Conservative management of diffuse cavernous haemangioma of the sigmoid and rectum

E Cotzias, SF Rehman, R Arsalani Zadeh, D Smith

Royal Bolton Hospital NHS Foundation Trust, Bolton, UK

ABSTRACT

Diffuse cavernous haemangioma is a rare disease of the rectum. It usually presents with a history of rectal bleeding in children and young adults. When conservative methods fail to control bleeding, traditionally resection is recommended. A 50-year-old man presented with per rectal bleeding and was diagnosed with diffuse cavernous haemangioma of the sigmoid and rectum extending up to 40 cm in the left colon through endoscopy, magnetic resonance imaging and computed tomography. The diagnosis was confirmed by biopsy. This patient was successfully managed conservatively with tranexamic acid as needed, avoiding the need for resection.

KEYWORDS

Colorectal – Surgery – Haemangioma

Accepted 5 May 2019

CORRESPONDENCE TO

S Rehman, E: sheik.rehman@nhs.net

Background

Cavernous haemangioma is a rare benign vascular malformation, with only approximately 350 cases being reported.¹ The diagnosis is often delayed and many patients will have been investigated and treated for other causes of rectal bleeding.^{1–4} It most frequently presents in children and young adults,^{4–7} with symptoms of recurrent painless rectal bleeding.^{5,8} Colonoscopy, magnetic resonance imaging (MRI) and computed tomography (CT) are all accepted investigations.⁸ Generally, diffuse cavernous haemangioma of the rectum (DCHR) should not be biopsied, as there is a risk of bleeding.^{3,8} The diagnosis should therefore be confirmed on pathology following surgical resection.⁹ The traditional approach in the published literature is to manage cavernous haemangioma with surgical resection,^{5,6,8} as conservative management poses the risk of recurrent bleeding.^{10,11} Various surgical approaches have been described, including low anterior resection, proctocolectomy abdominoperineal resection, and more recent cases reporting transanal total mesorectal excision.^{3,4} However, surgery is associated with increased morbidity and mortality.

We present a case report of a patient who has been successfully managed conservatively for over four years, with as needed tranexamic acid for the last two years.

Case history

A 50-year-old man was referred to the surgical department with a three-week history of fresh, painless rectal bleeding, weight loss and a reduced appetite. He underwent a colonoscopy (images 1). The first colonoscopy with biopsies

reported an ‘unusual appearance throughout the left colon (up to 40 cm) with evidence of multiple arteriovenous malformations’ and ‘prominent veins within the mucosa with a differential diagnosis of portal hypertension’. Other differential diagnosis included atypical ulcerative colitis, vasculitis and proctitis. In view of the colonoscopy findings, the patient went on to have CT abdomen/pelvis with contrast, which showed mild rectal wall thickening (Image 2), corresponding to the vascular abnormality seen on colonoscopy.

A few weeks later, the patient was admitted to hospital with more frequent, heavier rectal bleeding and symptoms of anaemia, with a haemoglobin of 69 g/l (normocytic). He was resuscitated and further imaging arranged.

The histology from the initial colonoscopy reported ‘thickened muscularis mucosae containing irregular shaped, greatly dilated vascular spaces lined with uniform endothelial cells’ and ‘occasional dilated vascular spaces deep in the lamina propria’. These appearances were in keeping with angioma or telangiectasis. He underwent MRI of the rectum (image 3) to further investigate the cause of his bleeding. This showed concentric mural thickening of the wall of the entire rectum with high signal intensity on T2. The appearances on MRI, in conjunction with histology, were thought to be in keeping with a diagnosis of diffuse cavernous haemangioma of the sigmoid and rectum. During this admission he required repeated blood transfusions (average two per week) and was commenced on oral tranexamic acid 1 g three times a day.

Management was multidisciplinary with the involvement of a radiologist, gastroenterologist and surgeons. The

Image 1 Endoscopic appearances.

Image 3 Magnetic resonance sagittal image showing thickened rectum with vascular malformations.

Image 2 Computed tomography; sagittal section during portovenous phase showing thickened rectum, phlebolith and enhancement.

agreed plan was for active observation and management with tranexamic acid, however, bowel resection would be considered if conservative management were to fail. CT angiography and embolisation was not an option as mainly venous malformations.

This patient has now been on as needed tranexamic acid for over two years, which he commences at the onset of bleeding and suspends once the bleeding has settled, usually only requiring a few days of treatment to control the bleeding. He has not required further blood or iron transfusions for over two years. Currently, he is followed up by flexisigmoidoscopy surveillance every three years.

Discussion

The traditional method of managing DCHR is surgical resection, which is tailored depending on the location and extent of the haemangioma. One case report described a method of transanal total mesorectal excision with a coloanal anastomosis and loop ileostomy, which appeared to be an effective technique.⁵ Another case report described performing three-dimensional laparoscopy-assisted abdominoperineal resection on a patient with DCHR invading the posterior wall of the vagina, and three-dimensional laparoscopy-assisted pull-through transection and coloanal anastomosis on a patient with a tortuous vein 2 cm above the dentate line. Both procedures appeared successful at a six-month follow-up.⁶ Although such cases have been successful and are arguably less invasive than open procedures, surgical interventions may not be suitable for all patients and are not without risk.

