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Case Report

Cystic Lymphangioma of the Colon: Case Report and Literature Review

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Abstract

An 18-year-old female presented with a history of pain in the abdomen with abdominal distension, vomiting, and fever for 2 weeks. Diagnostic imaging showed a multiseptate thin-walled cystic lesion in right lumbar region extending from subhepatic region superiorly to the urinary bladder inferiorly. On exploration, a huge transilluminant multiseptate cystic lesion was present arising from the serosal surface of ascending and transverse colon extending into the lesser sac with attachment to the greater curvature of stomach antrum and encroaching on the head of the pancreas. Histopathology showed multiple dilated lymphatic spaces suggestive of lymphangioma of colon. Lymphangiomas are rare asymptomatic lesion with low threshold for malignancy discovered accidentally with increasing use of colonoscopy. Clinical manifestations are highly variable. Diagnosis is difficult. Hence, it requires careful colonoscopy/endoscopic ultrasonography for proper evaluation. Complete surgical excision is the gold standard treatment to prevent recurrence as they have the potential to grow and can lead to serious complications.

Key words: Abdomen, Abdominal cystic lymphangioma, Case report, Colon, Histopathology, Surgical outcome

INTRODUCTION

Lymphangiomas are rare benign congenital malformations of the lymphatic vasculature with a rate of hospitalization of 1/250,000–1/20,000.[1] Lymphangiomas occur mostly in the head, neck, and axilla region in children with 90% of these becoming evident in few years after birth. However, occurrence in the abdominal or mediastinal cavity is extremely rare, particularly in adults, constituting approximately 5% of all lymphangiomas [Figure 1].[1,2]

The etiology is unclear, but probable causes for intestinal lymphangiomas could be intramural lymphatic obstruction, disturbed endothelial permeability, inflammation, congenital absence of lymphatics, and aging of the bowel wall.[3] Patients may present with a variety of non-specific symptoms such as abdominal distension, abdominal pain, constipation, loss of appetite, nausea, and vomiting.[4] Its pre-operative diagnosis is important so as to occurrence of associated complications.

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CASE REPORT

An 18-year-old female came to tertiary care hospital with the history of pain in the abdomen on the right side with abdominal distension, vomiting, and fever for 2 weeks. On per abdomen examination, right hypochondriac region was distended and tender. Reported weight loss was 15 pounds along with loss of appetite for the past 2 months. She had no history of fever, chills, hemoptysis, hematochezia, and change in bowel habit. There was no history of trauma and previous surgeries. No derangements were noted in general blood test, blood chemistry, or urinalysis. Ultrasonography (USG) was suggestive of gross free fluid in the abdomen seen with multiple thick septa within and abdominal lymphadenopathy. Computed tomography (CT) abdomen showed a 9.5 × 15.9 × 23.7 cm sized multiseptated thin-walled cystic lesion with enhancing septa noted in the right lumbar region extending from subhepatic region superiorly to the urinary bladder inferiorly. The lesion was

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seen displacing the bowel loops, compressing the head of the pancreas, inferior vena cava, and right ureter leading to mild hydroureter and hydronephrosis. Anteriorly, it was seen scalloping posterior and inferior surface of liver and gallbladder. No solid component was noted. Decision was taken to carry out an exploratory laparotomy. On exploration, a huge transilluminant multiseptate cystic lesion was present arising from the serosal surface of ascending and transverse colon extending into the lesser sac with attachment to the greater curvature of stomach antrum and encroaching on the head of the pancreas [Figure 2].

Incomplete excision of the cyst leaving behind the posterior wall, which was encroaching over the head of pancreas along with the resection of the bowel, which was densely adhered to the cystic wall, was done for the patient [Figure 3].

Histopathology confirmed multiple multilocular cysts ranging from 1 to 10 cm on serosal aspects of ascending and transverse colon. Cysts contained seromucinous exudate. Intestinal mucosa was flattened at places. On microscopic examination, cysts on serosal wall showed multiple dilated spaces lined by flattened epithelium, underlying tissue showed lymphocytic infiltrate, and at places, lymphoid aggregates were seen suggestive of lymphangioma [Figure 4]. Post-operative course was uneventful. The patient is currently disease free.

1-year follow-up with USG showed no recurrence.

DISCUSSION

Lymphangiomas of the gastrointestinal (GI) tract are very rare, most of them arise in the mesentery, omentum, mesocolon, and retroperitoneum, and 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] The above case highlights the rarity as well as the importance of cystic lymphangioma of colon occurring on the right side to be kept as an important differential when a surgeon encounters a cystic lesion per the abdomen or in pelvis. Intra-abdominal cystic lymphangioma can present as diagnostic challenge on imaging and can easily be confused with other cystic intra-abdominal lesions, ranging from pancreatic pseudocysts to abdominal tuberculosis, Hydatid disease, or malignancies such as mucinous carcinomatosis. Histologically, lymphangiomas could be capillary, cavernous lymphangiomas, or cystic hygroma. Rarely, a fourth subtype, the hemangio-lymphangioma also occurs.[6] The most common lymphangioma type in the colon was found to be the cystic hygroma type as found in our case.

