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Intermittent melena and refractory anemia due to jejunal cavernous lymphangioma: An unusual case presentation

Jejunal cavernous lymphangioma

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Abstract

BACKGROUND

Lymphangiomas in the gastrointestinal tract are extremely rare in adults. As a benign lesion, small intestine lymphangiomas often remain asymptomatic and pose challenges for definitive diagnosis. However, lymphangiomas can give rise to complications such as abdominal pain, bleeding, volvulus, and intussusception. Here, we report a case of jejunal cavernous lymphangioma that presented with intermittent melena and refractory anemia in a male adult.

CASE SUMMARY

A 66-year-old man presented with intermittent melena, fatigue and refractory anemia nine months prior. Esophagogastroduodenoscopy and colonoscopy were performed many times and revealed no apparent bleeding. Conservative management, including transfusion, hemostasis, gastric acid secretion inhibition and symptomatic treatment, was performed, but the lesions tended to recur shortly after surgery. Ultimately, the patient underwent capsule endoscopy, which revealed a more than 10 cm lesion accompanied by active bleeding. After single-balloon enteroscopy and biopsy, a diagnosis of jejunal cavernous lymphangioma was confirmed, and the patient underwent surgical resection. No complications or recurrences were observed postoperatively.

CONCLUSION

Jejunal cavernous lymphangioma should be considered a cause of obscure gastrointestinal bleeding. Capsule endoscopy and single-balloon enteroscopy can facilitate diagnosis. Surgical resection is an effective management method.

Key Words: Key words: Intermittent melena; Refractory anemia; Capsule endoscopy; Single-balloon enteroscopy; Jejunal cavernous lymphangioma; Enterectomy; Case report

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Core Tip: Core Tip: We report a patient with recurrent melena and refractory anemia that was misdiagnosed as obscure gastrointestinal bleeding or intestinal ischemia for 9 months. A definite diagnosis of jejunal cavernous lymphangioma was made through capsule endoscopy, double-balloon enteroscopy, and biopsy. After surgical resection of the lesion, both the melena and anemia resolved.

INTRODUCTION

Lymphangioma is an uncommon benign neoplasm that arises from congenital dysplasia of the lymphatic system and occurs more rarely within the small intestine in adults, accounting for less than 1% of all cases[1, 2]. Small bowel lymphangiomas lack characteristic manifestations and symptoms. The clinical presentations of asymptomatic chronic wasting diseases vary and include weight loss, hypoproteinemia, lymphopenia, hypogammaglobulinemia and iron deficiency anemia; acute abdominalgia, for instance, volvulus; intussusception; and acute intestinal obstruction[1, 3-7].In this report, we present a case of recurrent melena and refractory anemia caused by jejunal cavernous lymphangioma, which was initially misdiagnosed for nine months but then was successfully diagnosed through capsule endoscopy, single-balloon enteroscopy and biopsy at our hospital. Following surgical resection, there has been no recurrence of symptoms.

CASE PRESENTATION

Chief complaints

A 66-year-old man presented with recurrent melena and fatigue for 9 months.

History of present illness

The patient presented with melena approximately of unknown etiology for 3 to 4 times a day and that started nine months earlier, with approximately 100 mL of melena per occurrence, and the melena was accompanied by dull pain below the xiphoid and fatigue, without other significant discomfort. The symptoms resolved spontaneously after a few days. He underwent multiple hospital visits for treatment, during which esophagogastroduodenoscopy and colonoscopy were performed, revealing no evident bleeding lesions. He was diagnosed with moderate to severe anemia. The lowest recorded hemoglobin level was 5.5 g/dL. Following conservative management, including transfusion, hemostasis, and gastric acid secretion inhibition, his symptoms were temporarily alleviated. However, the symptoms recurred after discharge, leading to his presentation at our hospital in March 2023.

History of past illness

The patient had a history of hypertension, which was under control with medical treatment. He denied any history of trauma or surgery.

Personal and family history

No hereditary diseases or history of allergies, alcoholism or epidemiologic linkage were described.

