



## The Gargantuan Ooze-Dieulafoy's Lesion

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### Preface

Dieulafoy's lesion is an exceptional condition engendering massive haemorrhage of the upper gastrointestinal tract on account of vascular anomalies which initiate an acute, fatal condition with inadequate haemostasis. The disorder configures around < 2% of gastrointestinal haemorrhages and is associated with significant morbidity and mortality when appropriate discernment and efficacious treatment or standard endoscopic manoeuvre is delayed.

Dieulafoy's lesion was initially scripted by Dr M.T. Gallard in 1884 who designated the vascular articulations as "military aneurysms of the stomach" [1]. Subsequently, French surgeon Dr Paul Georges Dieulafoy in 1898 nomenclated the lesion as "exulceratio simplex" and precisely characterised the atypical vascular configurations of undetermined aetiology which generated upper gastrointestinal haemorrhage [2].

The condition is additionally denominated as calibre persistent artery, Dieulafoy's disease or Dieulafoy's lesion.

### Disease characteristics

Of indeterminate pathophysiology, Dieulafoy's lesion is categorized as the occurrence of "dilated, tortuous, histologically normal, submucosal arteries". The tortuous arteries predominantly appear within the lesser curvature of stomach although may be discerned within the colon, duodenum or exceptionally within the oesophagus or jejunum [3,4].

As the submucosal gastric vascular articulations directly emerge from right and left gastric arteries, are imbued within the hepatogastric ligament and appear adjacent to lesser curvature, Dieulafoy's lesion is posited to emerge from aforementioned vascular configurations [3,4]. Nevertheless, an estimated one third of

Dieulafoy's lesions are of non-gastric lesions of obscure aetiology and predominantly originate within the duodenum, colon, oesophagus or jejunum. Incriminated arteries may undergo perforation or haemorrhage [3,4].

Dieulafoy's lesion represents an acute process which emerges in diverse body locations and contributes to ~ 5% of gastrointestinal haemorrhages although the true incidence remains obscure.

The disorder demonstrates a predilection for elderly individuals. A male predominance is observed with a male to female proportion of 2:1.

The vascular configurations are typically delineated in the gastric (72%) region although extra-gastric lesions appear within the duodenum (15%), oesophagus (8%), colon (2%) and rectum (2%) [3,4].

An estimated 1% of Dieulafoy's lesion are discovered within the jejunum, a site which is inaccessible with conventional endoscopic measures. Jejunal Dieulafoy's lesion depicts a minimal male predominance with a male to female proportion of nearly 1.2:1 [3,4].

Majority of incriminated subjects appear within seventh or eighth decade. Age of disease occurrence varies from 10 years to 95 years with a median at 82 years. A disease of the developed world, Dieulafoy's lesion incriminates individuals of advanced age with associated chronic diseases or subjects on anticoagulants and nonsteroidal anti-inflammatory drugs (NSAIDs) [3,4].

### Disease pathogenesis

Dieulafoy's lesion is a vascular anomaly which incriminates vascular articulations ten times the magnitude of normal submucosal arteries. Implicated vessels vary from one millimetre to 3

millimetre calibre. Characteristically, a miniature mucosal defect with an absence of inflammation within circumscribing mucosa is observed. Aneurysmal, arteriosclerotic or adjunctive vascular modifications are usually absent. The tortuous, aberrant vessels may bulge through the mucosal defect of around 2 millimetres or 5 millimetres with consequent exposure to mechanical trauma [5,6].

Spontaneous rupture and haemorrhage from Dieulafoy's lesion is posited to arise from consistent pressure which is exerted by pulsatile large-calibre vascular arrangements engendering minute mucosal erosions with subsequent gastrointestinal haemorrhage [5,6].

Additionally, vascular deterioration of submucosal vessels in advancing age may initiate thrombosis, ischemia and hypo-perfusion of encompassing mucosa. Referred to as "vascular steal phenomenon", a pale mucosal halo is observed in incriminated vascular configurations or angiodysplasia and possibly initiates bleeding from Dieulafoy's lesion in subjects exceeding >70 years [5,6].

Jejunal Dieulafoy's lesion may appear as a congenital lesion in new-born babies. Ingestion of alcohol or antiplatelet agents contribute to configuration of Dieulafoy's lesion within the upper gastrointestinal tract [5,6].

Occurrence of chronic disorders, concurrent anticoagulants and non steroidal anti-inflammatory drugs (NSAIDs) contribute to senile mucosal atrophy with possible emergence of Dieulafoy's lesions [5,6].

