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## Subserous lymphangioma of the sigmoid colon: an uncommon cause of acute abdomen in pediatric patients

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### ABSTRACT

Lymphangioma is a rare, benign lesion derived from a malformation of the lymphatic system, which is more frequently found in the head, neck, and axilla. However, it may be present anywhere in the body, and the diagnosis involves adults as children with some distinct clinical features among them. In pediatric patients, abdominal cystic lymphangioma occurs mostly in the mesentery presenting abdominal pain, intestinal obstruction, or, more rarely, hemorrhage. The authors report the case of a child with a short-course history of fever, abdominal pain, and constipation. The physical examination disclosed the presence of an abdominal mass and signs of peritoneal irritation. Imaging was consistent with a cystic lesion compressing the sigmoid colon and laterally displacing the remaining loops. Exploratory laparotomy was undertaken, and a sigmoidectomy, followed by Hartman's colostomy, was performed. Histological examination revealed the nature of the lesion as a cystic lymphangioma. The authors highlight the clinical features of this entity and call attention to this disease in the differential diagnosis of acute abdomen or abdominal pain, mainly in pediatric patients.

### Keywords

Lymphangioma, Cystic; Colon, Sigmoid; Abdomen, Acute, Surgical Procedure, Operative.

### CASE REPORT

An 11-month-old girl was brought to the emergency facility with a 7-day history of fever (maximum 38 °C), abdominal pain, irritability, and a 3-day-history of constipation. On clinical examination, the child was cranky, complaining of severe pain in

the lower left abdominal quadrant and pelvis where a mobile cystic mass was easily palpable, and signs of peritonitis were present.

The plain abdominal x-ray showed a distended large bowel and a collapsed distal colon, which are

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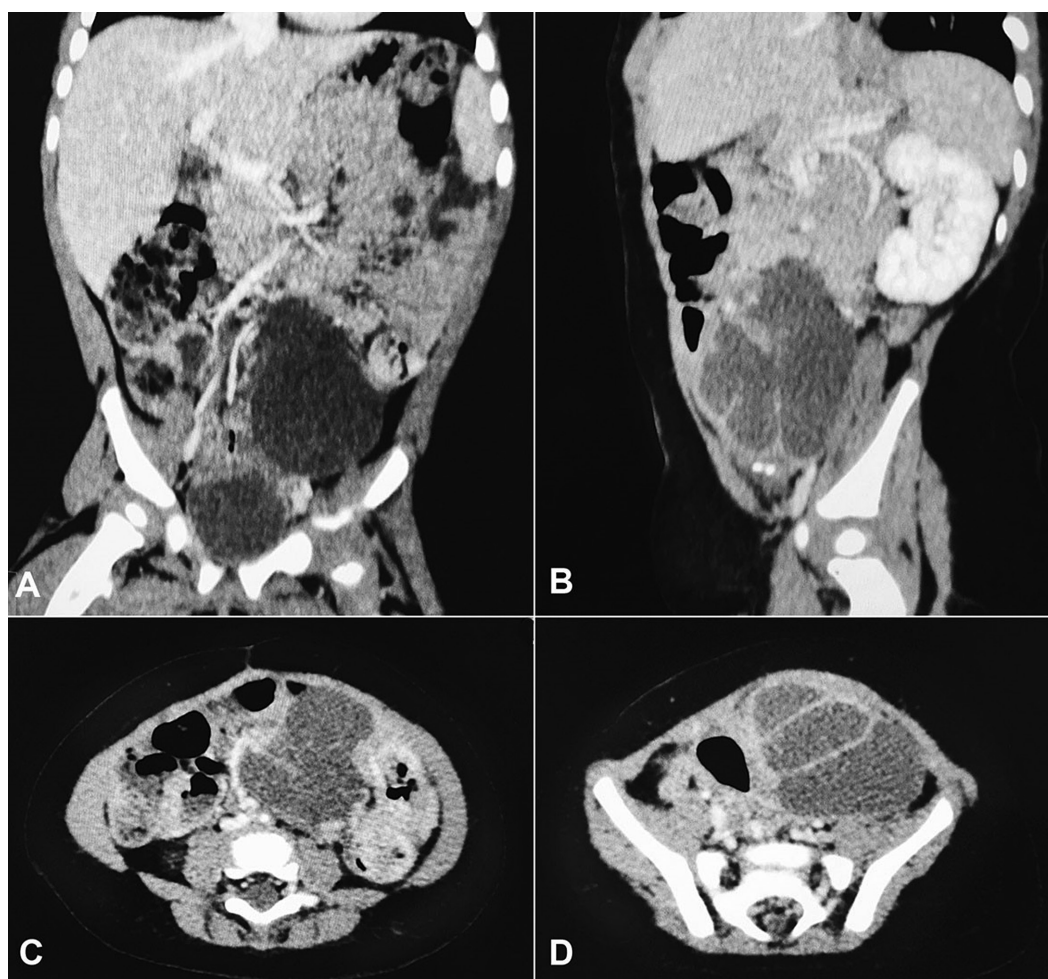
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consistent with a large bowel obstruction. Therefore, a glycerin enema was performed to rule out fecal impaction. An abdominal ultrasound showed a pelvic mass with a liquid content and possible septations, which were interpreted as possible grouped intestinal loops or a cystic lesion. The abdominal computed tomography (CT) diagnosed a pelvic/hypogastric, expansive, loculated cystic lesion with lobulated contour extending to the mesogastrium, measuring  $5.5 \times 4.1 \times 5.2$  cm, with parietal enhancement after contrast infusion, displacing the intestinal loops to the right side, and compressing the sigmoid colon (Figure 1).

