Case Report

Journal of INTERNATIONAL MEDICAL RESEARCH

Journal of International Medical Research
49(1) 1–8

DOI: 10.1177/0300060520986677
journals.sagepub.com/home/imr
SAGE

Haemorrhagic ileal haemolymphangioma: a case report and review of the literature

Shou-Xing Yang*, Yu-Hui Zhou*, Jing Zhang*, Lei Miao, Jing-Wei Zhong, Wen-Xing Wang, Chang-Long Xu, Zhen-Zhai Cai and Guang-Rong Lu

Abstract

We describe herein a 37-year-old woman with a 2-week history of melena who was eventually diagnosed with ileal haemolymphangioma, a rare benign tumour. Local mucosal congestion and swelling were found through single-balloon enteroscopy, which showed an irregular protuberance approximately 10 cm long, located 3.2 m from the Treitz ligament. We performed a laparoscopic-assisted partial resection of the small intestine combined with intestinal adhesiolysis. According to postoperative pathology, the final diagnosis was ileal haemolymphangioma with haemorrhage.

Keywords

Haemolymphangioma, ileal, haemorrhage, lymphangioma, case report, enteroscopy

Date received: 17 July 2020; accepted: 15 December 2020

Introduction

Haemolymphangioma is a rare benign tumour characterised by both malformed venous and lymphatic histological components.[1,2] According to a review of the literature, haemolymphangioma occurs primarily in the head, neck, and armpit, as well as in the spleen, pancreas, liver, spine, and other areas, but rarely in the

Department of Gastroenterology, The Second Affiliated Hospital of Wenzhou Medical University, Wenzhou, Zhejiang, China

*These authors contributed equally to this work.

Corresponding author:
Guang-Rong Lu, Department of Gastroenterology, The Second Affiliated Hospital of Wenzhou Medical University, 109 West Lucheng College, Wenzhou, Zhejiang 325000, China.
Email: 290636246@qq.com

gastrointestinal tract.[3–6] Haemolymphangioma can be congenital or acquired, but they are most frequently congenital,[7] occurring in an estimated 1.2 to 2.8 per 1000 newborn infants.[8] In view of its low incidence rate, reports of this tumour are rare, and reaching the correct diagnosis preoperatively remains challenging. Recently, a patient presenting with gastrointestinal haemorrhage was admitted to the Department of Gastroenterology in our hospital. Local mucosal congestion and swelling were found through single-balloon enteroscopy, which showed an irregular protuberance approximately 10 cm long. The lesion was 3.2 m from the Treitz ligament. After performing a laparoscopic-assisted partial resection of the small intestine combined with intestinal adhesiolysis, the patient was diagnosed with ileal haemolymphangioma. The case report follows and the related literature is reviewed.

Case report

A previously healthy 37-year-old woman was admitted to our hospital on 30 March 2020 with a 2-week history of melena. The patient had begun to defecate shaped, tarry black stool 2 weeks previously (1 to 2 times/day, about 200 g each time) without haematemesis, accompanied by dizziness and fatigue. A routine blood count showed that the patient's haemoglobin was 43 g/L (normal range: 110–150 g/L). Blood biochemistry indicated no abnormal liver or kidney function, and the serum potassium level was 3.43 mmol/L (normal range: 3.50–5.30 mmol/L). Cardiac function and immunological indicators were also normal. The patient was admitted to our hospital because of gastrointestinal haemorrhage and severe anaemia. A physical examination at admission revealed the presence of severe anaemia and a soft abdomen, with no tenderness, rebound pain, or palpable mass.

On 2 April 2020, gastroscopy revealed a possible chronic gastritis; when the colonoscope was inserted 40 cm from the terminal ileum, fresh blood was observed in the small intestine, with blood in the entrance side of the large intestine, suggesting the possibility of small intestine haemorrhage. A capsule endoscopy was immediately performed, and the small bowel transit time was 5 hours and 5 minutes. Rough mucosa of the small intestine wall was observed 1 hour and 40 minutes after the capsule passed through the pylorus, and a local irregular protuberance with scattered white spots on the surface was observed, but no active bleeding was seen. Subsequent intestinal computed tomographic (CT) angiography on 6 April 2020 revealed no abnormalities.

