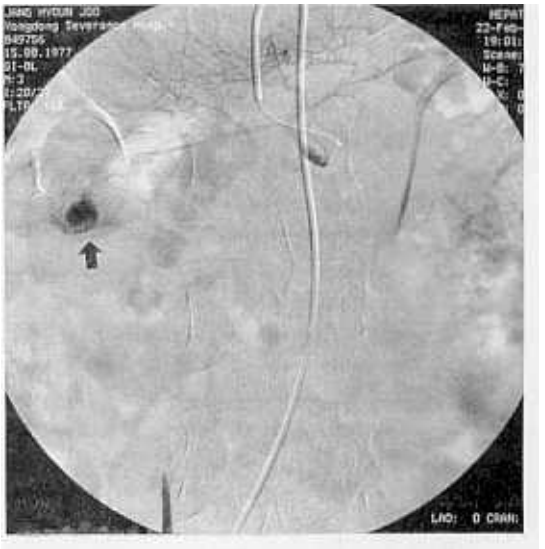

Fig. 2. Angiogram of the superior mesenteric artery revealed active extravasation of contrast media with pseudoaneurysmal dilatation from the third branch of the jejunal artery.

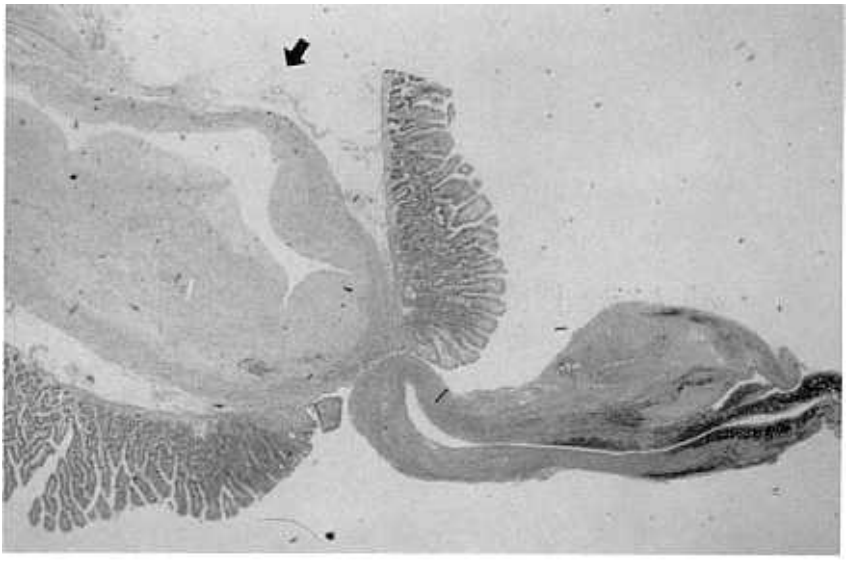

Fig. 3. The lesion consisted of a small mucosal erosion in close contact with a large, tortuous blood vessel in the submucosa, which was filled by a thrombus (H & E stain, X 40).

mm^3. Emergent visceral angiography was performed. A superior mesenteric artery injection revealed active extravasation of contrast media with pseudoaneurysmal dilatation from the third branch of the jejunal artery (Fig. 2). The radiological impression was an unusual type of arteriovenous malformation. We tried to embolize with gelform, but failed. So, to localize the source of bleeding, we left the arterial catheter in the third branch of the jejunal artery and surgical exploration was performed. When jejunum was opened, oozing was noted from a small exposed vessel, which was surrounded by tiny mucosal defect. There was no other macroscopic mucosal abnormality in the remaining mucosa. Histology confirmed the diagnosis of Dieulafoy lesion in the jejunum. The lesion consisted of a small mucosal erosion in close contact with a large, tortuous blood vessel in the submucosa (Fig. 3). The patient did very well postoperatively and had no further bleeding. She was discharged on the nineth postoperative day and reported no further bleeding episodes at a 2-month follow-up visit.

DISCUSSION

The source or etiology of acute massive gastrointestinal hemorrhage can be identified in between 80 and 95% of patients after the introduction of fiberoptic endoscopy. One of the rarer causes of massive bleeding is Dieulafoy lesion (Dieulafoy, 1897~1898). This lesion was first described by Gallard (1884) and has been described under the labels "aneurysm" (Gallard, 1984), "submucosal arterial malformation" (Goldman, 1964), "Dieulafoy's exulceratio simplex" (Dieulafoy, 1897~1898) and "gastric arteriosclerosis" (Antonie, 1961).

