CASE REPORT

Caecal lymphangioma: a rare cause of gastrointestinal blood loss

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SUMMARY

Lymphangioma is the malformation of the lymphatic system. Lymphangioma is a benign tumour and most colonic lymphangiomas do not cause symptoms and do not require treatment. Resection is required in patients with bleeding or intussusceptions. We report a case of intestinal lymphangiomatosis in a 31-year-old man who presented with recurrent melena and anaemia, which were diagnosed endoscopically and treated with surgical resection.

BACKGROUND

Lymphangioma is the benign proliferation of lymph vessels. Lymphangioma is a submucosal tumour covered with normal mucosa.^{1 2} It can occur anywhere in the body and only rarely affects the intestinal tract. However, it has been reported more frequently nowadays because of the widespread use of endoscopy.

This case is important as it highlights that caecal lymphangioma, an uncommon entity, can be a cause for gastrointestinal blood loss and should be considered in the differential diagnosis of anaemia.

CASE PRESENTATION

A 31-year-old man was admitted in our hospital with a history of recurrent melena for the last 8 years and iron deficiency anaemia. The patient had a total of eight episodes of melena requiring 10 units of packed red blood cell transfusions. There was no history of abdominal pain, fever, abdominal distension and abdominal trauma.

He had no significant medical history, and no history suggestive of pancreatitis, tuberculosis, Wegener's granulomatosis and polyarteritis nodosa. The patient was non-alcoholic and a non-smoker. There was no significant family history. Physical examination revealed no remarkable abnormality in the abdomen.

INVESTIGATIONS

Laboratory tests showed hypochromic microcytic anaemia, 4.4 g/dl haemoglobin, 15.7% haematocrit, 66.5 fl mean corpuscular volume (normal=85–99), 17.3 pg mean corpuscular haemoglobin (normal=26–34), 26% mean corpuscular haemoglobin concentration (normal=32–36%), 15 µg/dl iron (normal=65–157) and 3.59 ng/ml ferritin (normal=16.4–323). Liver function test revealed serum albumin 2.3 g/dl (3.5–5.0 g/dl) and the rest of the liver and renal function tests were normal.

Chest x-ray was normal. Supine and upright views of the abdomen disclosed a non-specific pattern of

bowel gas. Barium enteroclysis done 1 year ago was normal. Oesophagogastroduodenoscopy showed normal study on two separate occasions. Colonoscopy revealed protruding submucosal lesions of approximately 20–30 mm in diameter at the caecum (figure 1). The surface was covered with normal mucosa. This lesion was smooth, glossy and translucent, and the cushion sign was positive. Histopathology of endoscopic biopsy showed mild mononuclear infiltrate in lamina with focal prominence of goblet cells and no evidence of granuloma or malignant cells. Contrast-enhanced CT abdomen revealed small cystic lesion in ascending colon with few small mesenteric lymph node.

Capsule endoscopy showed a normal small bowel study.

TREATMENT

Patient was kept on follow-up, but there was a gradual drop in haemoglobin. Colonoscopy after 6 months showed a similar finding. Repeat histopathological examination was inconclusive. Patient was planned for laparotomy with intraoperative endoscopy. Enteroscopy revealed multiple diffuse submucosal soft nodular lesions of varying sizes (5–10 mm) with overlying normal mucosa extending from mid-jejunum to ileocecal junction: one fleshy polypoidal lesion in caecum of 40–50 mm size with ulcerated mucosa. No active bleeding lesion was seen. Laparotomy revealed a leash of blood vessels and multiple fleshy sessile pedunculated lesions overlying serosal aspect of small bowel and small bowel mesentery starting from mid-jejunum to ileocecal junction (figure 2). Limited ileocecal resection was performed. An end ileostomy and distal mucus fistula was done as the patient's haemoglobin and

Figure 1 Colonoscopy showing lymphangioma in caecum.

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Figure 2 Laparoscopy showing lymphangioma.

albumin were low. Postoperatively, there was no gastrointestinal (GI) bleed. After few days, when haemoglobin and albumin normalised, a revision surgery was done and continuity was restored. Histopathological examination of sections from caecal polypoidal lesion and serosal pedunculated lesions showed multiple irregular dilated space lined by endothelial cells with focal lymphoid aggregates; few were thick walled with muscular media, filled with acellular proteinaceous material and no evidence of malignant cells (figure 3). Based on the clinical and histopathological findings, a diagnosis of intestinal lymphangiomatosis was made.

OUTCOME AND FOLLOW-UP

The patient was healthy with maintenance of normal haemoglobin on subsequent 6 months and 1 year of follow-up.