On 7 April 2020, a single-balloon enteroscopy was performed via the upper gastrointestinal tract. The lesion was located 3.2 m from the Treitz ligament, determined by the entry depth of the enteroscope. Local mucosal congestion and swelling were found, showing an irregular protuberance approximately 10 cm long. (Figure 1). The protuberance had white, spot-like changes on the surface, with local ulceration, fresh blood stains and blood clots covering the surface, and active bleeding. The ileal area was sprayed with adrenaline (1:10,000) to stop the bleeding, and titanium clips were placed at the distal and proximal parts, 5 cm from the lesion, to mark the location.

Subsequently, preoperative preparations were made, and no obvious abnormalities were found on enhanced CT of the whole abdomen. Laparoscopic-assisted partial resection of the small intestine combined with intestinal adhesiolysis was performed on 10 April 2020. A mass about 4×3 cm was found in the ileum, approximately 1.5 m from the ileocecal junction. Following intestinal dissection, the mass appeared clustered, with multiple granular nodules (Figure 2). Postoperative pathology

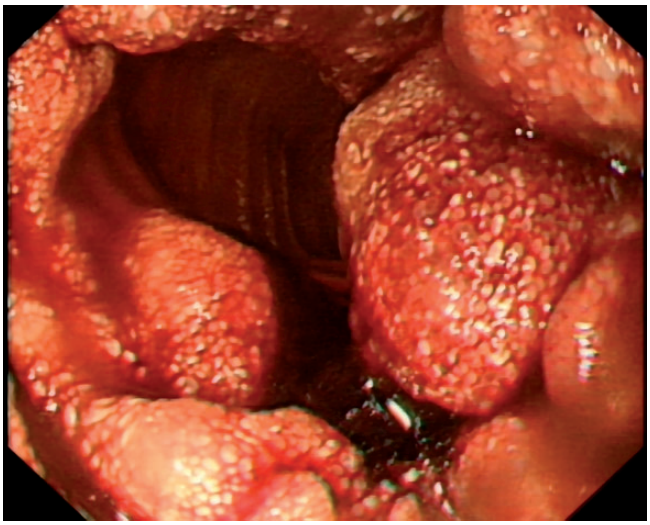

Figure 1. Single-balloon enteroscopy image showing congestion and swelling of the local mucosa with white spot-like changes on the surface.

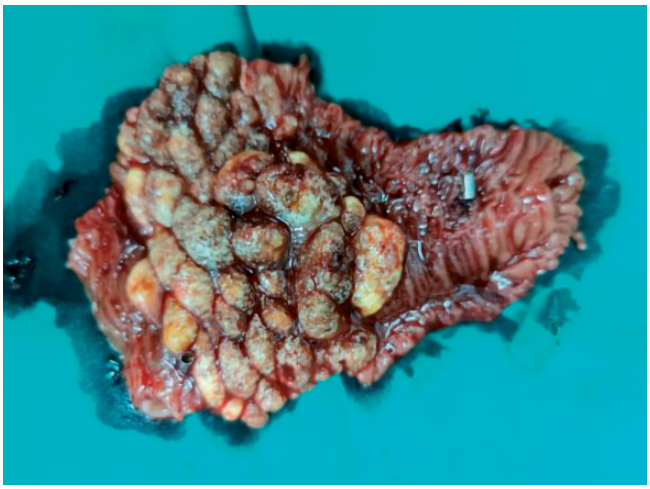

Figure 2. Intestinal dissection revealed a clustered mass measuring approximately 6.5 × 4.5 × 0.9 cm, with multiple granular nodules.

showed abundant diffuse proliferative blood and lymphatic vessels in the stroma of the small intestine mucosa and submucosal areas, with partial dilatation (Figure 3). A diagnosis of haemolymphangioma of the small intestine (approximately 6.5 × 4.5 × 0.9 cm) was considered. In addition, two reactive hyperplastic lymph nodes were detected in the surrounding mesentery. The diagnosis was ileal haemolymphangioma with haemorrhage.