The blood cell count revealed hemoglobin of 10.9 g/dL (reference value [RV]: 10.5-13.5 g/dL), leukocytes of  $17,020/\text{mm}^3$  (RV:  $6000\text{--}17,500/\text{mm}^3$ ), and C-reactive protein of 117 mg/L (RV:  $< 5$  mg/L). Platelet count, renal function, and electrolytes were

within the normal range. Two sets of blood cultures were sampled with negative results (posteriorly); however, ceftriaxone and metronidazole were prescribed. Acute abdomen, due to a pelvic abscess (appendicitis, Meckel's diverticulitis) or gynecological complication like ovarian torsion, was the clinical suspicion and therefore the patient underwent an exploratory laparotomy. Surgical findings included: (i) a loculated cystic tumor in the mesosigmoid seeming to infiltrate a friable portion of the sigmoid, which drained milky fluid resembling pus; (ii) serous ascites; and (iii) enlarged mesosigmoid lymph nodes. Some of the cystic areas were translucent while others were turbid, which is the reason why an intestinal perforation was suspected. Sigmoidectomy (Figure 2A) followed by a terminal colostomy by Hartmann's technique was performed. Culture of the peritoneal fluid was negative.



**Figure 1.** Multidetector computed tomography of the abdomen. **A** - Coronal reformation showing a cystic lesion over the urinary bladder and displacing mesenteric vessels to the right; **B** - Sagittal reformation depicting tiny parietal calcifications on the tumor wall; **C** and **D** - Axial images showing an abdominal cystic lesion displacing intestinal loops and mesenteric vessels in C, and thin septa within the lesion in D.

A 7.6 cm length of formalin-fixed large bowel segment with preserved mucosa and a whitish lobulated tumor measuring 4.0 × 3.0 cm, which was adhered to the mesenteric face, represented the surgical specimen (Figure 2B). At the cut surface, translucent cysts (measuring up to 2.5 cm) and a solid area of 1.5 × 1.3 cm represented the tumor. Most cysts contained watery fluid except one, which exhibited a milky content. Tiny lymph nodes were also identified surrounding the tumor.

The histology showed a subserous multiloculated cystic lesion partially lined by flat cells without atypia (consistent with endothelium), and cysts presenting erosion/ulceration surrounded by granulation tissue, mixed inflammatory infiltrate, and lymphoid aggregates. The intestine wall was free of invasion. Acute serositis and lymphoid hyperplasia in adjacent lymph nodes was present. The adjacent mucosa showed foci of microabscess within the crypts. The immunohistochemical profile was consistent with the diagnosis of lymphangioma (CD31<sup>[JC/70A clone]</sup> positive, CD34<sup>[QBEnd10]</sup> partially positive and AE1/AE3 negative) (Figure 3).