The etiology and pathogenesis of Dieulafoy lesion has remained obscure and is still the subject of controversy. One hypothesis is that a submucosal artery elongates and becomes tortuous with aging. The tortuous artery may cause pressure on the overlying mucosa leading to mucosal ischemia, erosion and then arterial rupture. Some reports support that Dieulafoy lesion is a disorder related to aging (Miko and Thomazy, 1988; Lin *et al.* 1989). Another hypothesis is that these large vessels do not erode, but rather develop a weakening in the wall with gradual dilatation, followed by thrombus formation before rupture. This is suggested by the localized subintimal fibrosis of the artery adjacent to the site of perforation as well as the thickening of the wall of the adjacent veins (Juler *et al.* 1984; Edius *et al.*

1990). The other hypothesis is a congenital vascular lesion, ie. the large submucosal artery follows a congenital abnormal course, which is abnormally close to the mucosa and an erosion of the overlying mucosa leads to arterial rupture (Hoffmann *et al.* 1984; Veldhuyzen *et al.* 1986). But this hypothesis has difficulty explaining why the lesion does not become clinically apparent until old age and why Dieulafoy's disease sometimes involves the colon or jejunum.

Some reports have described the factors associated with Dieulafoy lesion as the consumption of large amounts of alcohol and tobacco. It is well known that alcohol abusers are prone to chronic gastritis. Chronic gastritis predisposes to vascular dysplasia which leads to thrombosis and necrosis of the arterial wall before rupture. These findings preclude the diagnosis of "ulceroatio simplex" (Dieulafoy, 1897~1898; Veldhuyzen *et al.* 1986).

Histologically, there are no signs of deep ulceration with penetration of the muscularis mucosa and submucosal muscular arteries at the level of the muscularis mucosa that are abnormally large compared with normal vessels found at this level. Under normal conditions, the gastric arteries reveal a striking reduction in their caliber while crossing the submucosa, but with Dieulafoy lesion, the arteries essentially retain their caliber while running an oblique course from the true muscularis mucosa. The arterial segment of persistent caliber is often long, and gradually becomes tortuous. Therefore, Dieulafoy lesion is often called "caliber persistent artery" (Miko and Thomazy, 1988). A superficial mucosal defect, or erosion, is present in the mucosa overlying the artery with focal necrosis and rupture of the artery at the base of the erosion. Bleeding occurs from a small mucosal erosion involving an unusually large submucosal artery (Juler *et al.* 1984).

The clinical picture of this disease is described by the following: a negative past medical history, independence from peptic ulcer disease, sudden onset, usually with the presenting symptom in combination with the passage of tarry stools and circulatory shock (Miko and Thomazy, 1988; Pointer *et al.* 1988b). The true incidence of Dieulafoy is not known. Veldhuyzen *et al.* reviewed massive hematemesis from Dieulafoy lesion in 101 cases (Veldhuyzen *et al.* 1986). Among these, the youngest patient described was 20-months-old, the eldest was 93 and the median was 54. The lesion were twice as frequent in men as in women. Heavy alcohol intake was implicated as a contributing factor in bleeding.

In the past, the diagnosis of Dieulafoy lesion was very difficult and rarely made preoperatively, but these lesions are being diagnosed more frequently today by emergency endoscope. Repeated endoscopic examination may be necessary because of the intermittent character of bleeding. The endoscopic appearance of Dieulafoy lesion is variable. Sometimes no abnormality is seen on upon meticulous inspection. In most cases, there is a small erosive defect with a tiny visible vessel and adherent clot. The lesion is usually located in the proximal stomach, but extragastric locations have also been described (Jaspersen *et al.* 1994; Dy *et al.* 1995). On rare occasions, diagnosis has been made by angiography, which is effective only during active bleeding episodes (Sherman *et al.* 1972).

Dieulafoy lesion of the small intestine or colon is very rare (Matuchansky *et al.* 1985; Vetto *et al.* 1989). Norman *et al.* reported the incidence of Dieulafoy lesion of the small intestine or colon was 9 of 3,059 patients (0.3%) by endoscopic diagnosis (Norman *et al.* 1995). Among these 9 patients, only one patient presented Dieulafoy lesion from jejunum and the others from colon. The most common presenting symptom was hematochezia and other symptoms included melena, presyncope and fatigue. The endoscopic finding of Dieulafoy lesion in the small intestine and colon is identical to gastric lesion: 1) active arterial spurting or micropulsatile streaming from a minute (less than 3 mm) mucosal defect or through the normal surrounding mucosa; 2) visualization of a protruding vessel with or without active bleeding from a minute mucosal defect or through the normal surrounding mucosa; 3) a fresh, densely adherent clot with a narrow point of attachment to a minute mucosal defect or through the normal surrounding mucosa.

Surgery is the treatment of choice in Dieulafoy lesion. During the operation, careful inspection of the mucosa is necessary in order to find the lesion (Veldhuyzen *et al.* 1986). Selective arterial embolization was introduced as a treatment modality by Sherman *et al.* (1972). but it is rarely used today. Recently, a variety of endoscopic therapeutic modalities have

been reported with success rates ranging from 75-to-98%: epinephrine, ethanol, polidocanol, glucose injection, heater probe coagulation, Nd: Yag laser photocoagulation and endoscopic band ligation (Pointer *et al.* 1988a; Lin *et al.* 1989; Stark *et al.* 1992; Jaspersen *et al.* 1994; Norman *et al.* 1995). So, endoscopic therapy can spare a surgical intervention.