DISCUSSION

Lymphangioma, a benign tumour of the lymphatics, is commonly found in the head and neck or axillary region. Lymphangioma occurs rarely in abdomen and most of them arise in the mesentery, omentum, mesocolon and retroperitoneum. Those arising in the wall of the intestine are considered to be even rarer.¹

Lymphangiomas occur mainly in children; approximately 80–90% is diagnosed within the first few years of life and adult cases are rare.^{3–4}

Figure 3 Photomicrograph of lymphangioma.

Lymphangioma of the colon was first described by Chisholm and Hillkowitz in 1932.⁵

The patients are usually asymptomatic and are often discovered during the evaluation of an unrelated condition. Some patients may have abdominal pain and alteration in bowel habits. Lower GI bleeding, obstruction, intussusception and protein-losing enteropathy are rare complications.¹

Traditionally, lymphangiomas are classified as simple, cavernous or cystic. The simple type is usually situated superficially in the skin and composed of small thin-walled lymphatic vessels. The cavernous type is composed of dilated lymphatic vessels and lymphoid stroma, and has a connection with spaces of various normal adjacent lymphatics. Finally, the cystic type consists of lymphatic spaces of various sizes and contains fascicles of smooth muscle and collagen bundles, but has no connection with the bowel.

The differential diagnosis for submucosal tumours of the intestine includes lipoma, lymphangioma and leiomyoma. Carcinoid tumours and metastatic lesions can also manifest as submucosal masses. Barium enema, colonoscopy, abdomen CT, endoscopic ultrasonography are useful in its diagnosis.^{6–8} Endoscopic ultrasound helps in differentiating among these lesions by determining their layer of origin when they are accessible.

In some cases, such as ours, surgical resection may be required to provide definitive diagnosis and treatment.

Various forms of treatment have been described. Pedunculated or semipedunculated colonic lymphangioma can be managed by endoscopic polypectomy.^{9–10} But, colonic perforation is a well-known complication. Other authors have developed a technique of deroofting the lymphangioma lesion with cyst drainage. In our case, recurrent GI bleed, the endoscopic appearance and the large tumour size guided our choice of surgical resection.

Learning points

- ▶ Caecal lymphangioma is the cause of gastrointestinal blood loss.
- ▶ Caecal lymphangioma has a benign course.
- ▶ Large and symptomatic lymphangiomas need surgery.

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REFERENCES

- 1 Matsuda T, Matsutani T, Tsuchiya Y, *et al.* A clinical evaluation of lymphangioma of the large intestine: a case presentation of lymphangioma of the descending colon and a review of 279 Japanese cases. *J Nippon Med Sch* 2001;68:262–5.
- 2 Wildhaber BE, Chardot C, Coulter CL, *et al.* Total laparoscopic excision of retroperitoneal cystic lymphangioma. *J Laparoendosc Adv Surg Tech A* 2006;16:530–3.
- 3 Hanagiri T, Baba M, Shimabukuro T, *et al.* Lymphangioma in the small intestine: report of a case and review of the Japanese literature. *Surg Today* 1992;22:363–7.
- 4 Chen CW, Hsu SD, Lin CH, *et al.* Cystic lymphangioma of the jejunal mesentery in an adult: a case report. *World J Gastroenterol* 2005;11:5084–6.

- 5 Chisholm AJ, Hillkowitz P. Lymphangioma of the rectum. *Am J Surg* 1932;17:281–2.
- 6 Kawamoto K, Ueyama T, Iwashita I, *et al.* Colonic submucosal tumors: comparison of endoscopic US and target air-enema CT with barium enema study and colonoscopy. *Radiology* 1994;192:697–702.
- 7 Fujimura Y, Nishishita C, Iida M, *et al.* Lymphangioma of the colon diagnosed with an endoscopic ultrasound probe and dynamic CT. *Gastrointest Endosc* 1995;41:252–4.
- 8 Kochman ML, Wiersema MJ, Hawes RH, *et al.* Preoperative diagnosis of cystic lymphangioma of the colon by endoscopic ultrasound. *Gastrointest Endosc* 1997;45:204–6.
- 9 Sato K, Maekawa T, Yabuki K, *et al.* Cystic lymphangiomas of the colon. *J Gastroenterol* 1999;34:520–4.
- 10 Nakagawara G, Kojima Y, Mai M, *et al.* Lymphangioma of the transverse colon treated by transendoscopic polypectomy: report of a case and review of literature. *Dis Colon Rectum* 1981;24:291–5.

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