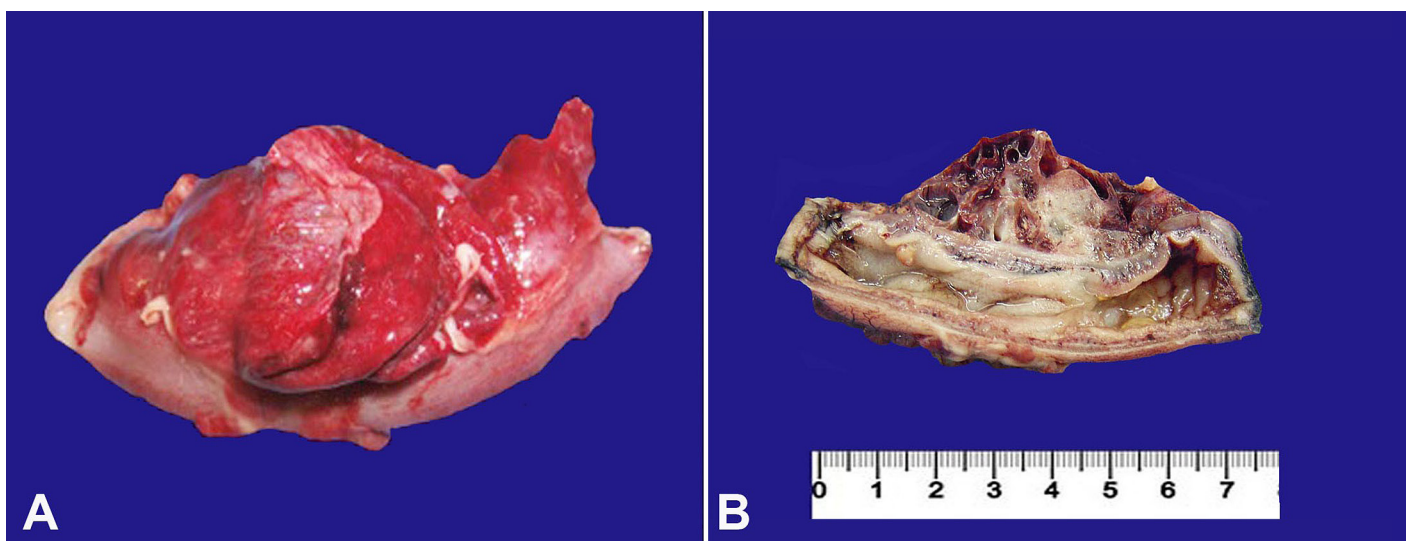
The outcome was favorable and the colostomy was functioning 2 days after surgery. The patient completed a 10-day course of ceftriaxone and metronidazole and was discharged 11 days after admission. She was referred to an outpatient clinic.

## DISCUSSION

Abdominal lymphangioma is a rare benign congenital lymphatic system malformation that can be locally invasive or can encroach on vital structures.<sup>1-3</sup> They are more commonly diagnosed in children younger than 1 year of age, although it may occur at any age. Lymphangiomas, in turn, occur more frequently in the head, neck, and axilla (in up to 95% of cases), but may occur in almost any anatomic site. They have been reported in the lungs, mediastinum, pleura, pericardium, kidneys, bone, adrenal glands, gallbladder, and stomach.<sup>4-6</sup> The incidence of abdominal lymphangioma varies from 1:23,000 to 1:120,000 acute pediatric hospital admissions.<sup>7-9</sup>