Identification of Dieulafoy lesion is important because it may be associated with massive, life-threatening bleeding. Before the endoscopic era, the prognosis was very poor. Goldman, in his 1964 review, reported a mortality rate of 79%.

In a subset of patients with gastrointestinal bleeding who have no readily identifiable lesions by routine endoscope and colonoscope, small-bowel bleeding must be considered. Especially if the patient presents hematochezia and hemodynamic instability, an aggressive diagnostic approach that includes enteroscopy and angiography should be performed and the clinician must consider the bleeding from a vascular origin in the small bowel. Because the character of Dieulafoy lesion is intermittent bleeding, routine endoscope can be normal. Therefore, repeat endoscope must be performed if bleeding from a Dieulafoy lesion in the gastrointestinal tract is suspected.

In our case, we did not find the bleeding site by routine endoscope and colonoscope. We did not perform the enteroscope, but took emergent angiography during active bleeding, which demonstrated a bleeding lesion on the jejunum. Embolization was tried but failed. Emergent surgical exploration revealed bleeding from a jejunal Dieulafoy lesion, which was confirmed by histological examination.

REFERENCES

Antonie T: Arteriosclerosis of the arteries of the stomach. *Med J Aust* 48: 210, 1961

Dieulafoy G: Exulceratio simplex. *Clin Med Hotel-Dieu Paris* 2: 1 (Abstract), 1897-1898

Dy NM, Gostout CJ, Balm RK: Bleeding from the endoscopic treatment of Dieulafoy lesion of the proximal intestine and colon. *Am J Gastroenterol* 90: 108-111, 1995

Eidus L, Rasuli P, Manion D: Caliber persistent artery of the stomach (Dieulafoy's vascular malformation). *Gastroenterology* 99: 1507-1510, 1990

Gallard MT: Aneurysmes miliarires de l'estomac donnantieu a des hematemeses mortelles. *Bull Mem Soc Med Hop Paris* 1: 84-91, 1884

Goldman RL: Submucosal arterial malformation ("aneurysm") of the stomach with fatal hemorrhage. *Gastroenterology* 46: 589-594, 1964

Hoffmann J, Beck H, Jensen HE: Dieulafoy's lesion. *Surg Gynecol Obstet* 159: 537-540, 1984

Jaspersen D, Korner, T, Schorr W, Brennenstuhl M, Hammar CH: Extragastric Dieulafoy's disease as unusual source of intestinal bleeding. esophageal visible vessel *Dig Dis Sci* 39: 12: 2258-2560, 1994

Juler GL, Labitzke HG, lamb R, Allen R: The pathogenesis of Dieulafoy's gastric erosion. *Am J Gastroenterol* 79: 195-200, 1984

Lin HJ, Lee FY, Tsai YT, Lee SD, Lee CH, Kan WM: Therapeutic endoscopy for Dieulafoy's disease. *J Clin Gastroenterol* 11: 507-510, 1989

Matuchansky C, Babin P, Abadie JC, Payen J, Gostout C, Barbier J: Jejunal bleeding from a solitary large submucosal artery. *Gastroenterology* 88: 1065-1068, 1985

Miko TL, Thomazy VA: Caliber persistent artery of the stomach. *Hum Pathol* 19: 914-921, 1988

Norman MD, Gostout CJ, Balm RK: Bleeding from the endoscopically-identified Dieulafoy lesion of the proximal small intestine and colon. *Am J Gastroenterol* 90: 108-111, 1995

Pointer R, Schwab G, Konigsrainer A, Dietze O: Endoscopic treatment of Dieulafoy's disease. *Gastroenterology* 94: 563-566, 1988a

Pointer R, Schwab G, Konigsrainer A, Dietze O: Dieulafoy's disease. *Occurrence, Diagnosis, Therapy. Endoscopy* 20: 27 (abstract), 1988b

Sherman L, Sheeney S, Satchidanand SK, Neumann PR, Barrios GC, Peer RM: Arteriovenous malformation of the stomach. *Am J Gastroenterol* 72: 160-164, 1972

Stark ME, Gostout CJ, Balm R: Clinical features and endoscopic management of Dieulafoy's disease. *Gastrointest Endosc* 38: 545-550, 1992

Veldhuyzen van Zanten, Bartelsman J, Schipper M, Tytgat G: Recurrent massive hematemesis from Dieulafoy vascular malformation. A review of 101 cases. *Gut* 27: 213-222, 1986

Vetto JT, Richman PS, Kariger K, Passaro E Jr: Crisoid aneurysms of the jejunum. *Arch Surg* 124: 1460-1462, 1989