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CASE REPORT Open Access

Ileal Dieulafoy Lesion: a rare case report

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Abstract

Background: Dieulafoy lesion, also known as calibre persistent artery is a rare cause of massive GI bleeding. It's an abnormal sub-mucosal artery protruding from a minute mucosal defect (≤3 mm). Commonest location is in proximal stomach while occurrence in small intestine, especially ileum is extremely rare.

Case presentation: A 26 year old female presented with lower gastrointestinal bleeding. Preliminary investigations failed to locate the exact source of bleed. At laparotomy, an ulcerated nodular lesion, approximately 0.8 cm in diameter was identified in distal ileum. Histology revealed it to be Dieulafoy lesion.

Conclusion: Although uncommon, ileal dieulafoy is one of the causes of obscure gastrointestinal bleeding that could result in treacherous and life-threatening gastrointestinal haemorrhage. Hence, it should be considered in the differential diagnosis of active GI bleeding. The definitive diagnosis is based only on histopathology.

Keywords: Ileum, Dieulafoy, Bleeding, Gastrointestinal

Background

Dieulafoy lesion (DL), also called calibre persistent artery/submucosal arterial malformation or solitary exulceration simplex, is a rare cause of massive GI bleed (0.3–6%) (Christopher Ma & Edwin, 2014). It's an abnormal sub-mucosal artery protruding from a minute mucosal defect (≤3 mm) with normal surrounding mucosa or active arterial bleeding without an ulcer base (Gustavo, 2008). DLs are located in proximal stomach in more than three-quarters of affected patients and occurrence in intestine is extremely rare (Christopher Ma & Edwin, 2014; Gustavo, 2008; Shibutani et al., 2011). Herewith, we report a rare case of lower GI bleed secondary to an ileal DL in a young female.

Case history

A 26 year female presented to emergency department with complaints of bleeding per rectum since 3 days. There were five episodes of passage of moderate volume bright red blood per rectum. There was no associated nausea, vomiting, diarrhea, abdominal pain, loss of weight or appetite. She had no past history of GI hemorrhage prior to presentation. There was no previous significant medical history or any drug intake such as non-steroidal anti-inflammatory drugs or alcohol.

On admission, blood pressure was 110/70 mmHg, and pulse was 92/min. Per-abdominal examination revealed soft, non-tender abdomen with no guarding, rigidity or organomegaly. Investigations revealed hemoglobin of 6.4 g/dl. All the other haematological and biochemical parameters were within normal limits. Supportive treatment with intravenous crystalloids and multiple units of packed red cell transfusion was given, however post-transfusion haemoglobin values were constantly below 6 g/dl.

Colonoscopy revealed large blood clots within the bowel, with no obvious bleeding source. (Figure 1a, b) Esophagogastroduodenoscopy was normal. Ultrasound abdomen and CT angiography showed no abnormality. RBC nuclear scan demonstrated active bleeding from the distal small intestine. Exploratory laparotomy was done and intra-operatively, distal ileum was cut along the anti-mesenteric border to localize the bleeding source. An actively bleeding haemorrhagic lesion was noted in the distal ileum, 70 cm (cms) from the ileocaecal junction. Resection of this segment was done and sent for histopathological examination.

On gross examination, an ileal segment of 8 cm length was received. It revealed a submucosal nodule measuring 0.8 cm in diameter on the anti-mesenteric border. Mucosa over the nodule appeared denuded and haemorrhagic. Cut surface showed presence of a blood clot. The serosa and rest of the ileum were unremarkable.