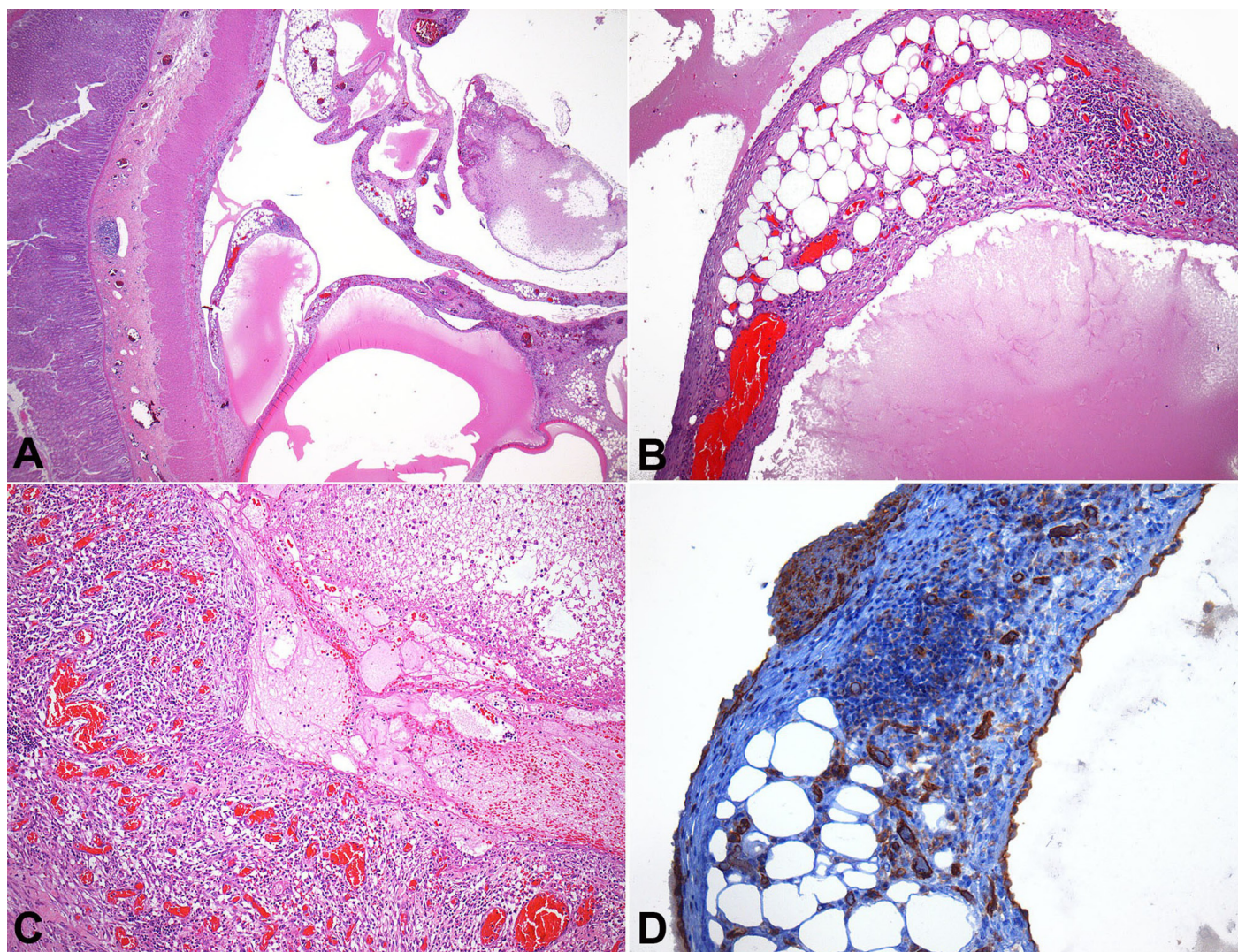
In pediatric patients, the mesentery is the most common localization of abdominal lymphangioma (50-70%), followed by the greater omentum (10-30%), the mesocolon (10-30%), and the retroperitoneal space (10%). Internal forms of lymphangiomas may be associated with cutaneous/subcutaneous forms as well as lympho-hemangiomas with or without artery-venous fistula.<sup>10,11</sup>

The clinical features of intra-abdominal lymphangiomas are variable – depending on the size and location of the tumor – and usually impose preoperative diagnostic difficulty. Most symptoms derive from the tumor's compressive effect. Among adults, some cases are asymptomatic or evolve



**Figure 2.** Gross findings of the surgical specimen. **A** - Note a lobulated mass in the mesenteric face of the sigmoid; **B** - Formalin-fixed of the longitudinal section of the sigmoid colon showing a multiloculated cystic mass within the adipose mesenteric tissue. The intestine wall is preserved without invasion.





