CASE STUDY: COLONIC LYMPHANGIOMA IN VIETNAMESE CHILDREN

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Abstract

Objectives: We presented a rare case of lymphangioma in the colon with successful surgical removal. ***Methods:*** A case report of a 13-year-old female patient who was brought to Vietnam National Children's Hospital with hematochezia severe normochromic microcytic anemia. Due to nonspecific symptoms, the patient was misdiagnosed with iron deficiency anemia and internal hemorrhoid. After approaching symptoms of lower gastrointestinal bleeding, the patient was done with colonoscopy and abdominal CT scanner, then was diagnosed with a lymphangioma of the sigmoid colon. Surgical resection of the bowel, which contains lymphatic malformation, was successful. ***Conclusion:*** Lower gastrointestinal bleeding and microcytic anemia are solitary colonic lymphangioma symptoms. Their diagnosis is established by colonoscopy and abdominal CT scanner. The main therapeutic option is surgical removal of the lesion.

Keywords: Colon; Lymphangioma; Children; Surgical resection.

INTRODUCTION

Lymphangioma is uncommon in children and presents 5% of benign tumors in childhood [1]. This is a malformation of the lymphatic system involving the cystic, dilated lymphatic vessels (lymphatic malformations). The lesion is frequently detected in the head, neck, and axilla [1, 2] and younger than two years of age. However, it can be observed anywhere with any age [3], and colonic location is predominant in women. The etiology of this disease needs to be more understood.

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There are some hypotheses of mechanism: (a) Lymphatic tissue is isolated during the embryonic without regular connection to the normal lymphatic and venous system; (b) obstruction of the efferent lymphatic vessels causes accumulation of lymph and dilatation of the lymph canals proximal to this obstruction [4]; (c) this malformation can be congenital, related to genetic abnormalities, and combined with Noonan, Turner, or Down syndromes [1]. Hence, the lymphatic blockade may appear after infections or traumas [4]. Clinical manifestations are based on the lymphatic tumor's location and volume. Colonic lymphangioma is extremely rare in children. The beginning of this disease is always asymptomatic. Abdominal pain and bloody stools may be presented in late episodes. Colonoscopy plays an important role in the diagnosis; however, histology provides valuable evidence to confirm it. The treatment is challenged with conservation by sclerotherapy or surgery. Although the symptoms are relieved by conservative treatment, complete resection may be a prioritized choice for this disease [1]. We conducted this study: *To report a Vietnamese child case with colonic lymphangioma and discussed the clinical, imaging features, and treatment approaches.*

CASE REPORT

A 13-year-old female patient whose 43 kilograms of weight had a 6-month history of unexplained anemia that was not improved by changing dietary, iron supplements, and transfusion of 500mL packed red cells in a rural hospital. Four months later, the patient accidentally observed her bloody stool. She was diagnosed with internal hemorrhoid stage III and underwent sclerotherapy in another local hospital. However, the patient still had hematochezia repeatedly after treatment.

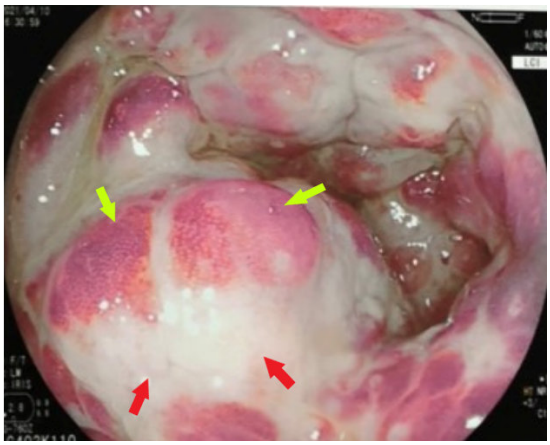

Figure 1. Colonoscopy findings.

(The entire sigmoid colonic mucosa is edematous and congested with small bleeding ulcers.

The yellow arrow indicated sigmoid colonic mucosa, and the red arrows indicated ulcer sites.)

The patient was transferred to a big clinic in Hanoi. She underwent an extensive evaluation for hematochezia and severe normochromic microcytic anemia with iron deficiency, including upper and lower gastrointestinal endoscopy and endoscopic ultrasound. The result was strange; the entire sigmoid colonic mucosa was edematous and congested with small bleeding ulcers. The patient was diagnosed with colitis, suspected Crohn, which was not proportional to her anemia status. That was the reason he was transferred to our department on April 10th, 2021.

