Heyde’s syndrome: diagnosis and management by the novel single-balloon enteroscopy

We present a case of obscure gastrointestinal bleeding due to small bowel angiodysplasia in a 68-year-old woman with severe aortic valve stenosis. The diagnosis was confirmed and subsequently managed using single-balloon enteroscopy.

Introduction

The association between bleeding gastrointestinal angiodysplasia in patients with aortic stenosis (Heyde’s syndrome) has been well recognised. Nevertheless, the diagnosis and treatment of small bowel angiodysplasia remains difficult, as these regions are often beyond the reach of conventional endoscopy. With the advent of capsule endoscopy and complimentary investigation with balloon-assisted enteroscopy, small bowels are no longer the ‘dark continent’ of endoscopy. To the best of our knowledge, this is the first report on the use of single-balloon enteroscopy (SBE) system for managing Heyde’s syndrome.

Case report

A 68-year-old woman presented with 1-year history of iron deficiency anaemia of unknown origin in December 2007. She had undergone mitral valvular replacement for chronic rheumatic heart disease 25 years earlier and was put on warfarin anticoagulation. Over the previous 10 years, she had concomitant aortic valve stenosis of mild-to-moderate severity. In 2006, it was noted that the degree of her aortic valve stenosis had progressed. A trans-oesophageal echocardiogram revealed severe aortic stenosis and the calculated aortic area (using the continuity principle) was around 0.50 cm². Towards the end of 2006, she developed exertional dyspnoea, decreased exercise tolerance, and the intermittent passage of tarry stools. There was no history of aspirin or non-steroidal anti-inflammatory drug intake. A stool sample was positive for occult blood. Laboratory results confirmed she was iron deficient with a haemoglobin level of 71 g/L (reference range, 117-149 g/L) and an elevated reticulocyte count of 4.1% (0.5-2.0%). Repeated upper gastrointestinal endoscopy and colonoscopy examinations failed to identify the bleeding source. Regular blood transfusions and iron supplements were required to control her anaemic symptoms. A wireless video capsule endoscopy examination was performed and this demonstrated fresh blood in the proximal part of the jejunum. Blood clots and coffee-grounds fluid were found throughout the small bowel but the underlying cause of the bleeding was not identified. An enteroscope was inserted to around 80 cm distal to the ligament of Treitz. The length of small bowel examined was measured according to the method described by May et al.² The enteroscope was inserted to around 80 cm distal to the ligament of Treitz. The length of small bowel entered was measured according to the method described by May et al.³ Upon withdrawal of the enteroscope, a bleeding proximal jejunal angiodysplasia was identified (Fig 1). Endoscopic injection with epinephrine solution was performed to arrest the haemorrhage and then the lesion (Fig 2) was ablated using argon plasma coagulation. No complications were noted after the procedure. Her haemoglobin level eventually stabilised at around 110 g/L and she required no further transfusions.

Discussion

The association of gastrointestinal bleeding from angiodysplasia with aortic valve stenosis is known as Heyde’s syndrome.¹ It was first reported in 1958, and since then many reports
本文報告一名患有嚴重主動脈瓣狹窄症的68歲女性。病人的腸道血管發育不良，引致腸胃出血的情況。我們使用單氣囊小腸鏡為病人確診及醫治。

Heyde症候群

have suggested an increased prevalence of gastro-intestinal angiodysplasia in patients with aortic stenosis. A similar association with angiodysplasia is also observed among patients with end-stage renal failure and von Willebrand’s disease. In this patient, it was postulated that the bleeding of angiodysplasia was triggered by her uncorrected severe aortic stenosis. When blood passed through the narrowed aortic valve, the turbulent flow generated caused mechanical disruption of the von Willebrand’s multimers. The deficiency of von Willebrand’s factor led to a secondary reduction in plasma factor VIII level and impaired platelet adherence to subendothelial components. Because of this acquired haematological defect, also known as von Willebrand’s syndrome, patients with uncorrected aortic stenosis are more prone to bleed from previously latent angiodysplasia.4,5 Another plausible explanation for Heyde’s syndrome is that the bleeding is the result of ischaemic necrosis caused by low cardiac output. But a similar phenomenon cannot be reproduced in other cardiac conditions causing low cardiac output.6 Nevertheless, the other compelling piece of evidence is that in the majority of patients, such haematological abnormalities and bleeding episodes are reversed by aortic valvular replacement.7

The diagnosis of obscure gastro-intestinal bleeding has always been difficult and definitive management is often delayed, as illustrated in this example. Wireless capsule endoscopy is a major breakthrough in the investigation of such small bowel lesions. Not only is it useful for detecting and localising small bowel lesions, it also helps to determine the route (antegrade or retrograde) of enteroscope insertion. The procedure is non-invasive and is currently the investigation of choice for obscure gastro-intestinal bleeding.8 The disadvantages of capsule endoscopy include: a risk of capsule retention; a biopsy cannot be performed; it has no therapeutic function; and the capsule direction cannot be purposely controlled. In this case, the angiodysplasia was obscured by blood during the capsule endoscopy examination so the exact source of bleeding could only be determined after an enteroscopic examination. Mesenteric angiography was not performed in this patient because the diagnostic yield is usually low in patients with obscure gastro-intestinal bleeding. In addition, potentially life-threatening complications (eg arterial dissection, bowel infarction and gangrene) may arise after vascular embolisation. Surgical resection during intra-operative enteroscopy can be considered for patients with large transfusion needs. Nevertheless, the concomitant severe aortic stenosis in Heyde’s syndrome may render the patient unfit for surgical intervention. Small bowel enteroscopy (push, ropeway and Sonde method) has been used for the last 30 years. Nevertheless, these methods are not widely practised because of the limited length of small bowel examined and the difficulties with manipulation. The procedure may also cause considerable pain and
discomfort due to overstretching of the intestine.

In 2001, the double-balloon enteroscopy (DBE) system introduced by Yamamoto et al. revolutionised the management of small bowel disease. Pressure-controlled balloons are attached to the overtube and the tip of the enteroscope. These help to shorten the small intestine and prevent the enteroscope from slipping out while the overtube is advanced. Through deep insertion of the endoscope via the oral and anal routes, safe inspection of the entire small bowel has now been made possible. Compared with capsule endoscopy, the DBE not only produces much higher quality images, it also permits concomitant use of a variety of diagnostic techniques such as tissue sampling, dye spraying, and contrast-enhanced examinations. Therapeutic interventions like achieving haemostasis, performing polypectomies, stricture dilatation and stenting can then be performed with this novel invention. Recently, a newer SBE system has been introduced for similar purposes. Instead of the balloon, the maximal angulations of a dedicated endoscope are used as an anchorage. The endoscopist thus needs to handle only one balloon from the overtube, which can save insertion time. Technically speaking, the SBE is easier to perform, requires a shorter preparation time, and is associated with a low complication rate. Nevertheless, the insertion efficiency of the SBE seems to be inferior to that of the DBE system. This was reflected by a lower rate of complete small bowel examinations in different clinical series (SBE 25% vs DBE 86%) but direct comparative studies are lacking.

In conclusion, this report serves to alert physicians to the possible association of severe aortic stenosis with bleeding angiodysplasia. At the same time, it illustrates that successful management of such lesions in patients with severe aortic stenosis can be safely accomplished with the novel SBE enteroscopy system.

References