

# Angiographic findings and endovascular embolization in Dieulafoy disease: a case report and literature review

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## ABSTRACT

Dieulafoy disease is an unusual cause of gastrointestinal (GI) hemorrhage that can be fatal. It arises from an abnormally large eroded submucosal artery commonly located in the proximal stomach. Since this disease first described in 1884, it has been rarely discussed in radiology literature. We describe a case of an elderly patient with multiple medical problems presented with upper and lower GI hemorrhage. Repeated upper GI endoscopies revealed a large fundal clot but otherwise were unremarkable. Angiography showed anomalous short gastric artery originating normally from the splenic artery. The latter was embolized with microcoils and Gelfoam®. Following embolization, the frequency and amount of bleeding decreased although did not totally stop and the patient underwent elective partial gastrectomy. The diagnosis was made histologically. Therefore, Dieulafoy disease can represent a diagnostic and therapeutic challenge. It should be suspected if a patient with GI hemorrhage is found to have extravasation from a dilated tortuous artery that appears normal otherwise with no associated large draining veins. Transcatheter selective arterial embolization has a reported role in the literature as a less invasive therapeutic option compared to surgery.

**Key words:** • Dieulafoy disease • gastrointestinal tract, hemorrhage • gastrointestinal tract, angiography • arteries, therapeutic blockade

**D**ieulafoy lesion is an uncommon etiology of gastrointestinal (GI) hemorrhage recently estimated to represent 4% of the cases of upper GI hemorrhage (1). The bleeding results from an abnormally large eroded submucosal artery commonly located in the proximal stomach (2-4). Endoscopy is the current standard method for diagnosis and treatment (1, 5, 6). Endovascular embolization and surgery are usually reserved for cases with diagnostic or therapeutic difficulties. Angiographic findings and transcatheter embolization of this disease are rarely discussed in radiology literature (7-11).

We describe the angiographic findings and transcatheter selective embolization in an elderly patient with Dieulafoy lesion in the proximal stomach that was not diagnosed neither by endoscopy nor angiography. Our aim is to increase the awareness to this potentially life-threatening entity and to discuss the therapeutic role of transcatheter embolization.

## Case report

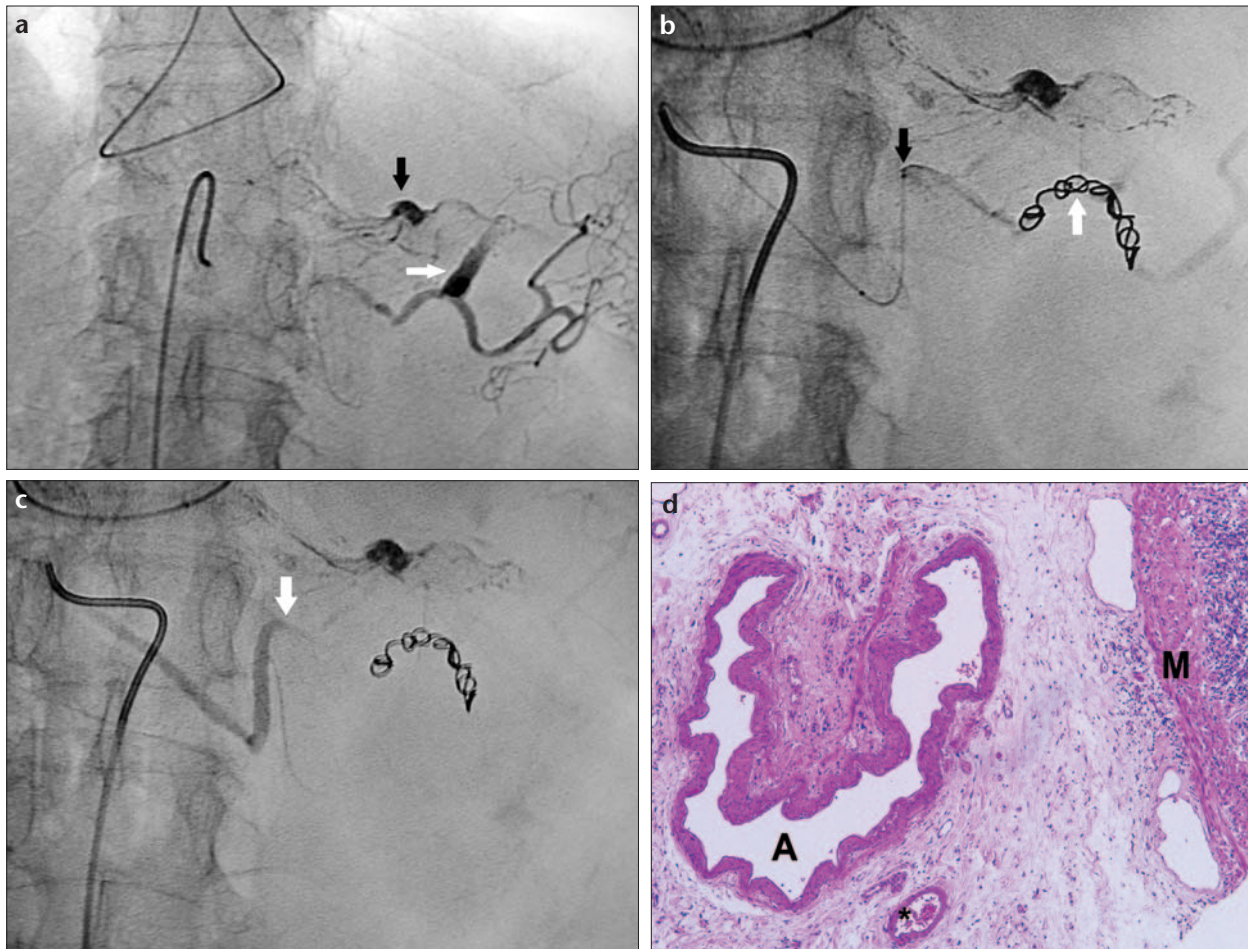
A 77-year-old man known to have diabetes mellitus, parkinsonism, hypothyroidism, sick sinus syndrome and advanced chronic obstructive pulmonary disease; presented with a 12-hour history of vomiting bright red blood. He gave a history of one-month dizziness, easy fatigability and black stools. There was a history of intermittent Aspirin® use for few months prior to presentation but no history of smoking or alcohol use. On examination he was pale with pulse rate of 85 beats per minute and blood pressure of 100/65 mmHg. The abdomen was soft and lax with normal bowel sounds. There were no signs of chronic liver disease, skin vascular malformations or collagen vascular diseases. Rectal examination revealed dark fecal material. His hemoglobin was 68 g/L with normal platelet count and coagulation profile.