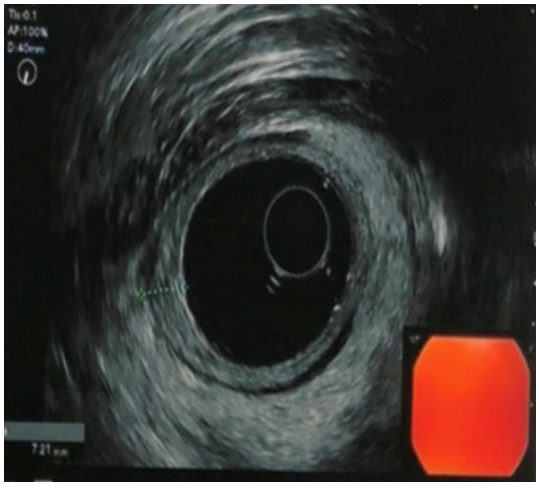

Figure 2. Endoscopic ultrasound finding.

(Abnormal thickening of submucosa in sigma colon around lesions.

There is vascular proliferation around the lesion site in the Doppler ultrasound.)

The only finding of note on examination was conjunctival pallor. Other physical examinations were no remarkable: Normal vital signs, no jaundice, soft abdomen, and yellow stool, no anus fissure in rectal examination. Laboratory results showed a severe anemia status with a low hemoglobin level of 5.3 g/dL (reference range, 11.5 - 15.0 g/dL), a low hematocrit level of 20.9% (reference range, 35 - 40%), a low mean corpuscular volume of 59fL (reference range, 77 - 95fL), low level of ferritin 15 ng/mL (reference range, 7 - 140 ng/mL), and a low serum iron of 2.5 µmol/L (reference range, 50 - 120 µmol/L). The other laboratory data, such as liver and renal function, were unremarkable. The patient was transfused with 500mL packed red cell to archive 9.8 g/dl.

Based on previous endoscopic results, the patient was diagnosed with severe anemia due to Crohn's disease. Seven days after the trial treatment of rectal mesalamine, her clinical status showed no improvement.

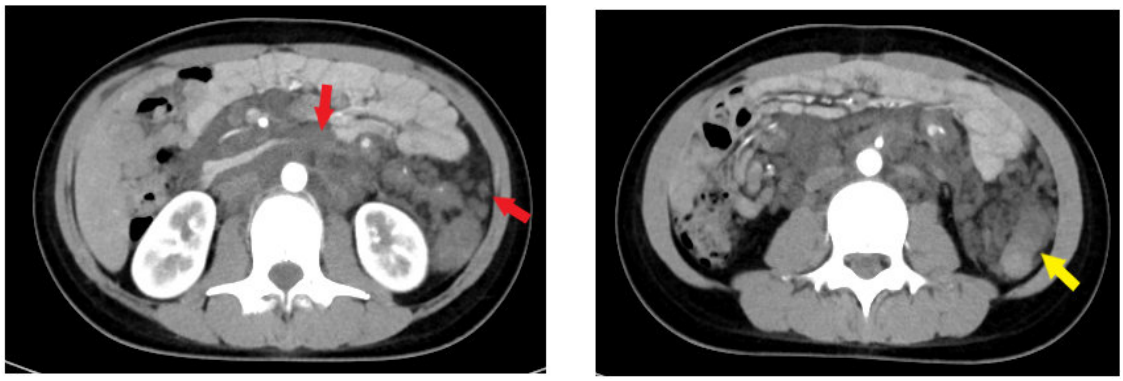

Figure 3. Abdominal CT findings.

(As the red arrows indicated, diffuse infiltrate without contrast enhancement in the mesentery.

Thickening of left colon wall as yellow arrow indicated.)

Abdominal CT with IV contrast was indicated to find other etiologies. Results show (1) thickening of the bowel wall in the left colon and sigma colon (2) diffuse infiltrate surrounding the pancreas, mesentery, and left colon gutter. These findings suggest abdominal lymphatic malformation. Secondary colon endoscopy demonstrated multiple submucosa lesions covered with normal mucosa in the sigma colon wall. These lesions were soft and could change shape when compressed by the endoscopic tool.