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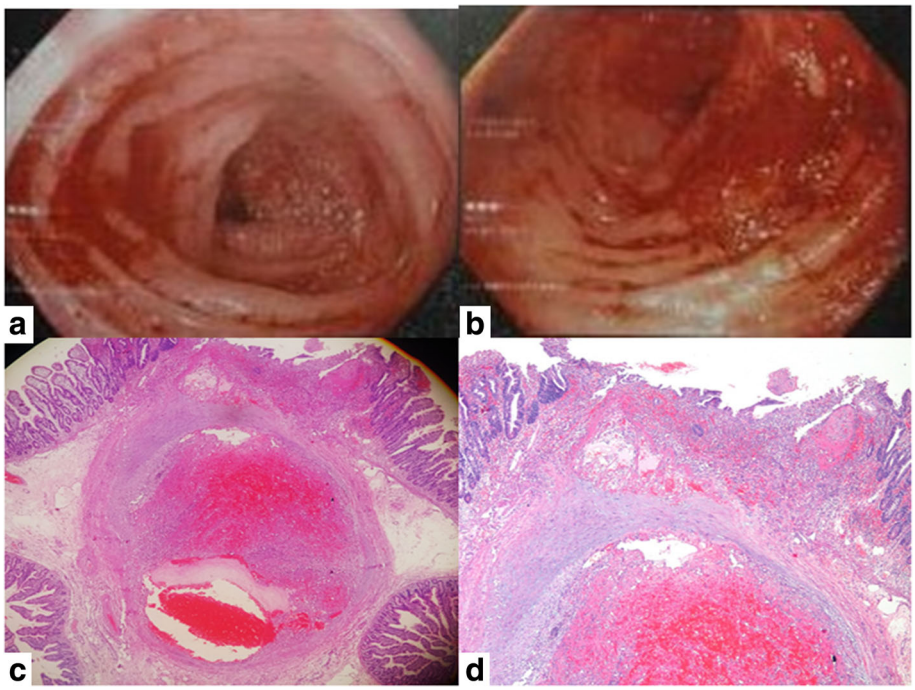

Fig. 1 **a**, **b** Ileo-colonoscopy shows presence of blood smeared entire colon and terminal ileum without any active bleeding or oozing lesion. **c**: Photomicrograph showing a dilated submucosal vessel with organising thrombus in lumen; H&E, 4×. **d**: Overlying mucosal ulceration; H&E, 40×

On microscopy, sections from the nodule revealed a large calibre artery in the sub-mucosa eroding the overlying mucosa (Fig. 1c, d). The lumen of the artery was obliterated by an organising thrombus (Fig. 1c). The arterial wall was unremarkable. A diagnosis of Dieulafoy lesion of ileum was rendered.

The patient had rapid and uncomplicated recovery with no further bleeds up to 6th months of follow up.

Discussion

Originally described by Gallard in 1884 as 'miliary aneurysms of the stomach', DL was distinguished by the French surgeon, Georges Dieulafoy in 1898 following his study of fatal gastric haemorrhage in three asymptomatic young men. He termed these lesions 'exulceratio simplex' based on his belief that these lesions were the early stage of peptic ulceration (Baxter & Aly, 2010). Although the pathogenesis is unclear, some authors consider the lesion to be congenital while others consider it to be acquired in origin (Jain & Chetty, 2009).

The true incidence is unknown. DL typically presents as acute, painless, massive, often recurrent haemorrhage, most commonly as malena followed by haematemesis, hematochezia, iron deficiency anemia or a combination of these features (Baxter & Aly, 2010). Our patient had hematochezia due to ileal lesion. It is clinically difficult to distinguish from other causes of lower gastrointestinal bleeding such as arterio-venous malformations or diverticular hemorrhage (Christopher Ma & Edwin, 2014). DL commonly presents in elderly (median age: 54 years), however it can occur in any age group. The male to female ratio is 2:1 (Baxter & Aly, 2010). Association with cardiovascular diseases, diabetes, chronic renal failure and hypertension or drug intake (anticoagulants/ antiplatelets) has been noted (Gustavo, 2008). However, no such condition was present in our patient.

Several mechanisms have been proposed for rupture and subsequent massive haemorrhage. Mechanical factors promote arterial thrombosis leading to subsequent necrosis. Bowel contents contribute to mucosal stercoral ulceration over an abnormally dilated submucosal vessel. Age-related mucosal atrophy and NSAIDs or alcohol induced mucosal injury can contribute. Ageing or cardiovascular disease weaken an intrinsically vulnerable point and unmask the silent anomaly (Baxter & Aly, 2010).

