Colonic mesenteric lymphatic malformation presenting as an intraabdominal abscess in an infant: A case report

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ABSTRACT

INTRODUCTION: Lymphatic malformations are low-flow vascular malformations most commonly located in the head and neck; isolated intraabdominal involvement is rare.

PRESENTATION OF CASE: An 8-month-old previously healthy male presented with a 9-day history of fevers. On examination, right-sided abdominal tenderness was noted. Ultrasound revealed a large heterogeneous mass, and CT scan revealed a rim-enhancing cystic mass adjacent to the right colon. Laboratory investigation including blood cultures was normal. His fever resolved with broad-spectrum antibiotics. Diagnostic laparoscopy revealed a large, firm mass arising from the mesentery of the right colon. An open right hemicolectomy with ileocolic anastomosis was performed. The infant tolerated the procedure well, and he was discharged home on postoperative day four, pathologic examination identified a mesenteric lymphatic malformation with secondary abscess formation.

CONCLUSION: This atypical presentation of an uncommon entity was instructive in several ways, particularly illustrating the diagnostic pitfalls that can be introduced by superinfection.

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1. Introduction

A lymphatic malformation is a type of low-flow vascular malformation that can occur in isolation or as part of a systemic disease. Lymphatic malformations are present at birth and become more apparent as an infant grows and the lesion enlarges in size. They are typically located in the head and neck region, sometimes in the axilla and chest wall [1], and rarely intra-abdominally [2]. Given their heterogeneous presentation and overall rare occurrence, a true prevalence estimate is elusive. Here we present the diagnostic approach and treatment, as well as lessons learned from an interesting case of an intra-abdominal colonic lymphatic malformation complicated by abscess formation. This work is reported in line with the SCARE criteria [3].

2. Case presentation

An 8-month-old male, ex-full-term, presented to the Emergency Department after referral from his primary care physician with a nine-day history of high grade fevers. Family members denied associated symptoms, particularly vomiting, diarrhea, rashes, and seizures. There was no sick contact or travel history. On physical examination, he was noted to have very dry and red lips. On evaluation, temperature was 36.5 °C; heart rate was 140 beats per minute; other vital signs were within normal limits for age. Laboratory findings included: hemoglobin 10.4 g/dL, mean corpuscular volume 81 fl, white blood cell count 31,500/mm³, platelets 618,000/mm³, C-reactive protein 21.53 mg/L, sodium 133 mmol/L, chloride 98 mmol/L, and a normal urinalysis. Blood cultures had been drawn at the primary care physician’s office following by initiation of intravenous ceftriaxone; they subsequently showed no growth. A chest radiograph was normal.

With an initial diagnostic suspicion of Kawasaki disease, the child was admitted to our Rheumatology service. Echocardiogram was normal and showed no aneurysms. Further detailed physical examination revealed mild right abdominal pain. An ultrasound of the abdomen revealed a large heterogeneous right-sided intraabdominal mass, concerning for neoplasm versus abscess. Further laboratory evaluations, including serum alpha-fetoprotein and β-human chorionic gonadotropin, urine homovanillic acid, vanillylmandelic acid, liver function tests and coagulation profile were normal. A computed tomography (CT) scan revealed a rim-enhancing cystic mass (Fig. 1), thought more likely to be abscess or superinfected duplication cyst, and less likely malignancy. Given his ongoing fevers, his antibiotic regimen was changed to piperacillin and tazobactam in anticipation of eventual abdominal exploration, ideally once fevers or inflammatory markers improved. Over the course of a week, his fevers resolved. Diagnostic laparoscopy revealed a large, firm mass emanating from the mesen-

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tery of the right colon (Fig. 2); given its size and relative immobility, an open laparotomy was then pursued through a limited periumbilical midline incision. Resection was via a right hemicolectomy with side-to-side, functional end-to-end, stapled ileocolonic anastomosis. The mass and the right colon were densely adherent to the retroperitoneum (Fig. 3, video), but no adjacent organs were involved.

