Dieulafoy Disease of the Colon

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Dieulafoy disease is a rare cause of upper gastrointestinal hemorrhage (usually gastric) and an even rarer cause of colonic bleeding. There is a sudden onset of catastrophic bleeding in a previously healthy individual, which cannot be managed conservatively. Although this entity can be diagnosed and treated by endoscopy and angiography, the knowledge of its existence in colon is critical to making the diagnosis because of its small size. In an era of widespread use of endoscopy, the pathologists uncommonly receive resection specimens for Dieulafoy lesions. However, the diagnosis can be missed because of the small size of the lesion and the almost normal appearance of the mucosa. Awareness of its existence will provide an answer for otherwise unexplained gastrointestinal hemorrhage.

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Dieulafoy disease, first described by Gallard in 1884, and named so, by Dieulafoy, in 1898, is a rare cause of sudden onset, catastrophic upper gastrointestinal bleeding. It is defined as an abnormally large artery that retains a large caliber as it approaches the mucosa. The reported incidence as a cause of upper gastrointestinal hemorrhage varies from 0.3% to 6.7% and 1% to 2% in resection specimens. Although rare, this entity is being reported more frequently because of widespread use of endoscopy. Only 38 cases were reported, to our knowledge, before 1973, but since then, there has been an exponential increase, and more than 200 cases of Dieulafoy disease have been reported.3

Dieulafoy disease is also known as Dieulafoy lesion or ulcer, caliber persistent artery, gastric aneurysm, gastric arteriosclerosis, submucosal arterial malformation, cirsoid aneurysm, and solitary exulceration simplex. The most common location is the lesser curvature of stomach, within 6 cm of the cardia. The reason for the predominance of location at this site is that the submucosal arteries in the lesser curvature are direct branches of the left gastric artery, whereas in the rest of the stomach, the submucosal vessels are of much smaller caliber because of their serial branching. Interestingly, coexistence of the lesion with prior gastric surgery has been noted with increased frequency.4 The cause of the disease is unknown, with some believing it could be congenital, whereas others propose it to be an acquired condition.2,4,5

Of late, there have been increasing reports of extragastric Dieulafoy disease. It has been reported in the lip, esophagus, duodenum, jejunum, ileum, cecum, right transverse, left colon, rectum, anal canal, gall bladder, and bronchus. In fact, one-third of cases in a recent series were extragastric in location.6 Barbier7 reported colonic Dieulafoy disease for the first time in 1985. Since then, 10 cases have been reported in colon, with 9 involving the right colon.1,2,6,7 Thus, left-sided colonic involvement is quite rare.

The disease usually occurs in males, and there is a wide age range at time of occurrence, with reports of cases occurring in infants as well as individuals up to 93 years old.2 The presentation in all ages is the same, with sudden onset of extensive bleeding. Because the bleeding is arterial, the amount of blood lost is large. The bleeding is usually intermittent, and even if it stops spontaneously, the likelihood that the patient will rebleed is high. Depending upon the site of bleeding, the patients present with hematemesis, melena, hematochezia, hemoptysis, or hemodynamic instability only. There is a report of a patient bleeding from a surgically created rectal mucous fistula.10

Although Dieulafoy disease has no direct disease associations, alcohol intake and nonsteroidal antiinflammatory drug use are said to increase the chances of bleeding from a gastric lesion by causing mucosal erosions. In the colon, solid bowel contents lead to stercoral ulceration with resultant exposure of the abnormal artery and hemorrhage.2 There are reports of Dieulafoy disease occurring in patients with classic polyarteritis nodosa11 and abnormal von Willebrand factor.12 Interestingly, there have been 3 recurrences, to our knowledge, of Dieulafoy lesion.11

PATHOGENESIS

In 1962, Voth13 put forward the theory of the caliber persistent artery. Voth postulated that the artery fails to taper as it reaches the mucosa but is otherwise normal. Miko and Thomazy,5 who carried out measurements of the diameter of mucosal and submucosal arteries of patients with Dieulafoy lesion as well as control subjects, support that theory. The mean diameter of pathologic arteries at the level of muscularis mucosae was found to be 1.08 mm (0.39 SD), versus normal arteries having 0.1 mm (0.01 SD) diameter. By serial sectioning, they also demonstrated that there is an accompanying vein, which may also play a role in bleeding.