Typical location is in the proximal stomach (75–95%), commonest site being within 6 cm of the gastro-oesophageal junction on the lesser curvature. This is followed by duodenum (18%), colon (10%), jejunum and esophagus (2% each) (Fox et al., 2001). They also have been found in the ano-rectum and bronchus. DL in the ileum are extraordinarily rare (< 1% of all DLs) (Baxter & Aly, 2010). To the best of our knowledge, less than 10 cases of histologically proven ileal DL in an adult have been reported so far (Shibutani et al., 2011; Fox et al., 2001; Choi, 2012)

DL has a dilated aberrant sub-mucosal vessel that does not undergo normal distal branching or tapering, and maintains a constant diameter of 1–5 mm (about 10 times that of mucosal capillaries). It runs a tortuous course within the submucosa and protrudes through a 2–5 mm mucosal defect with fibrinoid necrosis at its base (Baxter & Aly, 2010). Vascular lesions of small bowel have been classified into six groups (Table 1) (Baxter & Aly, 2010; Choi, 2012). According to this classification, our case was type 2b.

Association with aneurysms, atherosclerosis, arteritis or inflammation is generally absent (Christopher Ma & Edwin, 2014). The diagnosis of DL is based upon classical histological features which are usually unavailable, as recently most vascular lesions have been treated by nonsurgical modalities (Choi, 2012).

Rapid advances in endoscopic technology have increased the opportunity to precisely diagnose DL. Initial endoscopy is effective in 70% of patients, however several endoscopies (≥3) have been required in 6% (Joarder et al., 2014). DLs are challenging to diagnose as they are subtle and can be overlooked on endoscopy (Nojkov & Cappell, 2015). Moreover, an ileal DL is inaccessible to conventional endoscopy (Gustavo, 2008). CT angiography and red blood cell (RBC) scintigraphy also aid in diagnosis, as in our case (Christopher Ma & Edwin, 2014). Technetium-99 m labelled RBC scans can identify the location of bleeding DL when not detected by endoscopy. It is advantageous as the threshold for detecting extravasation in gut is only 20% of that required by angiography (Baxter & Aly, 2010; Joarder et al., 2014).

There is no definite protocol for treatment of DLs is available; management depends on the presentation, site of lesion and available expertise. Therapeutic endoscopy remains the first line of treatment for controlling bleeding while angiography is considered as a valuable alternative (Nojkov & Cappell, 2015). Due to its low incidence, there are only few articles describing the role of endoscopic treatment. It is suggested by some authors that surgical resection is still preferable, especially in lesions exposed to hard stools, increasing their likelihood of re-bleeding (Baxter & Aly, 2010). The mortality rate has declined dramatically from 80 to 8.6% with diagnostic and therapeutic advancement (Choi, 2012).

Table 1 Types of vascular lesions of small bowel

Types	Features
1a	Punctulate erythema (< 1 mm), regardless of oozing
1b	Patchy erythema (a few mm), regardless of oozing
2a-DL	Punctulate lesions (< 1 mm) with pulsatile bleeding
2b-DL	Pulsatile red protrusion without surrounding venous dilatation
3-AVM	Pulsatile red protrusion with surrounding venous dilatation
4	Unclassified lesion

Conclusion

Although rare, ileal DL can cause life-threatening hemorrhage and should be considered in differential diagnosis of GI bleeding. An aggressive multidisciplinary approach including radiologic intervention, enteroscopy and surgery is required for proper diagnosis and treatment.

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Authors' contributions
AJa carried out concepts and design, literature search, participated in clinical study, data acquisition, data analysis and manuscript preparation will stand as guarantor also. MK carried out concepts and design, literature search, manuscript review. AJo participated in clinical study, data acquisition and manuscript review. AR carried out literature search, clinical study and data acquisition. All the authors read and approved the final manuscript.

Ethics approval and consent to participate
Available with author. Written informed consent to participate was obtained.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing interests
The authors declare that they have no competing interests.

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