Dieulafoy's Lesion

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Introduction
Incidence of acute gastrointestinal bleeding ranges from 50–150 per 100,000 of the population each year, commonly caused by peptic ulcers and esophageal or gastro duodenal erosions in 80% of patients. Obscure gastrointestinal bleeding accounts for up to 5% of all gastrointestinal haemorrhages. Dieulafoy's lesion is one of the causes of obscure gastrointestinal bleeding that can result in treacherous and life-threatening gastrointestinal haemorrhage. 2,3 It is recently estimated to represent 4% of the cases of upper GI hemorrhage4 and is reported in all age groups, but commonly affects patients above 60 years with male predominance.4-7 It is also known as Dieulafoy’s ulcer, cirrhotic aneurysm, gastric aneurysm, gastric arteriosclerosis, caliber-persistent artery, and submucosal arterial malformation.5,8

Dieulafoy’s lesion was first described by Gallard in 1884 as “miliary aneurysms of the stomach” followed by a French surgeon Georges Dieulafoy in 1898 following his study of fatal gastric hemorrhage in three asymptomatic young men. He termed these lesions ‘exulceratio simplex’ since he believed that these lesions were the early stage of peptic ulceration. In over 100 years since Dieulafoy’s original paper, there had been more than 280 reported cases world-wide. 9-14 Dieulafoy disease can represent a diagnostic and therapeutic challenge. We present case report of a 23 years old male who was identified with Dieulafoy’s lesion. 15

Case Report
A 23 year old male presented to emergency department with a self-limiting episode of hematemesis. On inquiry, he reported a similar episode 6 months back which settled without any intervention. He complained of on and off black tarry stools, easy fatigueability, dizziness and palpitations since 1 year. No history of chronic liver disease, peptic ulcer disease, NSAID use, smoking or alcohol abuse was reported. However there is history of multiple transfusions on account of low hemoglobin. Also he underwent an endoscopy which was unremarkable. Past medical and surgical history were unremarkable. On admission there were no significant findings on clinical examination apart from pallor and tachycardia. His laboratory investigations were inconclusive except for hemoglobin which was 4gm/dl. Coagulation parameters were normal. He was resuscitated with intravenous fluids and blood transfusion. Endoscopy was done when patient was stable but it remained unremarkable except for an adherent small clot in stomach fundal area. He was admitted for surveillance and another endoscopy was performed during active bleeding on second day of admission which revealed a bleeding vessel and a minute mucosal defect was unveiled in fundus of stomach. He was admitted for surveillance and another endoscopy was performed during active bleeding on second day of admission which revealed a bleeding vessel and a minute mucosal defect was unveiled in fundus of stomach after hemostasis was achieved with intra-lesional injection of epinephrine. The lesion was identified as Dieulafoy’s lesion. Band ligation was performed. Subsequent recovery was uneventful and no further episodes of bleeding were reported.

Discussion
A normal artery of the GI tract progressively narrows as it transverses the wall of its end organ. 2 Grossly, a Dieulafoy’s lesion consists of a protuberant, tortuous, serpiginous, and abnormally wide artery located in the submucosa of the gastrointestinal tract that approximates a submucosal tumor. 16-18 Histologically, the lesion is described as an abnormally large and tortuous submucosal artery that protrudes through a small mucosal defect 12 varying from 2-5 mm, and has fibrinoid necrosis at its base. 10 Origin of this lesion is controversial since some authors believe it to be congenital or anatomical variant and others think it is acquired or age-related. 12 It was believed to be acquired and aneurysmal in nature, but pathological reports have failed to associate these lesions to aneurysms, atherosclerosis, arteritis or inflammation. There are cases in the literature of newborns being affected, while rare, this could support congenital nature of these lesions. 1

Several mechanisms were proposed to explain the rupture and subsequent massive haemorrhage. One theory suggests that the pulsations in a large submucosal vessel lead to overlying epithelium disruption, causing localized ischemia and exposure to bowel contents resulting in erosion and rupture. 1 Another theory suggests that gastric wear and tear promotes thrombosis within the artery leading to the subsequent necrosis. No evidence was
found to support relationship of NSAIDs or alcohol use causing mucosal injury. Since the pathogenesis is poorly understood, consensus is that there is some form of mucosal erosion or ischaemic injury, possibly related to ageing or cardiovascular disease which weakens an intrinsically vulnerable point and unmask the silent anomaly.¹