The patient received three units of packed red blood cells. Endoscopies at admission and in the next day revealed a large fundal clot but were not able to identify the source of bleeding. He was admitted to the intensive care unit in a hypovolemic state and received blood again. The surgical team was consulted. A third endoscopy was performed with epinephrine injection and heat probe application but the bleeding continued.

Angiography was requested to identify the source of bleeding and for possible embolization. Selective angiography of the celiac artery was performed using a 5-F catheter (SOS Omni Selective, Angiodynamics, NY). It demonstrated an abnormally large short gastric artery arising from the mid splenic artery with free spillage of contrast in the gastric fundus (Figure, a). Subselection of the splenic artery was obtained with a 3-F microcatheter (Excelsior 1018 microcath, Boston Scientific - Target, Fremont, CA). Superselection of the short gastric artery was not

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**Figure a-d.** Pre-embolization selective angiography (a) of the celiac artery shows ectatic short gastric artery (white arrow) originating from the splenic artery. Contrast extravasation (black arrow) from a smaller branch indicating active bleeding is noted in the gastric fundus. Subselective catheterization and embolization of the splenic artery distal and proximal to the origin of the short gastric artery with microcoils (b) (white arrow). The tip of microcatheter (black arrow) is in the proximal splenic artery. Post-embolization angiography (c) shows contrast stagnation (white arrow) in the splenic artery indicating complete occlusion. Histological section from the resected proximal stomach (d) shows a large tortuous submucosal artery (A) below muscularis mucosa (M). A normal-sized submucosal arteriole (\*) is seen at the same level.

feasible as the patient became unstable. Three microcoils (Complex Helical Fibered Platinum Coil-18, 4x30 mm, Boston Scientific - Target Therapeutic, Fremont, CA) were used to embolize the splenic artery distal and proximal to the short gastric artery origin (Figure, b). Gelfoam (Pharmacia Corp, Kalamazoo, MI) was used to establish a complete occlusion as the area of contrast extravasation persisted in the fundal region (Figure, c). The patient received 1 g IV Ceftriaxone just prior to embolization and pneumococcal vaccine afterward.

Postembolization, the amount and frequency of upper GI bleeding decreased but did not stop totally. This allowed time for planning elective partial gastrectomy few days later because an underlying malignancy was

suspected. The diagnosis of Dieulafoy disease was made histologically (Figure, d). There was no further bleeding for the next 6 months.

#### Discussion

Dieulafoy disease was first described by Gallard in 1884 as "miliary aneurysms of the stomach" and characterized in 1898 by Dieulafoy under the name "exulceratio simplex" (12, 13). This abnormality has also been referred to as Dieulafoy's ulcer, cirroid aneurysm, gastric aneurysm, gastric arteriosclerosis, caliber-persistent artery, and submucosal arterial malformation (2, 3). Some of these names are misleading as aneurysms, arteriovenous malformations, and atherosclerosis are not features of this disease (3, 14). The reported incidence ranges

from 0.3 to 6.7% (2, 14). It has been reported in all age groups, but commonly affects patients above 60 years with male predominance (1, 5, 14, 15). The proximal stomach is the most common site, classically in the lesser curvature (14-16). About one third of cases occur outside the stomach with the duodenum being the second most common location followed by the colon (16, 17).