Physical examination

The patient presented with an anemic appearance. The abdomen was soft. No tenderness, peritonitis or palpable mass was observed. No abnormalities were detected during a digital rectal examination. Cardiopulmonary and neurological examinations revealed no abnormalities.

Laboratory examinations

His hemoglobin level was 6.7 g/dL, and his fecal occult blood test was strongly positive at admission. The C-reactive protein level was 11.7 mg/L. His leukocyte count and procalcitonin level were normal. The lymphocyte count and lymphocyte percentage were 0.47 □ 109/L and 14.6%, respectively. The thrombocyte count and coagulation function were also within the normal range. No positive laboratory results were detected for carcinoembryonic antigen, cancer antigen 19-9, tuberculosis antibody, autoimmune disease-related antibodies, immunoglobulin A (IgA), IgG, or IgM. There were no abnormalities in liver or renal function tests.

Imaging examinations

No hemorrhagic foci were detected during esophagogastroduodenoscopy or colonoscopy. Contrast-enhanced computed tomography (CT) of the entire abdomen revealed ischemic changes on the mesenteric side of the distal jejunal and adjacent mesenteric nonenhancing masses (Figure 1 A-C). During capsule endoscopy, distal jejunal mucosal erosion and active bleeding were found (Figure 2). Single-balloon enteroscopy was performed for biopsy. As revealed by hematoxylin and eosin (HE) staining, the biopsy specimens exhibited a significant abundance of buried tubular structures in the mucosal layer (Figure 3). Immunohistochemical analysis revealed negative CD34 expression and positive D2-40 expression in the biopsy specimen.

FINAL DIAGNOSIS

According to the enteroscopy findings and pathological results, a definitive diagnosis of jejunal cavernous lymphangioma was established.

TREATMENT

After positive preoperative preparation, exploratory laparotomy was performed. During intraoperative exploration, edema and thickening of the intestinal wall were observed approximately 2 meters away from the Treitz ligament. The length of the diseased jejunum exceeded 10 cm. Numerous cystic and hemorrhagic masses of varying

sizes were observed on the mesentery, and these lesions were infiltrating into the intestinal lumen (Figure 4 A-B). The patient underwent resection of the diseased jejunum and mesentery, followed by side-to-side jejunal anastomosis. Following a three-day fasting period postsurgery, accompanied by prophylactic anti-infection measures and parenteral nutrition support, the patient started a full-fluid diet and gradually transitioned to a normal diet. No postoperative complications were observed, and the patient was discharged seven days after the surgical procedure.

OUTCOME AND FOLLOW-UP

The anemia resolved, and 4 months after the operation, his hemoglobin level increased to 12.9 g/dL without transfusion. No complications or recurrences were observed during the 6-month follow-up period.

DISCUSSION

Lymphangioma can arise from malformations of the lymphatic system during embryonic development or from acquired factors such as trauma, surgery, inflammation, and parasitic infections[8]. Lymphangioma can be histopathologically classified into three subtypes: cavernous, capillary, and cystic lymphangioma, which may manifest in any part of the body[9, 10]. The preferential localization of the fungus is predominantly in the head, neck, and axilla, encompassing approximately 95% of all cases, especially in young children[11, 12]. However, limited reports on the small intestine, spleen, esophagus, and other organs exist[13]. Intestinal lymphangioma is extremely rare in adults. A mere total of 52 cases have been reported in the English literature since 1961[2, 3, 6, 8, 13-23].

There is no characteristic clinical presentation of lymphangioma. The manifestation may be asymptomatic at times and can occasionally encompass a range of symptoms, including abdominal pain, melena, and acute peritonitis. Hemorrhage, perforation, torsion, intussusception, and rupture are frequent causes of hospital admission[16]. Occult gastrointestinal bleeding or obscure intermittent melena may be the

predominant clinical manifestations, accounting for more than half of the cases (28 out of 52, 53.8%) reported in the English literature. The mechanism of bleeding may be attributed to the obstruction of lymphatic flow, resulting in increased pressure on lymphatic–venous connections and subsequent retrograde blood flow into lymphatic vessels [16]. In our case, the patient presented with undiagnosed intermittent melena, fatigue and refractory anemia for a duration of nine months, which highlights the necessity of considering intestinal lymphangioma as a potential etiology for obscure gastrointestinal bleeding.