Most common site for Dieulafoy's lesion is stomach especially lesser curvature, with 80% to 95% of these lesions being located within 6 cm of the gastro-esophageal junction¹. Duodenum is the commonest location (18%) of extra gastric Dieulafoy's lesions followed by colon (10%) and jejunum (2%) and esophagus (2%).¹⁹ Dieulafoy's lesions have also been described in the small intestine, rectum and anal canal. No history of dyspepsia or peptic ulcer disease in patients with Dieulafoy lesion have been reported²⁰. Classic presentations include haematemesis alone (28%), hematemesis with melena (51%), and melena alone (18%).²¹ Bleeding from Dieulafoy lesion is often self-limited, but life-threatening arterial bleeding occurs in about 10% of cases.

Current standard method for diagnosis and treatment is endoscopy.²² It is effective in diagnosing up to 70% of patients and 92.3% to 96.4% of cases can be identified if endoscopy is performed within the first two hours of bleeding.²² Several endoscopies may be required to establish the diagnosis. Reasons for unsuccessful initial endoscopy were attributed to excessive blood (44%) or the lesion was subtle and overlooked (56%).¹ In the latter situation, aids to endoscopic diagnosis have been suggested. Wright et al. have described provocation of bleeding using intravenous bolus of heparin¹. Endoscopic ultrasound has also been used to aid endoscopic diagnosis.²³ Wireless capsule endoscopy has also been used to localize Dieulafoy's lesion, which has the benefit of being minimally invasive but it does not allow therapeutic intervention. Enteroscopy can be used to identify Dieulafoy's lesion in proximal part of jejunum.¹⁷

Endoscopic criteria for diagnosing Dieulafoy's lesions include any one of the following three abnormalities: (1) active arterial spurting or micropulsatile streaming from a mucosal defect <3 mm or through normal surrounding mucosa; (2) visualization of a protruding vessel with or without bleeding, within a minute mucosal defect or through normal surrounding mucosa; (3) appearance of a fresh, densely adherent clot with a narrow point of attachment to a minute mucosal defect¹. Lesion was missed initially in our patient due to a clot which obscured the view. The correct diagnosis was made when endoscopy was carried out 1 day later during active bleeding. A bleeding vessel was identified and mucosal defect was unveiled during endoscopy hence two criterias for establishing diagnosis of Dieulafoy's lesions were met. An alternative diagnostic tool is angiography. Diagnosis is suggested on demonstration of a tortuous and ectatic artery. If a diagnosis remains uncertain after angiography, portable intraoperative technetium Tc 99m scintigraphy should be considered.²³

Although there is no consensus regarding treatment, endoscopic treatment has evolved over the years as the first-line therapy for Dieulafoy's lesion and is the treatment of choice in easily accessible lesions.²⁵ The reported success rate is in excess of 90%¹. Endoscopic hemostatic procedures can be classified into three groups: (i) thermal therapy – electrocoagulation, heat probe coagulation and argon plasma coagulation; (ii) regional injections – local epinephrine injection and sclerotherapy; and (iii) mechanical therapy– banding and haemoclip.¹ Evidence suggests that endoscopic mechanical hemostatic methods are more effective and successful in achieving hemostasis when compared to injection or thermal treatment methods.¹³ Epinephrine injection and electrocautery (heat probe) are most commonly used endotherapies.⁵ Following endoscopic management, 1 series found no recurrence of bleeding from Dieulafoy lesions over a mean follow-up period of 28 months; another series found no recurrence of bleeding over a mean follow-up period of 36 months.²²,²⁵ Angiography can be used to embolise actively bleeding Dieulafoy's lesions 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 Surgical resection was historically the first-line treatment of Dieulafoy's lesions, usually taking the form of gastrotomy and wide-wedge resection or gastrectomy. Currently it is reserved for 5% of cases that are refractive to endoscopic or angiographic methods.¹ The risk of re-bleeding from Dieulafoy's lesions is reported to be between 9–40% and is higher in endoscopic monotherapy compared with combined endoscopic therapies. Endoscopic methods of hemostasis are the preferred treatment in cases of re-bleeding from lesion. Mortality has been decreased from 80% to 8.6% due to advances in endoscopy.¹ Also improvement in the prognosis might also be explained by the increasing use of endoscopic rather than surgical intervention to control the bleeding.¹³
References