Histologically, the lesion is described as an abnormally large and tortuous submucosal artery that protrudes through a small mucosal defect (3, 18, 19). There is controversy related to the origin of this lesion. While some authors believe it is congenital or anatomical variant, others think it is acquired or age-related (3, 19). The pathogenesis of bleeding in this

disease is not well understood. Mechanical injury to the mucosa by the underlying large pulsating artery may, however, result in a tiny mucosal ulcer which exposes the artery to gastric or bowel contents. The artery, therefore, can be eroded chemically or mechanically and subsequently bleeds (4, 18, 19). Aspirin®; as in our case, non steroidal anti-inflammatory drugs, and alcohol have a reported association (4, 17, 18). Takayasu's arteritis and Behcet's syndrome have been reported in isolated cases (20, 21).

The classical clinical presentation is recurrent, often massive, GI hemorrhage sometimes associated with hypotension. The majority (51%) of patients presented, as in our case, with hematemesis accompanied with melena (14). Comorbid conditions including ischemic heart disease, hypertension, diabetes mellitus, liver disease, and renal failure; are present in up to 90% of patients (6, 17). Initial endoscopy is diagnostic in up to 71% and multiple endoscopies may be required to reach a definite diagnosis (1). Unsuccessful initial endoscopic diagnosis might be attributed to excessive blood (44%) or missed lesion (56%) (17). Endoscopic Doppler ultrasound has been used to show the dilated pulsating submucosal artery and to guide the endoscopic treatment (1, 22).

Dieulafoy disease was first discussed in radiology literature by Durham et al in 1990 (7). The angiographic findings include extravasation of contrast from an eroded artery that may appear normal otherwise (7). However, angiography should be indicative of gastric Dieulafoy disease if it shows tortuous ectatic arteries in the territory of the left gastric artery with no early venous return (3). In our case, there was an ectatic artery in the territory of the short gastric artery with one of its smaller branches showed contrast extravasation. There was no early venous return (Figure, a). Angiographic identification of a tuft of abnormal blood vessels with early venous filling of normal draining veins differentiates angiodysplasia from Dieulafoy lesion (4). In a review of 177 cases of upper GI hemorrhage due to Dieulafoy's lesion; angiography was used in about 8% as an adjunctive diagnostic tool and it was helpful in localizing the bleeding site in 78% (14). In a

series of nine cases angiography was performed in three of them and the findings enabled localization in each case (7). Grace et al. reported angiographic diagnosis in a case of Dieulafoy disease associated with vascular anomaly of the splenic artery (8). A recent case report has described the first case diagnosed by computed tomography angiography (23).

Endoscopy is the current initial therapeutic option (1, 6). The achieved endoscopic permanent hemostasis can be more than 90% (16). Epinephrine injection and electrocautery (heat probe) are probably the most commonly used endotherapies (5).

Selective arteriography with embolization might be the treatment of choice in patients who a) fail endoscopic therapy, b) have acute lower GI bleeding or lesions beyond the reach of therapeutic endoscope, or c) are poor surgical candidates (5). The reported outcome of gastric Dieulafoy's lesion embolization is not constant. In Reilly and Al-Kawas' review (14), three of four patients were treated successfully with gelfoam embolization. Embolization with gelfoam was successful in three patients with upper GI bleeding who appeared angiographically to have true gastric artery aneurysms (24). Successful embolization with polyvinyl alcohol particles was achieved in one out of two Dieulafoy's cases reported by Durham et al (7). However, as in our case, bleeding after embolization may occur and the patients may ultimately require surgery. In a series of 40 cases (5), all three patients who had embolization of Dieulafoy lesions required surgery. Rebleeding from the same lesion could be explained by collateral circulation or non-complete embolization of the feeding artery (3). Endovascular coil embolization of colonic Dieulafoy lesion has been reported with a favourable outcome (10, 11).

The long-term prognosis of Dieulafoy disease is good. The overall recently-estimated mortality rate is 8.6% which is almost the same in the endoscopically and surgically treated patients (1).

Dieulafoy disease can represent a diagnostic and therapeutic challenge. It should be suspected if a patient with GI hemorrhage is found to have extravasation from a dilated tortuous artery that appears normal otherwise with no associated large draining

veins. Even though more studies are required to establish the efficacy of transcatheter selective arterial embolization in Dieulafoy disease particularly in the stomach, there is enough evidence in the literature to justify its use prior to surgery as a less invasive therapeutic option.

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