On 25 May 2020, 1 month after the patient was discharged, routine blood examination showed a haemoglobin level

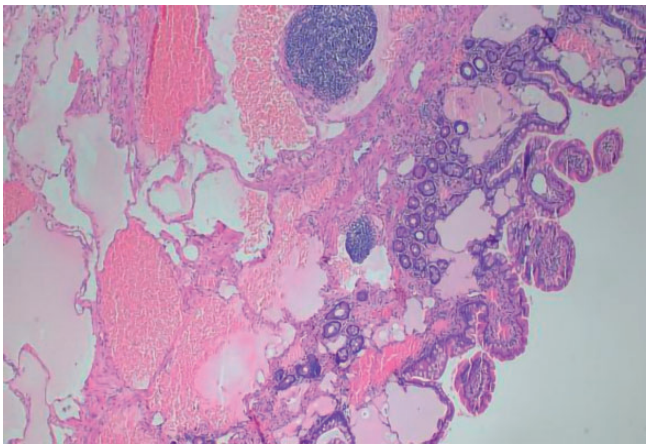

Figure 3. Partial small intestine specimens were observed by light microscopy following haematoxylin and eosin (H&E) staining (magnification = 100×) and showed abundant diffuse proliferative blood and lymphatic vessels in the stroma of the small intestine mucosa and submucosal areas, with partial dilatation.

of 127 g/L and a red blood cell count of 4.59×10^{12}/L (normal range: 3.5–5.0×10^{12}/L). The patient's blood biochemistry was normal. A CT of the chest and abdomen showed no obvious abnormalities.

Discussion

Lymphangioma is a rare type of benign lesion caused by abnormal development of lymphatic system. It may be caused by an abnormal interruption of the connection among lymphatic vessels, leading to lymphatic fluid reflux obstruction, accumulation of fluid in the local lymphatic vessels, and gradual formation of cystic tumour-like expansion.[9,10] Based on the degree of lymphatic dilation, lymphangioma can be divided into three types of capillary lymphangioma: cavernous lymphangioma, cystic lymphangioma, and haemolymphangioma.[3]

Haemolymphangioma, also known as vascular tumour, is a clinically rare lymphangioma that originates from mesenchymal tissue, and it is composed histologically of both vascular and lymphatic components.[11,12] Haemolymphangioma is now considered to be a congenital benign tumour.[13] It has an incidence of about 1.2% to 2.8% in neonates, occurring in both males and females, with no difference in incidence.[13] Approximately 90% of patients are diagnosed before the age of 2 years, and about 60% are born with corresponding symptoms such as gastrointestinal bleeding and intestinal obstruction. Furthermore, secondary haemolymphangioma can develop from poor drainage of lymphatic fluid caused by infection, trauma, or surgery.[12]

In our case, the possibility of primary haemolymphangioma was considered because the patient had no previous history of intestinal surgery or trauma. The patient may have had haemolymphangioma for many years without clinical symptoms and was eventually diagnosed because of ulceration and bleeding on the tumour surface. Our literature review suggests that haemolymphangioma can be located in the spleen, pancreas, liver, mediastinum, spine, and other areas, but is extremely rare in the small intestine. Fang et al.[14] first reported small intestinal haemolymphangioma in 2012. Xiao et al.[15] found six reports of

small intestinal haemolymphangioma from 2010 to 2019, in which the tumours were located in the proximal jejunum or duodenum, with anaemia caused by gastrointestinal haemorrhage as the primary clinical symptom. At present, we can find no relevant literature about ileal haemolymphangioma. In addition to causing gastrointestinal haemorrhage, small intestinal haemolymphangioma can also compress adjacent organs because of tumour growth and cause abdominal pain, abdominal distension, intussusception, intestinal obstruction, and other symptoms.[14,16] However, these symptoms lack specificity.