Gross examination of the resected specimen showed a 5-cm mesenteric cyst filled with viscid yellow material. The overlying mucosa was unremarkable. A representative section was frozen at the time of surgery for intraoperative consultation; this fragment showed a neutrophilic abscess (Fig. 4A). Microscopic examination of permanent sections confirmed the extensive intramural abscess formation and additionally showed uninflamed lymphatic malformation. The latter was characterized by aggregates of markedly dilated lymphatic channels, some lined with lymphoid aggregates and variably well-developed smooth muscle coats. Excess numbers of small lymphatic channels were also present, along with diffuse fibrosis (Fig. 4B and C). Gram-positive rods were highlighted by a Brown-Brenn stain in the areas of abscess; microbiologic cultures from this area showed no growth. The lymphatic endothelium was highlighted by a D2-40 (podoplanin) immunostain (Fig. 4D). Margins of resection were grossly uninvolved. The child tolerated the procedure well. His recovery was uneventful, and at last follow-up, eight months postoperatively, he was entirely well.
3. Discussion

We present the case of a lymphatic malformation emanating from the right colonic mesentery in an 8-month-old male infant. The observations from the case include: (a) The lymphatic malformation likely predisposed the patient to infection at this site. (b) This represents an atypical presentation of a lymphatic malformation. (c) Physical clues to intraabdominal involvement in an infant may be subtle. (d) In the setting of intraoperative frozen section, superinfection may represent a diagnostic pitfall attributable to incomplete sampling. (e) The differential diagnoses that are important to consider in this scenario are enteric duplication cyst, tumor, or abscess.

The incidence of intraabdominal lymphatic malformations has been reported to be around 4 per 100,000 admissions [4]. There appears to be slight male predominance with a male to female ratio of 3.2:1, and these lesions have been seen in patients between the ages of 3 months and 13 years (mean 5 years). The most common reported location for intraabdominal lymphatic malformations is the small bowel mesentery (60%), followed by the mesocolon (24%) as in our case, and the retroperitoneum (14.5%) [5]. Cystic lymphatic malformations are classified as macrocystic, microcystic, or mixed cystic [6]. The symptoms of an abdominal lymphatic malformation can range from asymptomatic, to abdominal distention, pain, fever, constipation, nausea and vomiting. Physical examination findings include fever, localized abdominal fullness or distention, and a palpable abdominal mass with or without signs of obstruction. The complications of such a malformation can include abscess formation (as in our patient), intestinal obstruction, volvulus, bleeding into the cyst resulting in a hematoma formation, and/or rupture.

Diagnosis is usually confirmed on radiologic studies. A plain X-ray of the abdomen may reveal lateral displacement of bowel loops or may show signs of bowel obstruction. Ultrasound and CT scan further define the anatomic relations for this condition [7]. An ultrasound will reveal a hypoechogenic, often multilocular, cystic structure. A CT can further characterise the size of the cyst, the content of the cyst, location, relation to surrounding structures and patterns of enhancement with intravenous contrast.

The definitive treatment of an intraabdominal lymphatic malformation is open or laparoscopic [9,10] excision of the cystic structure, with complete excision associated with a low recurrence rate. Though in general recurrence rates are generally believed to be low following gross total resection, the best method to follow these patients is not well established; hence one must remain vigilant of the possibility and perform a thorough physical exam at each encounter with the patient’s pediatrician. Other treatment options include simple deroofing of the cyst, excision of the cyst along with resection of bowel, and sclerotherapy using doxycycline [8]. Intraoperative frozen section analysis can help guide surgery, but as illustrated in our case, secondary abscess formation may result in incomplete characterization of the pathologic process. Resection of bowel may be necessary in cases where the mesentery is involved.

4. Conclusion

To summarize, this 8-month-old male with high grade fevers and right-sided abdominal tenderness had imaging findings suggestive of an abscess or a duplication cyst. Laparoscopy revealed a colonic mesenteric mass. Right hemicolectomy was undertaken and pathologic examination revealed a mesenteric lymphatic mal-
formation complicated by abscess formation. We found the atypical presentation of this uncommon pediatric condition to be educational, illustrating some potential diagnostic pitfalls associated with superinfection of a lymphatic malformation.

**Conflicts of interest**

No conflicts of interest to report from any of the authors.

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**Ethical approval**

Discussed with our IRB and no IRB approval required to proceed with publication.

**Consent**

Parental consent has been obtained.

**Author contribution**

S. Gonakoti, B. Zendejas, S. Vargas, C. Chen all contributed equally to the study concept, design, data collection, interpretation and writing of the manuscript.

**Guarantor**

Dr. Chen is the guarantor.

**Appendix A. Supplementary data**

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.jiscr.2017.07.055.

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