Recurrent colonic Dieulafoy's lesion associated with bizarre vascular malformations and abnormal Von Willebrand factor

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Abstract

Colonic Dieulafoy's lesion is an unusual source of massive lower gastrointestinal haemorrhage. It is characterized by severe bleeding from a minute submucosal arteriole that bleeds through a punctuate erosion in an otherwise normal mucosa. We describe an elderly man who presented recurrent rectal bleeding from a Dieulafoy's lesion in the sigmoid colon associated with bizarre colonic vascular malformations and an abnormal von Willebrand Factor. He was successfully treated by endoclips application. The clinicopathologic features of this unusual association are discussed and suggestions are made for diagnosis and management (Acta gastroenterol. belg., **2005**, 68, **443-445**).

Key words : Colonic Dieulafoy's lesion, bizarre vascular malformations, von Willebrand Factor.

Introduction

A Dieulafoy's lesion (DL) is an uncommon cause of gastrointestinal bleeding, although some authors consider it to be an under-recognized entity (1,2) . In all of the published series, the proximal stomach is by far the most common site for DL (2). The duodenum is the second most common site for DL, while 26 cases have been reported in segments of the small intestine distal to the ligament of Treitz, mostly in the jejunum (3,4). Oesophageal DLs have rarely been reported with the distal oesophagus being the most common site where they may be an alternative source of bleeding, despite the presence of varices (5). Colonic DLs are also uncommon. They are most frequently found in the proximal colon[1] and rectum (6) and rarely in the anal canal (7).

We report an elderly man who presented recurrent rectal bleeding from a Dieulafoy's lesion in the sigmoid colon associated with bizarre colonic vascular malformations and an abnormal von Willebrand factor (vWF). He was successfully treated by endoclips application. To our knowledge this is the first report of an association of recurrent colonic DL, bizarre vascular malformations and an abnormal vWF.

Case Report

A 71-year-old man presented with a 24-hour history of painless haematochezia. There was no history of GI haemorrhage, peptic ulcer disease, use of non steroidal anti-inflammatory drugs, alcohol abuse or chronic liver disease. No significant orthostatic changes were noted to the patient's pulse rate or blood pressure.

Digital rectal examination revealed bright red blood and no mass or anal fissure. Clinical examination revealed a systolic murmur on the heart's apex, and echocardiography showed a moderate aortic valve stenosis. Laboratory values were normal, apart from a low haemoglobin level (10.5g/L ; normal 12-16 g/L). Emergency colonoscopy showed residues of fresh blood in the rectum, sigmoid and descending colon, and oozing of blood from a minute mucosal defect without surrounding ulceration in the sigmoid colon. The lesion was treated by application of two clips and the bleeding stopped with a subsequent stabilization of the patient's haemoglobin. Post-procedure recovery was uneventful and the patient was discharged four days later.

He was readmitted two months later complaining about mild rectal bleeding. His haematocrit was normal. A colonoscopy was performed, after full bowel preparation, which demonstrated a lesion with active bleeding in the sigmoid colon (Fig. 1), having similar features and in the same region with the one previously described two months ago. We also observed bizarre vascular malformations in close proximity to the bleeding lesion (Fig. 2). Endoscopic haemostasis was achieved by using a clipping device (HX-5QR-1 ; Olympus, Tokyo, Japan) to apply 3 normal-sized clips (HX-600-135 ; Olympus, Tokyo, Japan) (Fig. 3). Subsequently, the bleeding stopped. Considering the recurrence of DL as an unusual phenomenon, associated to bizarre vascular malformations and aortic stenosis, standard haemostasis tests (prothrombin time, fibrinogen level, activated partial thromboplastin time, factor VIII activity, von Willebrand factor antigen (vWFAg) levels and von Willebrand factor ristocetin cofactor activity (vWFRCo)) were measured and were normal. The multicentre pattern and the proportion of high-molecular-weight multimers of vWF were also measured. A rate of 8% of high-molecular-weight multimers of vWF was found (normal 23-27% of

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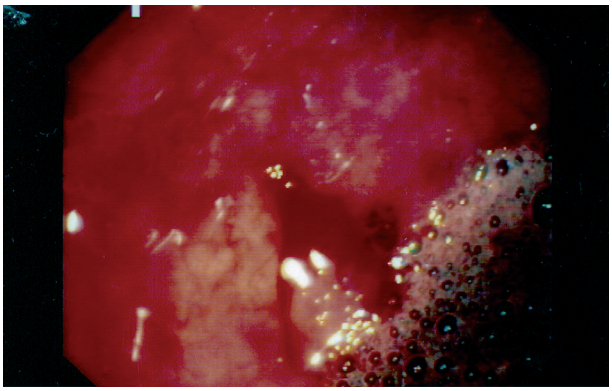

Fig. 1. — Endoscopic view of acute bleeding from Dieulafoy's lesion.

