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Emergency presentation of cystic lymphangioma of the colon: A case report and literature review

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ABSTRACT

INTRODUCTION: Colonic lymphangioma is an unusual benign malformation. The clinical presentation of lymphangiomas vary from incidental discovery on imaging to presenting with acute abdomen.**PRESENTATION OF CASE:** We present the case of a 73-year-old male, undergoing surgery due to acute abdomen associated with severe anemia, in whose case a lymphangioma of the cecum was recognised only in the postoperative histopathological examination.**DISCUSSION:** The management of colonic lymphangioma depends on the individual situation; close surveillance or endoscopic therapy may be appropriate for asymptomatic lesions smaller than 2.5 cm in diameter. Surgical intervention can be considered for larger lesions or in patients who develop complications.**CONCLUSION:** The interest in our case lies in the relatively rare diagnosis of colon lymphangioma and how the cardinal sign was anemia, which may be due to the serosanguineous cystic contents of lymphangiomatosis from the internal bleeding and can cause fatal complications that require emergency surgery.© 2016 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Lymphangiomas constitute a group of very rare diseases with a rate of hospitalization of 1/250,000 to 1/20,000. although being benign lesions, their complications may turn them into a life-threatening condition. They usually occur in children (90%) and they are generally found in the head and neck regions during the first few years of life. Other locations such as the abdominal or mediastinal cavity are rare, accounting for approximately 5% of all lymphangiomas. Complications, like intestinal obstruction and persistent bleeding, are the cause of exploratory laparotomy. Final diagnosis requires histological examination. We present the case of a 73-year-old male, undergoing surgery due to acute abdomen associated with severe anemia, in whose case a lymphangioma of the cecum was recognised only in the postoperative histopathological examination.

2. Case presentation

A 73-year-old male presented to the emergency department complaining of acute onset of diffuse abdominal pain and nau-

sea that began approximately 48 h earlier and fatigue that began one month before, associated with several episodes of melena. He denied any recent fever, chills, hemoptysis, hematochezia, or changes in bowel habits. He had no history of trauma, no history of surgery, and no other meaningful history. On presentation to the emergency room, he was pale with heart rate of 101 bpm, blood pressure of 140/60 mmHg, and respiratory rate of 14/min. Physical examination revealed abdominal distension with notable rebound tenderness and guarding. Rectal examination revealed normal sphincter tone, no palpable masses, and black tarry stools. Other features were unremarkable. Laboratory values upon admission showed 4 g/dL haemoglobin, 11.1% hematocrit, 124.7 fL mean corpuscular volume (normal = 83–97), 44.9 pg mean corpuscular haemoglobin (normal = 27–33), 36.0 g/dl mean corpuscular haemoglobin concentration (normal = 32–36), 12 ug/dL iron (normal = 65–157), and 3.59 ng/mL ferritin (normal = 16.4–323). The remaining laboratory data including electrolytes, urinalysis and coagulation factors were unremarkable. An abdominal CT scan documented a 42 mm Ø, low-density, non-enhancing cystic mass located at the ascending colon associated with free abdominal fluid, moderate pericardial and bilateral pleural effusion. (Fig. 1) No air-fluid levels, normal liver, spleen, pancreas, kidneys and adrenal glands. The patient was kept under close observation, but there was a gradual reduction in haemoglobin levels despite the transfusion of 4 packet red blood cell. Due to haemodynamic impairment

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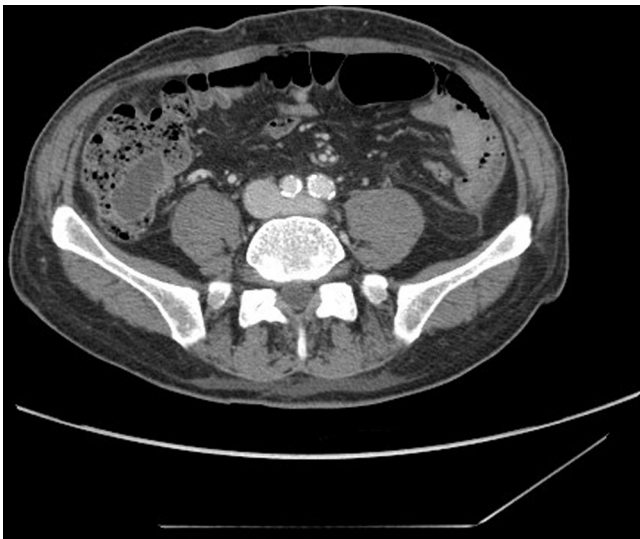


Fig. 1. Enhanced abdominal CT axial image shows a 42 mm Ø, low-density, non-enhancing cystic mass located at the ascending colon.