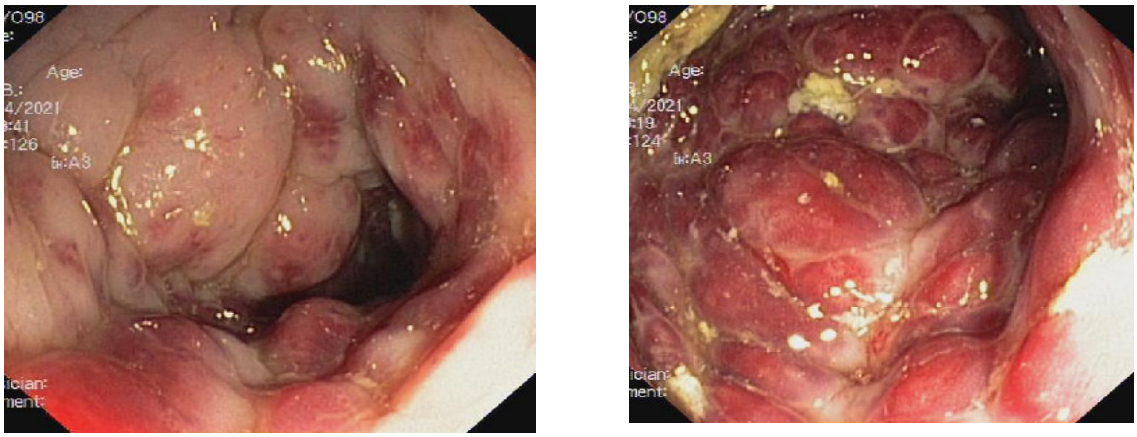

Figure 4. Colonoscopy findings for the second time.

(Multiple red mass tissues protruded into the lumen of the sigma colon, and there was no sign of inflammation.

The overlying mucosa was congested, which may indicate bleeding inside the tumor.)

Based on the colonoscopy and abdominal CT findings, the patient was diagnosed with lymphangiomatosis of the sigmoid colon and mesentery. Due to large tumors and diffuse lesions spread throughout the abdominal capacity, surgery was indicated for the patient. In laparoscopy, lymphatic malformation spreads throughout the mesentery from the descending colon to the rectal; the affected colon has a narrow lumen; grape-shaped cystic lymphangioma surrounds left ureters in retroperitoneal. After removing the adhesive and freeing the colonic mesentery, the tumor was profoundly bleeding, which challenged surgeons. The patient underwent left hemicolectomy, colostomy, and lymph node dissection. This operation lasted for 8 hours. The histopathological result was consistent with lymphangiomatosis *(Figure 3)*. The patient was treated with antibiotics and discharged after ten days of operation.

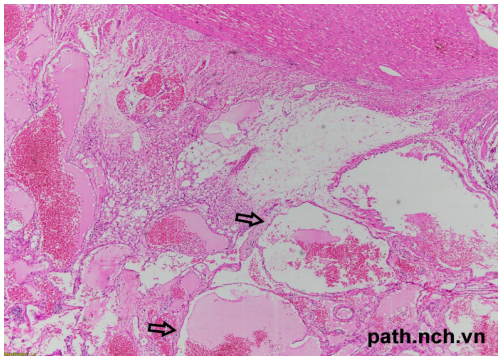

Figure 5. Histopathologic findings of resected tissue revealed

(Multiple thin-walled cystic lesions were covered with normal colonic mucosa.

The fluid in the cysts was crystal, with red blood cells indicating hemorrhage inside the tumor.)

DISCUSSION

Lymphangiomas are benign malformations characterized by abnormal dilatation and proliferation of lymphatic spaces. They are usually detected in the head, neck, and axillary areas, accounting for 50 - 75% of all cases, while only 5% occur in the abdomen. Intra-abdominal lymphangiomas are frequently located in the mesentery, greater omentum, and retroperitoneum but rarely in the colon. Matsuda et al. reviewed 279 cases of colonic lymphangioma for 70 years reported in the Japanese literature, with only two cases in children [5]. After 20 years of working in our department, this is the first case we diagnosed by colonoscopy.

Although it is generally agreed that lymphangiomas are hamartomatous or malformation lesions, most of which are thought to be present at birth. Our patient had the disease onset at 12 years of age when she was at puberty. This may be due to endogenous estrogens are hypothesized to play a

role in the enlargement or growth of lymphangiomas [5].