Regarding intestinal haemolymphangioma, reaching the correct diagnosis preoperatively is challenging because of the low incidence rate and the lack of specific clinical findings. Haemolymphangioma generally presents as a solid or cystic tumour (few of which are completely cystic). The cystic part may be caused by the expansion and fusion of the vascular cavity, whereas the solid part may be residual extruded vascular tissue.[17] For tumours with abundant blood vessels, enhancement in the cystic wall and septum can be observed by CT, especially in the venous and delayed phases.[18] However, in our patient, we observed no obvious abnormalities in the small intestine by CT angiography, which may be attributed to the small tumour volume, the relatively low degree of tumour vascularisation, or the slow blood flow caused by vascular dysplasia. Furthermore, magnetic resonance imaging (MRI) can be useful to determine the relationship between the tumour and its surrounding tissues, as well as the degree of infiltration. The cystic component shows mainly low or slightly low signal on T1-weighted imaging and high signal on T2-weighted imaging, which is generally associated with the proportion of blood and lymphatic vessels in the tumour. In addition, endoscopy plays an important role in diagnosing small intestinal haemolymphangioma. Among these methods, capsule endoscopy has advantages: it is a simple, non-invasive, painless, and relatively safe operation, and it has become a first-line examination approach for diseases of the small intestine.[19] However, there may be complications, including capsule retention, and its uncontrolled operation depends on gastrointestinal peristalsis, which may reduce the observational accuracy of capsule endoscopy, with blind areas preventing photography. Enteroscopy has a higher diagnostic value for small bowel diseases. According to the Chinese Guideline for Clinical Application of Enteroscopy, for patients with positive results during capsule endoscopy, enteroscopy not only yields a more intuitive and comprehensive picture of the lesion's characteristics but can also determine its nature through endoscopic biopsy.[20] Enteroscopy can be used to localize the lesion, such as by placing titanium clips around the lesion. If necessary, endoscopic treatment can be carried out simultaneously with the lesion located and marked.

In our case, no abnormality was found using gastroscopy, suggesting a small intestinal haemorrhage of unknown cause. Following localisation of the lesion by capsule endoscopy, we carried out single-balloon enteroscopy, discovering active bleeding on the lesion's surface. Adrenaline (1:10,000) spray was applied to stop the bleeding, and a titanium clip was used as an identification marker for surgery. We reviewed and summarised endoscopic images of small intestinal haemolymphangioma reported in the past 10 years. According to the results, the focus mainly manifests as multiple polypoid or an irregular yellowish-white protrusion, with localised erosion or ulceration, and dense, white spot-like surface changes.[1,14,15] Because there are abundant vascular components in haemolymphangioma tumour, biopsy can easily cause or aggravate bleeding. With respect

to the aforementioned details, considering the possibility of small intestinal haemolymphangioma and avoiding biopsy in this case are necessary.

Several studies indicate that haemolymphangioma is generally localised and non-invasive.[14–16] However, a few reports describe infiltration of adjacent organs in haemolymphangioma. For example, Sun et al.[21] found lesion invasion of the mesentery and omentum of the transverse colon with duodenal and superior mesenteric artery adhesions in a patient who underwent surgical excision for giant pancreatic haemolymphangioma. Toyoki et al.[22] reported a case of duodenal invasion by haemolymphangioma of the head of the pancreas, leading to gastrointestinal bleeding. Surgery is considered the most effective choice for treating haemolymphangioma, and radical resection is necessary to prevent its recurrence. In addition, complete resection should be considered in case of adhesion or invasion of tumour to surrounding organs.[14,23] The recurrence rate of haemolymphangioma is 10% to 27% after radical resection and 50% to 100% after partial resection.[6] In terms of haemolymphangioma of the digestive system, endoscopic treatment can be considered in addition to surgery. For instance, Xiao et al.[15] reported a case of jejunal haemolymphangioma with haemorrhage that was treated by endoscopic sclerotherapy. One-year postoperative follow-up results with endoscopy revealed that the focus had disappeared, and only a few white spots remained on the mucosal surface. Furthermore, in some cases, endoscopic mucosal resection or endoscopic submucosal dissection has been performed for oesophageal haemolymphangioma with submucosal eminence, with good results.[24,25]