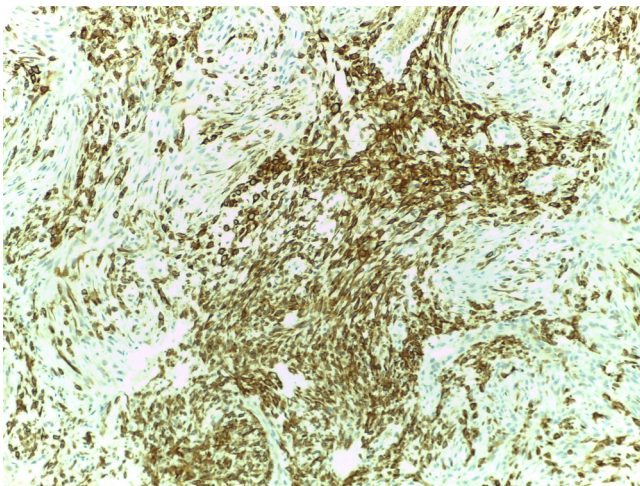


Fig. 2. Multiple lymphatic channels lined by benign-appearing endothelial cell (note the absence of red blood cells).

he underwent an emergency exploratory laparotomy. At laparotomy, a conspicuous abdominal clear not-hematic effusion was found with cecal mass measuring approximately 2,5 × 2 × 1,5 cm. Mass content was haematic. Upon these findings a right hemicolectomy was performed to provide definitive diagnosis and treatment. The postoperative course was complicated by a worsening of pericardial effusion, with signs of unstable hemodynamic and increase of the bilateral pleural effusion. The patient was then transferred to the cardiac ICU where underwent pericardiocentesis with drainage of 800 ml of serous fluid. The patient was discharged on the eighteenth postoperative day. The pathological examination documented an intraluminal pedunculated lesion of 5.5 cm, composed of multiple lymphatic channels lined by benign-appearing endothelial cell (note the absence of red blood cells) separated from each other by fibrous connective tissue septae with aggregate of lymphocytes. Based on these findings, colon lymphangioma was diagnosed. (Figs. 2 and 3). The patient is currently disease-free, with maintenance of normal haemoglobin on subsequent 6 months of follow-up.

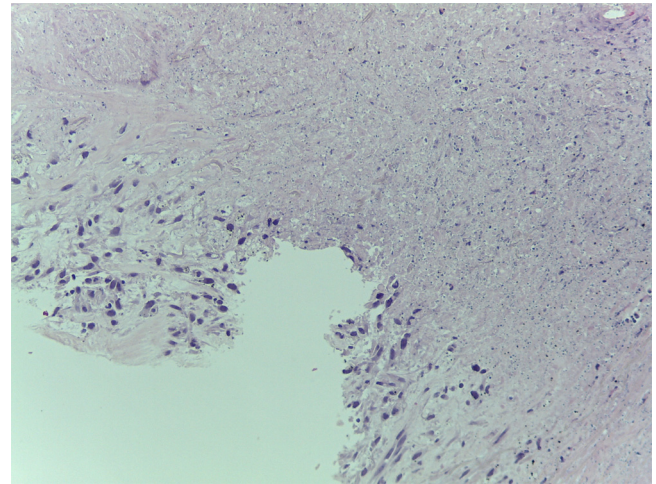


Fig. 3. Fibrous connective tissue septae with aggregate of lymphocytes.

3. Discussion

Lymphangioma of the colon was first described by Chisholm and Hillkowitz in 1932 [1], and is a submucosal tumour covered with normal mucosa. It occurs mainly in children [2]; approximately 80–90% is diagnosed within the first few years of life and adult cases are rare [3]. According to various reports, the age distribution was 1–83 years with the most frequent age being 60 years, and the incidence was higher in males with a sex ratio of 2–2.5:1 in Japan [4]. Lymphangiomas can occur anywhere in the body and only rarely affects the intestinal tract 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 and tend to be located in the right half of the colon [5]. It has been reported more frequently nowadays because of the widespread use of endoscopy. Specific etiology of this tumor is unknown. In a Japanese review of 279 cases Matsuda T. et al. [4] suggested that the occurrence of these lesions is connected with excessive proliferation of the endothelial cells of the lymphatic vessels walls. These vessels create vesicular or nodular lesions when growing, or when getting larger they cause deformation of the regions in which they occur. The literature presents the data indicating that some of the intestinal inflammations, surgical procedures performed and radiation may trigger the occurrence of these tumours. Macroscopically, colonic lymphangiomas are classified into one of the following three types: simple (capillary), cavernous and cystic [6]. No malignant case has been reported. Although cases complicated by colorectal carcinoma have been reported, the complications incidentally occurred, and the causal relationship was unclear. The cystic type is the most frequently reported. Cystic lymphangiomas could be yellow, greyish, or yellow-pink in colour, and they often appear as multiple cysts or spongy masses with cavities containing watery or milky fluids. Cystic lymphangiomas may be classified into microcystic, macrocystic and, mixed subtypes according to the cyst. The clinical symptoms of a mesenteric lymphangioma are non-specific and include abdominal pain, vomiting and constipation. Lower GI bleeding, obstruction, intussusception and protein-losing enteropathy are rare complications [7–12]. The cardinal sign of our case was anemia, which may be due to the serosanguinous cystic contents of lymphangiomatosis from the internal bleeding that in our case led to an impairment of the haemodynamic status. Making a specific diagnosis is practically impossible on clinical grounds. Imaging thus plays an indispensable role. The majority of lymphangiomas are discovered incidentally on imaging for the investigation of unrelated clinical indications [13–15]. The differential diagnosis includes a wide range of cys-