More conservative approaches, such as sclerotherapy, have been described in an case report of a seven-year-old with Klippel-Trenaunay-Weber syndrome had colonic haemangiomas ranging between 5 cm and 15 cm in size. These were successfully managed with sclerotherapy. The patient has had no further episodes of gastrointestinal bleeding over a three-year follow-up period.¹²

Exploring conservative methods remains prudent and our case is an example of a successful conservative management approach for DCHR with a four-year follow-up. This gentleman uses tranexamic acid as needed at the onset of bleeding. He experiences bleeding approximately every six weeks, which lasts a few days. It may be the case that if his symptoms worsen or he suffers with anaemia more definitive treatment may be required, but at present tranexamic acid has been an effective strategy to manage his condition. Haemangiomas generally do not continue to grow beyond a few days to months after birth, and this probably also applies to DCHR.² Similarly, there was no change in size in this case during the four-year observation period. Haemangiomas are also not suitable for angioembolisation as they are diffuse venous malformations.

This case also further highlights the known difficulties that arise in diagnosing gastrointestinal haemangiomas and the potential scope for misdiagnosis. Prior to the conclusive diagnosis this patient was investigated for vasculitis and ulcerative colitis among other differentials. The challenging nature of his diagnosis may have been further compounded by the fact that cavernous haemangiomas usually present in children and young adults.⁶ The difficulty in diagnosis is reflected in much of the literature,¹ and other cases report patients with colonic cavernous haemangiomas who were diagnosed and treated as haemorrhoids in the initial stages.⁵ To avoid delayed or misdiagnosis, a diagnosis of haemangioma should be considered in patients who present with a long history of painless rectal bleeding, while remaining mindful of the rarity of this condition as a cause of rectal bleeding in comparison with other diagnoses. Arguably, in this case, MRI was the most useful investigation. Other studies also report that MRI can be a useful diagnostic tool for cavernous haemangioma.^{2,3,13–15}

Conclusion

DCHR can be a delayed and difficult diagnosis. MRI is a sound diagnostic investigation. Although traditionally

surgical resection has been preferred, a more conservative treatment with as needed tranexamic acid is a safe alternative to surgery. However, further evidence is required, with a longer follow-up period, to establish whether this is an effective treatment option.

References

1. Wang H, Gao X, Fu C *et al.* Diagnosis and treatment of diffuse cavernous hemangioma of the rectum: report of 17 cases. *World J Surg* 2010; **34**(10): 2,477–2,486.
2. Osaki K, Mori Y, Ozaki Y *et al.* Successful conservative management of diffuse cavernous hemangioma of the rectum. *Int Cancer Conf J* 2016; **6**(1): 8–10.
3. Rodríguez-Zentner H, Castañeda-Argáiz R, Tapia H *et al.* Colonic cavernous hemangiomas: a report of two cases treated by laparoscopic surgery. *Asian J Endosc Surg* 2011; **4**(2): 78–81.
4. Maddah G, Abdollahi A, Rouhbakhshfar Z *et al.* Arterio-venous malformations of the colon: a report of two cases and review of the literature. *Caspian J Intern Med* 2017; **8**(1): 52–55.
5. Yoo S. GI-associated hemangiomas and vascular malformations. *Clin Colon Rectal Surg* 2011; **24**(3): 193–200.
6. Fu Z, Wang L, Zhang Z *et al.* Three-dimensional laparoscopy-assisted bowel resection for cavernous hemangioma of the rectum: report of two cases. *Asian J Endosc Surg* 2018; Aug 9. doi: 10.1111/ases.12636.
7. Ugolini G, Rosati G, Montroni I *et al.* Diffuse cavernous haemangioma of the rectum and anus: an unusual case of rectal bleeding with challenging management. *Case Rep* 2009; **2009**: pii bcr02.2009.1545.
8. Yorozuya K, Watanabe M, Kitajima M *et al.* Diffuse cavernous hemangioma of the rectum: report of a case. *Surg Today* 2003; **33**(4): 309–311.
9. Z Oner, G Altaca. Diffuse cavernous rectal hemangioma clinical appearance, diagnostic modalities and sphincter saving approach to therapy: report of 2 and a collective review of 79 cases. *Acta Chir Belg* 1993; **93**: 173–176.
10. Sylla P, Deutsch G, Luo J *et al.* Cavernous, arteriovenous, and mixed hemangioma-lymphangioma of the rectosigmoids: Rare causes of rectal bleeding: case series and review of the literature. *Int J Colorectal Dis* 2008; **23**: 653–658.
11. Wu X, Liang W, Zhang X *et al.* Transanal total mesorectal excision as a surgical procedure for diffuse cavernous hemangioma of the rectum: a case report. *Int J Surg Case Rep* 2017; **39**: 164–167.
12. Parashette KR, Cuffari C. Sclerotherapy of rectal hemangiomas in a child with Klippel-Trenaunay-Weber syndrome. *J Pediatr Gastroenterol Nutr* 2011; **52**(1): 111–112.
13. Aylward C, Orangio G, Lucas G, Fazio V. Diffuse cavernous hemangioma of the rectosigmoid: CT scan, a new diagnostic modality, and surgical management using sphincter-saving procedures. *Dis Colon Rectum* 1988; **31**(10): 797–802.
14. Hakeem A, Shafi H, Gojwari T *et al.* CT Findings in diffuse rectosigmoid cavernous haemangioma: a case report. *J Gastrointest Surg* 2008; **13**(5): 1,017–1,018.
15. Kandpal H, Sharma R, Srivastava D *et al.* Diffuse cavernous haemangioma of colon: magnetic resonance imaging features. Report of two cases. *Australas Radiol* 2007; **51**: B147–B151.