In summary, we report here a rare case of ileal haemolymphangioma with nonspecific clinical signs. Gastrointestinal bleeding was our patient's initial symptom, and was not accompanied by abdominal pain and distension and intestinal obstruction. Capsule endoscopy and enteroscopy are valuable in reaching a diagnosis for this tumour type. A diagnosis of haemolymphangioma can be considered in case of multiple irregular bumps and dense white spots on the lesion's surface; however, pathological examination is still required for final diagnostic confirmation. At present, surgical resection is the main choice for treating haemolymphangioma, although endoscopic treatment such as argon plasma coagulation or sclerotherapy can be attempted. Long-term follow-up is important in the later stages of the disease.

Author contributions

Lei Miao, Jing-Wei Zhong, Wen-Xing Wang, and Chang-Long Xu collected the clinical data; Shou-Xing Yang, Yu-Hui Zhou, and Jing Zhang wrote and revised the article; Zhen-Zhen Cai and Guang-Rong Lu modified and edited the article.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images. The research was approved by the Ethics Committee of the Second Affiliated Hospital of Wenzhou Medical University.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

ORCID iD

Jing Zhang https://orcid.org/0000-0001-9899-4549

References

1. Antonino A, Gragnano E, Sangiuliano N, et al. A very rare case of duodenal hemolymphangioma presenting with iron deficiency