tic intraabdominal lesions, ranging from pancreatic pseudocysts to abdominal tuberculosis, hydatid disease, or malignancies such as mucinous carcinomatosis. Romeo V. et al. [13] suggest that the appropriate diagnostic imaging protocol in patients with cystic lymphangioma should initially include the US study, followed by a MRI scan with contrast administration. US is considered as being the first level of imaging investigation for a suspected mass suggestive of cystic lymphangioma because of its non-invasiveness, low cost, and non-use of ionizing radiation, in order to identify the lesion and to define its structural cystic type characteristics as well as the size. However, this technique needs to be integrated with CT or MRI scans because of its non-panoramic view and also for obtaining additional information such as structural feature, internal and peripheral contrast enhancement, as well as loco-regional lesion spread. Although less available than CT, especially in an emergency setting, MRI provides a better preoperative differentiation from other cyst-like masses. In an emergency setting CT scan play a central role in the diagnosis. On CT, these masses show densitometric characteristics of the fluid type, regular margins, and only a peripheral contrast enhancement. The value of imaging in cystic lymphangiomas is to exclude malignancy and to offer the exact anatomic location of the tumor before surgery. US and MRI may also be useful in the follow-up of patients who refuse surgical resection or in whom surgery is contraindicated or postponed, as well as to early detect a possible disease relapse. Even though these tumours are benign, they may often lead, as in our experience, to life-threatening conditions, such as infection, volvulus, obstruction or bleeding into the lumen of a cyst that are usually very difficult to manage. Various forms of treatment have been described [16–20]. Pedunculated or semipedunculated colonic lymphangioma can be managed by endoscopic polypectomy, with colonic perforation being a well-known complication. Karasawa et al. [20] reported that polypectomy as diagnostic treatment is applicable for 2-cm or smaller pedunculated and semipendunculated types, and polypectomy is applicable after lymph drainage by endoscopic puncture for larger and sessile-type tumors, because lymphangioma is benign, and no malignant case has been reported. Other authors [21] have developed a technique of deroofing the lymphangioma lesion with cyst drainage. Many cases of lymphangioma larger than 3 cm were surgically resected in Japan [4]. Wang et al. [22] reported a case of laparoscopy-assisted resection of colon lymphangioma, in which low-invasive surgery was performed because differentiation from malignant diseases was difficult due to a large tumor size. In our case, recurrent drop in haemoglobin with haemodynamic impairment and CT appearance that could not rule out the possibility of malignant tumor guided our choice for surgical resection. According to the literature review we can say that surgical resection is a valid therapeutic option only when a preoperative diagnosis could not be performed and complication such as bleeding occurs.

4. Conclusion

Our report describes the case of a patient with persistent drop in haemoglobin due to an internal bleeding of cystic lymphangioma of the colon that led to hemodynamic impairment with no characteristic CT findings. Thus we decided that a surgical resection was the most effective treatment, and the case was associated with a good prognosis. Despite its low frequency, this disease should be considered because colon lymphangioma could be the cause of gastrointestinal blood loss.

Competing interests

The authors declare no potential financial conflict of interest related to this manuscript.

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Ethical approval

The ethical approval was obtained from the San Filippo Neri hospital's Ethics Committee.

Consent

Patient consent: Obtained.

Author contribution

Study Conception and Design: Fransvea, Cortese, Saputelli
Acquisition of Data: Lepre Analysis and interpretation of data: Fransvea, Cortese Drafting of Manuscript: Cortese, Saputelli
Revision: Giofrè, Sganga Costa.

Guarantor

The Manuscript has been seen and approved by all authors.

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