Aforesaid factors may precipitate haemorrhage from existing lesions. Hypertension, previous gastrointestinal surgical intervention with stress induced vascular injury, contact with intestinal contents or minor trauma may engender a mucosal defect with rupture of aberrant vessels and consequent occurrence of massive haemorrhage [5,6].

Congestive heart failure with fluid overload state may generate Dieulafoy's lesion within enlarged, dilated, tortuous, malformed vascular articulations. Also, hypertrophic obstructive cardiomyopathy, endocarditis, pericarditis, congenital heart disease, abdominal aortic aneurysm, arteriovenous malformation, hepatitis, polycystic ovarian syndrome, dysfunctional uterine bleeding, osteoarthritis, obstructive sleep apnoea, tuberculosis and human immunodeficiency syndrome (HIV) may engender the vascular Dieulafoy's lesion [5,6].

Alcoholism, drug abuse, diverticulosis, asthma, diabetes mellitus, obesity, chronic renal disease, ischemic heart disease, rheu-

matic fever, gastrointestinal angiodysplasia and hyperlipidemia are infrequently associated comorbid conditions [5,6].

Pre-existing gastrointestinal conditions as the peptic ulcer, gastritis, gastroesophageal reflux disease (GERD) or mucosa-associated lymphoid tissue lymphoma (MALT) may be associated with Dieulafoy's lesion [5,6].

### Clinical elucidation

Typically, an acute onset with voluminous hematemesis or melena is delineated. Haemorrhage from Dieulafoy's lesion is unaccompanied by previous retching or vomiting. Clinically, a painless, intermittent, massive gastrointestinal haemorrhage is observed [5,6].

Dieulafoy's lesion is comprised of vascular articulations of significant calibre with uniform patency and unremarkable morphology. Adjoining gastrointestinal vascular articulations appear narrow as vessels traverse the wall of recipient organ. Enlarged submucosal vessels with significant diameter may erode the superimposed mucosa due to vigorous pulsation. With deterioration of superficial mucosa, submucosal vascular articulations are exposed, traumatized and may haemorrhage in the absence of preceding symptoms [7,8].

Antecedent, nonspecific clinical symptoms such as abdominal discomfort can be encountered [7,8].

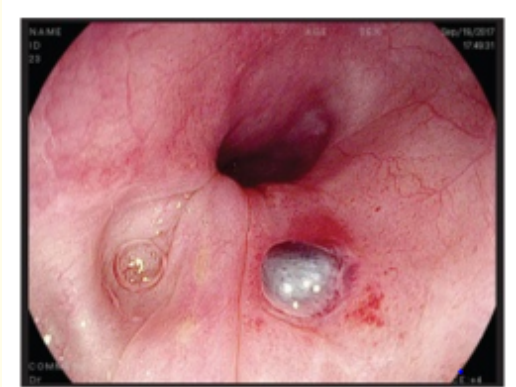
Classically, Dieulafoy's lesion of the jejunum depicts painless gastrointestinal haemorrhage, melena, haematochezia or gastrointestinal bleeding of obscure origin. Haemorrhage may be severe and intermittent due to enlarged diameter of aberrant, pulsatile submucosal vessels. The lesion may be discovered incidentally or may engender intestinal intussusception [7,8].

### Histological elucidation

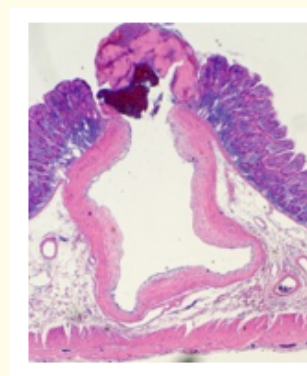
On microscopic examination, enlarged, tortuous vascular articulations are accompanied by miniature discontinuity of the superimposed mucosa. The vessel wall may be incorporated with amyloid [7,8].

### Investigative assay

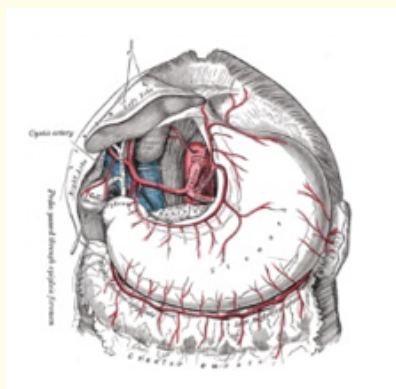
Site, extent and duration of haemorrhage require evaluation. Generally, endoscopy is beneficial for appropriate disease discernment and therapeutic purposes. Cogent angiography can be adopted in instances where endoscopy is inconclusive. Diagnosis of Dieulafoy's disease may be challenging to achieve with intermittent haemorrhage and sites inaccessible upon conventional endoscopy.