anemia. *Int J Surg Case Rep* 2014; 5: 118–121. DOI: 10.1016/j.ijscr.2013.12.026.
2. Ahmad K, Vad H, Christensen TD. Mediastino-hepato-renal cystic lymphangiomas-diagnostic and surgical considerations. *J Thorac Dis* 2014; 6: E173–175. DOI: 10.3978/j.issn.2072-1439.2014.08.47.
3. Kosmidis I, Vlachou M, Koutroufinis A, et al. Hemolymphangioma of the lower extremities in children: two case reports. *J Orthop Surg Res* 2010; 5: 56. DOI: 10.1186/1749-799x-5-56.
4. Bosdure E, Mates M, Mely L, et al. Cystic intrathoracic hemolymphangioma: a rare differential diagnosis of acute bronchiolitis in an infant. *Arch Pediatr* 2005; 12: 168–172. DOI: 10.1016/j.arcped.2004.11.012.
5. Cophignon J, d'Hermies F, Civit T. Vascular tumors of the orbit. *Neurochirurgie* 2010; 56: 197–212. DOI: 10.1016/j.neuchi.2010.02.028.
6. Wang LC, Krunic AL, Medenica MM, et al. Treatment of hemorrhagic lymphatic malformation of the tongue with a pulsed-dye laser. *J Am Acad Dermatol* 2005; 52: 1088–1090. DOI: 10.1016/j.jaad.2005.03.014.
7. Rai HC, Krishnamoorthy A, Dayakar M. Hemolymphangioma of the neck: a rare vascular malformation in children. *Internet J Pediatr Neonatol* 2013; 16.
8. Filston HC. Hemangiomas, cystic hygromas, and teratomas of the head and neck. *Semin Pediatr Surg* 1994; 3: 147–159.
9. Richter GT and Friedman AB. Hemangiomas and vascular malformations: current theory and management. *Int J Pediatr* 2012; 2012: 645678. DOI: 10.1155/2012/645678.
10. Giuliani A, Romano L, Coletti G, et al. Lymphangiomatosis of the ileum with perforation: A case report and review of the literature. *Ann Med Surg (Lond)* 2019; 41: 6–10. DOI: 10.1016/j.amsu.2019.03.010.
11. Zhang Y, Chen XM, Sun DL, et al. Treatment of hemolymphangioma of the spleen by laparoscopic partial splenectomy: a case report. *World J Surg Oncol* 2014; 12: 60. DOI: 10.1186/1477-7819-12-60.
12. Chen G, Cui W, Ji XQ, et al. Diffuse hemolymphangioma of the rectum: a report of a rare case. *World J Gastroenterol* 2013; 19: 1494–1497. DOI: 10.3748/wjg.v19.i9.1494.
13. Figueroa RM, Lopez GJ, Servin TE, et al. Pancreatic hemolymphangioma. *JOP* 2014; 15: 399–402. DOI: 10.6092/1590-8577/2649.
14. Fang YF, Qiu LF, Du Y, et al. Small intestinal hemolymphangioma with bleeding: a case report. *World J Gastroenterol* 2012; 18: 2145–2146. DOI: 10.3748/wjg.v18.i17.2145.
15. Xiao NJ, Ning SB, Li T, et al. Small intestinal hemolymphangioma treated with enteroscopic injection sclerotherapy: A case report and review of literature. *World J Gastroenterol* 2020; 26: 1540–1545. DOI: 10.3748/wjg.v26.i13.1540.
16. Teng Y, Wang J, Xi Q. Jejunal hemolymphangioma: A case report. *Medicine (Baltimore)* 2020; 99: e18863. DOI: 10.1097/MD.0000000000018863.
17. Dong F, Zheng Y, Wu JJ, et al. Hemolymphangioma: a rare differential diagnosis of cystic-solid or cystic tumors of the pancreas. *World J Gastroenterol* 2013; 19: 3520–3523. DOI: 10.3748/wjg.v19.i22.3520.
18. Mao CP, Jin YF, Yang QX, et al. Radiographic findings of hemolymphangioma in four patients: A case report. *Oncol Lett* 2018; 15: 69–74. DOI: 10.3892/ol.2017.7268.
19. Makins R and Blanshard C. Guidelines for capsule endoscopy: diagnoses will be missed. *Aliment Pharmacol Ther* 2006; 24: 293–297. DOI: 10.1111/j.1365-2036.2006.02991.x.
20. The role of colonoscopy in the management of patients with colonic polyps. Guidelines for clinical application. *Gastrointest Endosc* 1988; 34: 6s–7s.
21. Sun LF, Ye HL, Zhou QY, et al. A giant hemolymphangioma of the pancreas in a 20-year-old girl: a report of one case and review of the literature. *World J Surg Oncol* 2009; 7: 31. DOI: 10.1186/1477-7819-7-31.
22. Toyoki Y, Hakamada K, Narumi S, et al. A case of invasive hemolymphangioma of the pancreas. *World J Gastroenterol* 2008; 14: 2932–2934. DOI: 10.3748/wjg.14.2932.
23. Hancock BJ, St-Vil D, Luks FI, et al. Complications of lymphangiomas in children. *J Pediatr Surg* 1992; 27: 220–224;

discussion 224–226. DOI: 10.1016/0022-3468(92)90316-y.
24. Cheng Y, Zhou X, Xu K, et al. Esophageal lymphangioma: a case report and review of literature. *BMC Gastroenterol* 2019; 19: 107. DOI: 10.1186/s12876-019-1026-9.
25. Suwa T, Ozawa S, Ando N, et al. Case report: lymphangioma of the oesophagus endoscopically resected. *J Gastroenterol Hepatol* 1996; 11: 786–788. DOI: 10.1111/j.1440-1746.1996.tb00333.x.