A Dieulafoy lesion remains asymptomatic unless mucosal erosion exposes the underlying artery, causing it to
bleed. Normally, a network of musculoelastic fibers surrounds and tethers the big branches of arteries entering the submucosa to the muscularis propria. Because the artery reaches close to the muscularis mucosae in Dieulafoy disease, such fibers hold the artery to the muscularis mucosae as well. There is no free, overlying submucosa to allow movement of the mucosa over the submucosa, and the pulsations of the artery cause mucosal damage. Mucosal erosions expose the artery, predisposing it to further injury and bleeding. Also, the artery raises the mucosa overlying it, making it vulnerable to injury.

All these studies have been done on gastric lesions. Because gastric and colonic lesions are identical histologically, the same mechanisms may be extrapolated to play a role in colonic Dieulafoy disease.

**PATHOLOGIC FEATURES**

Macroscopically, dormant colonic Dieulafoy lesion appears like a pseudopolyp at colonoscopy. Characteristically, attempted polypectomy at colonoscopy by electro-snare leads to heavy bleeding. Actively bleeding lesions show arterial spurting, and the caliber of the artery may approach 1 to 2 mm. The mucosal erosion usually measures 3 to 5 mm, and the surrounding mucosa typically appears normal endoscopically.¹⁻⁴

Microscopically, the submucosal artery is seen impinging on, and often eroding through, the muscularis mucosae (Figure 1). Apart from this artery having an abnormally large caliber at this site, there is no obvious abnormality of the arterial wall, and there is no vasculitis. The lesion is usually a single one and always shows an elastic lamina (Figures 2 and 3). Only the overlying mucosa shows erosion, and the surrounding mucosa appears normal histologically. If the artery has bled, an attached thrombus may be seen. No other microscopic abnormalities have been documented, to our knowledge.

**DIFFERENTIAL DIAGNOSIS**

The most important differential diagnosis for Dieulafoy disease is angiodysplasia because both are right sided and present with sudden onset of massive lower gastrointestinal bleeding. On angiography, vascular ectasia and arteriovenous shunting is seen in angiodysplasia, but these features are absent in Dieulafoy disease. Histologically, findings of abnormal submucosal arteries and veins are seen in angiodysplasia, whereas Dieulafoy disease is characterized by mucosal erosion overlying an eroded, large, submucosal, abnormally superficial artery.

Dieulafoy disease should also be separated from other causes of lower gastrointestinal bleeding, like diverticular disease; telangiectasias (which may be associated with Osler-Weber-Rendu syndrome, Turner syndrome, and systemic sclerosis); vascular neoplasms; advanced liver disease causing mucosal spider nevi, which can cause bleeding similar to Dieulafoy disease⁵⁻⁷; and disorders of connective tissues affecting blood vessels. All these conditions can be distinguished from each other on the basis of clin-

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Figure 1. Low-power image, illustrating mucosal erosion overlying a pouting, eroded, large, submucosal, abnormally superficial artery. Note the surrounding mucosa, which is normal in appearance (hematoxylin-eosin, original magnification ×50).

Figure 2. Higher magnification of the superficial artery at the base of the erosion (hematoxylin-eosin, original magnification ×200).

Figure 3. Elastic van Gieson stain at high magnification (original magnification ×300).
Dieulafoy disease of the colon is a rare cause of massive lower gastrointestinal hemorrhage. The treatment is endoscopic or arteriographic, and resection is performed in only 5% of cases when control of bleeding has not been achieved by other first-line modalities. Pathologists need to be aware of the existence of this rare entity as a possible cause of extensive lower gastrointestinal tract bleeding and to differentiate it from other etiologies.

References