The laboratory tests conducted for lymphangiomas lack specificity. In this case, we observed a mild decrease in the lymphocyte count, which has been previously reported in two similar cases[3, 4]. It is speculated that lymphopenia may serve as one of the clinical features associated with lymphangioma. However, the underlying mechanism is unclear. One postulation is that this condition could be due to the excessive loss of lymphatic fluid containing lymphocytes into the intestinal lumen[24].

The use of CT, MRI, and ultrasound, although lacking specificity and having limited accuracy, is advantageous for facilitating the detection of lesions and differential diagnosis. On CT, intestinal lymphangiomas are nonenhanced, well-demarcated and low-density oval masses beneath the submucous membrane[8]. CT scans may yield inconclusive results, with a positive rate of 39.1%[3]. However, it plays a crucial role in the detection of masses that manifest as asymptomatic large abdominal masses or those that give rise to complications, such as perforation, volvulus, and intussusception. MRI scans often reveal a prolonged T1 and T2 signal, a high compression lipid signal and homogeneous/hypodense lesions[22]. Ultrasound can detect multiple cystic anechoic-hypoechoic masses that exhibit thickening of the mucosal and submucosal layers and that display a hypoechoic heterogeneous component[15]. PET/CT is helpful for identifying malignant tumors.

The utilization of capsule endoscopy may represent one of the most efficacious approaches for the diagnosis of lymphangioma. Characteristic endoscopic manifestations include edematous white–yellow mucosa accompanied by hemorrhagic

red spots[14]. Once the typical features are identified, a single or double enteroscopy can be performed to conduct a biopsy and establish a definitive diagnosis. Among the 52 cases reported in the English literature, 40 included a definitive diagnosis established using capsule endoscopy or enteroscopy (76.9%, 40/52), and 42 patients (80.8%, 42/52) were diagnosed since 2010. This finding may be attributed to the widespread clinical utilization of endoscopy in the past decade and indicates the indispensability of capsule endoscopy for diagnosing lymphangioma.

Histopathologic presentation of cavernous lymphangioma revealed large dilated and congested lymphatic lumens expanding to the mucosa of the intestine[6, 23]. D2-40, CD31, VEGFR3 and ERG expression is valuable for demonstrating lymphangioma endothelial cells[4, 25, 26]. The expression of CD34 can serve as a reliable marker for distinguishing lymphangioma tissue from hemangioma tissue, particularly in lymphangioma tissue with hemorrhage, where the dilated vessels are filled with erythrocytes[27]. Immunohistochemical analysis of this patient revealed D2-40 expression and negative CD34 expression, consistent with a diagnosis of cavernous lymphangioma.

Specific interventions are rarely necessary for asymptomatic lymphangiomas, whereas surgical resection remains the established approach for individuals experiencing symptoms.

Recently, compelling evidence has demonstrated the feasibility, efficacy, and safety of endoscopic resection for patients with lymphangiomas smaller than 2 cm[16, 18, 19, 21, 28]. Conservative therapies, such as cryotherapy, laser therapy, and local administration of sclerosing agents, are disfavored[29]. The implementation of meticulous follow-up is crucial, as recurrence rates range from approximately 10% to 27% for completely excised lesions and from 50% to 100% for partially resected lesions[13, 29].

CONCLUSION

Jejunal cavernous lymphangioma, albeit rare, should be considered one of the etiologies of obscure gastrointestinal bleeding. Capsule endoscopy and single-balloon enteroscopy

play pivotal roles in facilitating diagnosis. Surgical resection of the lesion represents a highly effective therapeutic intervention.

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