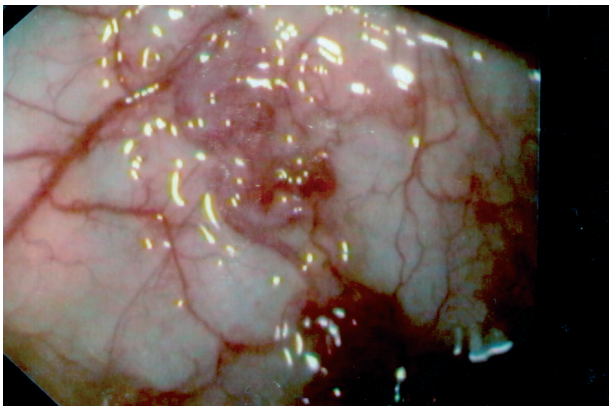

Fig. 2. — Endoscopic view showing excessively tortuous, protuberant and dilated vessels.

total multimers), indicating deficiency in haemostasis. We suggested operation for aortic stenosis but he refused.

Thereafter, he has remained stable without evidence of GI haemorrhage during a follow – up period of six months.

Discussion

Despite the fact that DL has been reported rarely in neonates and young infants (8) considering it to be a congenital lesion, in most published series there is a preponderance of men of advanced age (9). Consequently, most authors now believe that DL is acquired in origin and related to old age. Normally, when muscular arteries penetrate the bowel wall there is gradual tapering as the arterioles ramify and transverse successive layers. In DL, the natural tapering fails to occur and an abnormally large muscular arteriole reaches the submucosa (10-11). A variety of local factors may then cause mucosal ulceration and rupture of the submucosal arteriole. Continued pulsatile arterial compression has been implicated as a cause of localized ischemia of the overlying mucosa (12). The ischemic mucosa erodes, exposing the

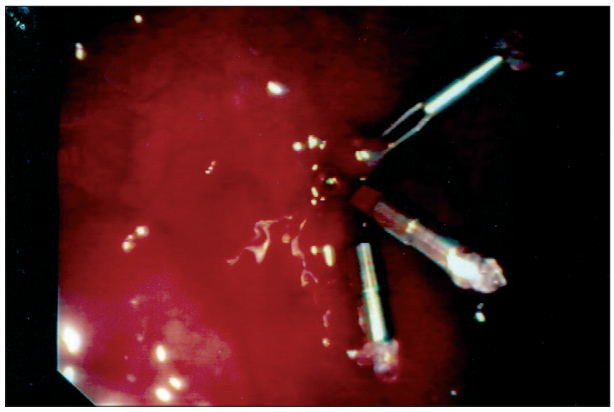

Fig. 3. — The bleeding stopped after the application of three clips.

underlying arteriole to luminal contents which may eventually lead to erosion and rupture of the arteriole. Age-related mucosal atrophy and medial degeneration may contribute to the process. The final rupture of the vessel results from the compound effects of the vascular ectasia, mucosal atrophy, and possibly ischemic injury related to aging and cardiovascular diseases, which weaken an inherently vulnerable point (10). In the colon, impacted faeces may contribute to mucosal stercoral ulceration over an abnormally dilated submucosal arteriole. This condition is especially true in the anal canal, where a scybalous mass may erode into the arteriole.

The case presented here is unusual in several aspects. The site of DL in the sigmoid colon is uncommon, as DLs are most frequently found in the proximal colon and rectum. The bleeding had arisen from a lesion with the typical features of DL like micropulsatile streaming of blood from a minute (< 3mm) mucosal defect with normal surrounding mucosa, in close proximity to the bizarre vascular malformations. The bleeding recurred, despite initially successful endoscopic treatment, which is unusual for DLs. Also, measurement of several factors of haemostasis revealed a variable selective loss of the largest multimeric forms of vWF, a finding described in patients with bleeding gastrointestinal angiodysplasias and aortic stenosis.

The term "bizarre vascular malformations" is used for excessively tortuous protuberant dilated vascular structures that are occasionally encountered in the colon and may be responsible for chronic blood loss (13) ; they are difficult to classify at present, but we believe they represent an early stage before the full development of angiodysplasias. Therefore, the existence of bizarre vascular malformations and DL in close proximity in our patient, suggests that both are age related phenomena in the blood vessels of the colonic wall.

The finding of an abnormal vWF in our patient is an acquired trait related to his aortic stenosis, as inherited von Willebrand disease would have been symptomatic since early childhood. Veyradier *et al*. (14) reported that

a variable deficiency of the largest multimers of vWF is likely to play a key rote in the bleeding expression of GI angiodysplasias and they suggested that patients with bleeding angiodysplasias should be screened for vWF abnormalities, including a study of vWF multimers and testing for the most common disorders associated with acquired von Willebrand disease, especially aortic stenosis in elderly patients.

The confirmation of our findings in further studies would be of great importance because the correction of aortic stenosis in patients with acquired abnormalities of vWF and bleeding angiodysplasias has resulted in normalization of vWF and reduction of blood loss. Thereafter, in patients with Dieulafoy's lesion, abnormal vWF and aortic stenosis, the treatment of the acquired haemostatic defect, via operation of aortic stenosis, could contribute to the avoidance of recurrent bleeding.

In summary, further awareness of this unusual disease is required, which should include its relation with the idiopathic vascular disorders involving the GI tract and an abnormal vWF associated to aortic stenosis.

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