**Figure 3.** Photomicrography of the cystic lesion. **A** - Presence of multiple cystic lesions within the subserous layer of the sigmoid segment filled by amorphous and proteinaceous fluid (H&E, 25X); **B** - The cysts are lined by a flat unilayered cells without atypia, similar to endothelium. Note a lymphoid aggregate in the cyst wall (H&E, 100X); **C** - Ulcerative area with mixed inflammatory infiltrate, granulation tissue, and fibrin plug (H&E, 100X); **D** - Positive immunohistochemical reaction for CD-31 in the cystic endothelial lining and in the small vessels of the wall (200X).

over months or years before diagnosis; however, in pediatric cases, up to 88% are symptomatic and the course is of short duration.<sup>5</sup> Some cases are incidentally diagnosed during surgery or autopsy. Abdominal pain seems to be the most frequent symptom, sometimes mimicking acute appendicitis, Meckel's diverticulitis, constipation, bowel obstruction, volvulus, and infarction may be the presenting symptoms—mainly when within the mesentery.<sup>12-16</sup> In the colon, lymphangioma are usually submucosal and may present as polypoid lesions that may bleed and cause hemorrhage or anemia.<sup>17</sup> Insidious, but progressive, abdominal distention resembling ascites occurs in 18-20% of the cases.<sup>18</sup> Malnourishment is

uncommon, but undernutrition may be present in cases where intestinal lymphangiectasis is present, leading to refractory diarrhea and malabsorption syndrome. Acute abdominal pain may be present as a result of bowel torsion or lymphangioma infection.<sup>19</sup> The presence of a palpable mass is quite variable in the literature, ranging from 25% to 77%.<sup>20</sup> In the case presented herein, the lymphangioma occurred in the mesosigmoid and compressed the sigmoid causing pain and constipation. Signs of peritoneal irritation, the presence of a palpable mass, a cystic lesion on imaging examinations, and the deteriorating clinical status were sufficient to indicate the laparotomy without a precise preoperative diagnosis.



However, physical examination, and imaging work-up are usually enough to establish the diagnosis in most cases. Depending on the tumor size, a visible and/or palpable painless, mobile, flat tumor with defined limits may be detected on the physical examination.<sup>1,9,21</sup>

The abdominal x-ray may show an opacification that displaces the intestinal loops or signs of bowel obstruction. Ultrasonography is diagnostic – in most cases – showing single or multiple, varying-sized, thin layered, septated cysts with internal echos.<sup>4,5,22</sup> CT or nuclear magnetic resonance imaging of the abdomen is useful when diagnosis is doubtful or for surgical purposes, since surgical excision may be difficult in some cases due to the insinuating nature of the tumor.<sup>4,6,7,22</sup>

Macroscopically, abdominal lymphangiomas are thin-walled cystic multilocular masses of various sizes, reaching up to 15 cm (6 in). Their external surface is smooth, thin walled, yellowish, grayish, or yellow-pink in color, appearing as multiple cysts or a spongy mass containing watery or milky fluid.<sup>17</sup> The cut surface may show interconnecting cysts, which may contain a clear, chylous, serous, or gelatinous liquid where foamy macrophages, leukocytes, or hemorrhage may be found. Microscopically, the cysts are lined by a flat epithelial endothelium, circumscribed by connective tissue, small lymphatic spaces, smooth muscular fibers, and lymphoid follicles or aggregates, and foam cells.<sup>6,7,23</sup> Malignant transformation does not occur, but in the case of colonic lymphangioma, colon cancer or adenoma may be concurrently diagnosed.<sup>17</sup>

The recommended treatment for the intra-abdominal cystic lymphangioma is a complete surgical resection. The intervention should be performed as soon as possible because of the risk of infection, torsion, hemorrhage, or obstruction.<sup>20</sup> The correct technique depends on the size and shape of the tumor, which can be resected with or without the adjacent intestinal loop or other adjacent involved organ.<sup>24</sup> Prognosis is excellent when the complete resection is feasible. However, relapses may occur if vesicles or part of the tumor remain unresectable.<sup>1,20</sup>

Prognosis is generally favorable; however, local relapse after partial resection, diffuse lymphangiectasis of the adjacent intestinal wall, infection, and myxoid degeneration may be responsible for a poor prognosis.<sup>7</sup>

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