Difficulties in diagnosing colonic lymphangiomas are due to the fact that clinical signs and symptoms are highly variable and depend on the size and location of the lesion. Colonic lymphangiomas are usually present as submucosal polypoid lesions without any symptomatic presentation. Some patients may experience episodic constipation with vague abdominal distress, diarrhea, or other nonspecific symptoms [6]. Hemorrhage or anemia [7] and protein-losing enteropathy [8] are always associated with surface ulceration or erosion. In acute abdomen obstruction of the volvulus, intussusceptions may occur when a mass is sufficiently enlarged [9]. The clinical findings in our case are nonspecific, with refractory microcytic anemia. Prolonged bloody stool was a late finding that led to her long misdiagnosis. These symptoms suggested lower gastrointestinal bleeding, and vascular malformation is a rare cause.

Colonoscopy has a high value in the definitive diagnosis of colonic lymphangioma. Lymphangioma characteristics on endoscopy are soft, round cystic masses in the submucosa of the colon, which a forceps may push. These tumors have translucent surfaces covered with normal colon mucosa. Our case also has these features, but the overlying mucosa was congested with dark red. This finding may suggest bleeding complications inside a tumor.

Abdominal CT is a valuable imaging tool for diagnosing abdominal lymphangioma. Moreover, CT imaging may provide detailed information about tumor size, location, and lesion extent, which is critically important to decide the treatment plan. Imaging features of abdominal lymphangioma typically are low attenuation lesions in the colonic wall and mesentery. Cystic wall and septa are normally enhanced in IV contrast-enhanced CT. In our case, the lesion in CT imaging is also low attenuation, spreading in the descending colon and mesentery. Nevertheless, when administering intravenous contrast, these tumors were not enhanced in the wall.

Colonic lymphangiomas are submucosa lesions, and endoscopic biopsies are usually not deep enough to approach these masses. Besides, massive bleeding may occur, which is seriously threatened. In general, the pathological examination is not commended.

Histopathological findings after the operation I are the gold standard for diagnosis. Our patient had postoperative histopathological findings consistent with typical lymphangiomas.

Regarding treatment, colonic lymphangioma management depends on the symptoms, size, growth speed, and location. About 10% can be relieved naturally [10]. Kochman et al. recommended regular follow-ups rather than intervention for asymptomatic patients. Treatment methods include surgery, endoscopic resection, and drug injection. Complete surgical resection is the first choice for large lymphangiomas accompanied by complications such as bowel obstruction, bleeding, volvulus, or intussusception [11, 12]. Laparotomy is considered a principal treatment in this field, but endoscopic therapy has been developed recently and has shown efficacy. Lymphangioma is a benign disease; therefore, endoscopic resection may be sufficient, and excessive invasive treatment should be avoided. Endoscopic intervention can be reserved for gastrointestinal tumors with a maximum diameter of 2.5cm or smaller [5, 11]. Matsuda et al. [5] reviewed 279 Japanese cases of colorectal lymphangiomas. Of them, 104 patients (37.4%) successfully underwent endoscopic resection; however, there is no information about long-term outcomes. In 2017, Kanithi et al. presented a case of a 69-year-old patient without symptoms, who had a colonic lymphangioma measuring 5cm in diameter. It was successfully resected by end-loop endoscopic with no complications. Endoscopic therapy is a less invasive method with lower costs.

Based on the volume and location of lymphangioma, the endoscopic intervention is also a new direction and can be replaced for surgical evaluation. Until now, however, the gold standard of treatment is complete surgical resection. The literature does not show endoscopic sclerotherapy (agents such as Bleomycin, sodium tetradecyl sulfate, or OK-432). Non-invasive therapy, including steroids, fibrin glue, or Ethibloc, has not been established as superior to surgery. Hence, the risk hazard of intra-abdominal leakage and the extended lesion can appear and lack gold standard criteria for diagnosis. Our patient had a large colonic lymphangioma associated with mesenteric lesions and bleeding complications, so the best treatment is surgical resection. She underwent

a successful operation and was confirmed with a diagnosis of colonic lymphangioma in operation and histology. In this case, surgical intervention with histology is the best choice for diagnosis and treatment.

CONCLUSION

Lymphangioma is an uncommon benign tumor in children. Lower gastrointestinal bleeding and microcytic anemia are symptoms of colonic lymphangioma. Their diagnosis is established by colonoscopy and abdominal CT scanner. The main therapeutic option is surgical removal of the lesion.

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