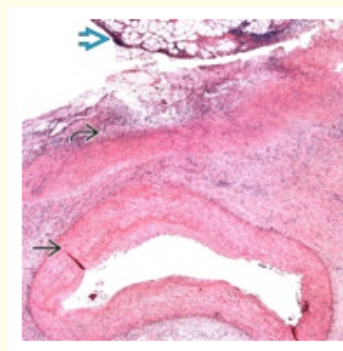
**Figure 1:** Dieulafoy's lesion of the oesophagus depicting an enlarged, tortuous lesion confined to the submucosa [11].



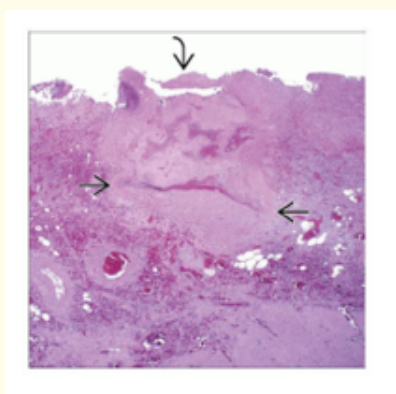
**Figure 4:** Dieulafoy's lesion delineating enlarged, dilated, pulsatile vascular arrangements within the submucosa [14].



**Figure 2:** Dieulafoy's lesion exhibiting tortuous, pulsatile arterial vascular configurations with dilatation [12].



**Figure 5:** Dieulafoy's lesion exhibiting a singular, dilated, pulsatile vascular channel confined to the submucosa [15].



**Figure 3:** Dieulafoy's lesion demonstrating enlarged vascular articulations with haemorrhage confined to the submucosa [13].

Endoscopy of narrow intestinal lumen, discernment of mucosal anomalies, inflammation, vascular clotting or imprecise clinical features can obscure adequate discernment. Therefore, conventional endoscopy may not be efficacious [9,10].

Capsule endoscopy or push enteroscopy are associated with variable efficacies. Mesenteric angiography is beneficial in undiagnosed instances [9,10].

Computed tomography angiography can suitably discern extra-gastric vascular articulations. Heparin therapy with angiography is adopted where failure of standard angiography is encountered. Disease discernment and therapy is significantly ameliorated with non-operative, endoscopic manipulation. Contemporary mortality

rate of haemorrhage encountered with Dieulafoy's lesion is around 8.6% [9,10].

Surgical evaluation of the lesion is recommended with emergence of massive haemorrhage. Intraoperative single or double balloon enteroscopy is safe and beneficial in localizing haemorrhaging blood vessels and may be utilized to complement procedures like capsule endoscopy for cogent diagnostic evaluation and therapeutic interventions [9,10].

### Therapeutic options

Guidelines for managing haemorrhaging Dieulafoy's lesion remain undefined. Endoscopic therapy, direct surgical intervention and angiographic embolization may be advantageous. Although employment of optimal endoscopic monotherapy lacks consensus, endoscopic techniques combined with manoeuvres such as injection sclerotherapy, heater probe, electrocoagulation and mechanical methodologies are adopted [9,10].

Non contact thermo-coagulation is preferable as contact techniques engender transmural injury within thin-walled gastrointestinal segments such as the small bowel. Singular adoption of endoscopic haemo-clipping is associated with significant primary haemostasis and minimal repeat haemorrhage. Endoscopic band ligation is recommended for accessing difficult gastrointestinal sites [9,10].

Surgical intervention is indicated where failure of adjunctive diagnostic modalities is encountered. Intraoperative visualization of intestinal lesions may be challenging on account of accompanying inflammation, polyposis, pertinent mucosal alterations or lack of significant fibrosis thereby necessitating preoperative assessment of lesions. Adequate preoperative evaluation may be achieved with the employment of intraoperative enteroscopy with methylene blue dye or Technetium 99m labelled red blood corpuscles [9,10].

Minimally invasive approach is recommended besides the frequently employed segmental resection with end-to-end anastomosis. Surgical intervention accomplishes a comprehensive (100%) vascular haemostasis [9,10].

Reoccurrence within Dieulafoy's lesion is non concurrent to age, gender or site of lesion within the incriminated subject although appears pertinent to associated comorbidities, infection or adrenaline monotherapy. Repetitive haemorrhage may occur within hours to weeks following initiation of treatment. Thus, monitoring for minimally six months is recommended. Surveillance endoscopy may be optimally adopted [9,10].

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11. Image 1 Courtesy: Practical gastroenterology.
12. Image 2 Courtesy: Wikipedia.com.
13. Image 3 Courtesy: Basic Medical Key.

14. Image 4 Courtesy: Journal of Paediatric Surgery.

15. Image 5 Courtesy: Science Direct.

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