In the article on review of 279 cases of lymphangioma of colon by Matsuda *et al*., the mean age of onset was 55.2 ± 14.1 years and the male-to-female ratio was 150:92 indicating a higher incidence in males. However, Goh *et al*. series demonstrated that abdominal lymphangiomas have a male preponderance and present more acutely in pediatric patients, whereas in adults, female patients predominate and the history is more chronic as present in our case.[7] It commonly affects the transverse colon followed by the ascending colon, cecum, and descending colon.[8] Although they less commonly involve the GI tract, the clinical presentations of GI lesions are myriad, ranging from asymptomatic adenoma-like polypoid lesions to large obstructing masses. Abdominal pain was the most frequent symptom, followed by bloody stool, constipation, and diarrhea. Weight loss seen in this patient is not a typical finding, but protein-losing enteropathy associated with a large tumor has been reported,[9] which may have resulted in weight loss in our case as well. Complications include compression of surrounding structures, rupture, secondary infection, volvulus, intestinal obstruction, GI

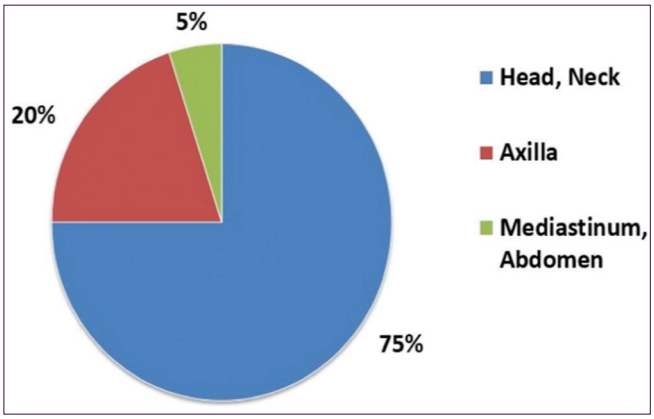

Figure 1: Distribution of lymphangiomas

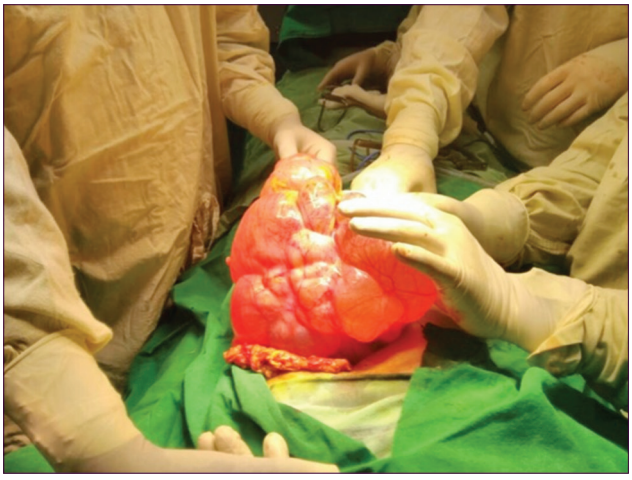

Figure 2: Exploratory laparotomy showing a huge multiseptate cystic lesion

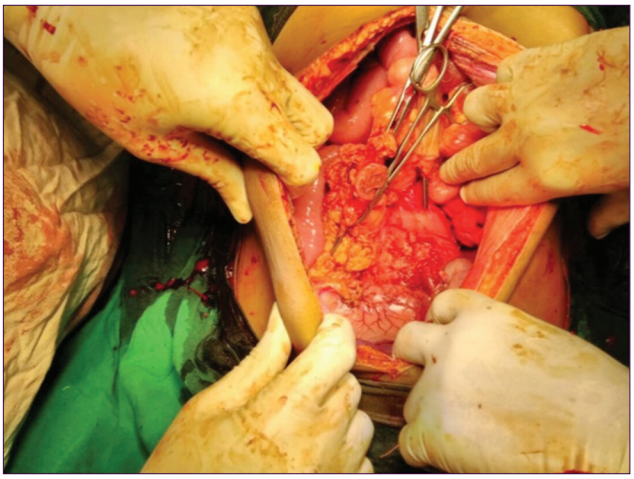

Figure 3: Excision of cystic lesion along with resection of terminal ileum up to right one-third of transverse colon

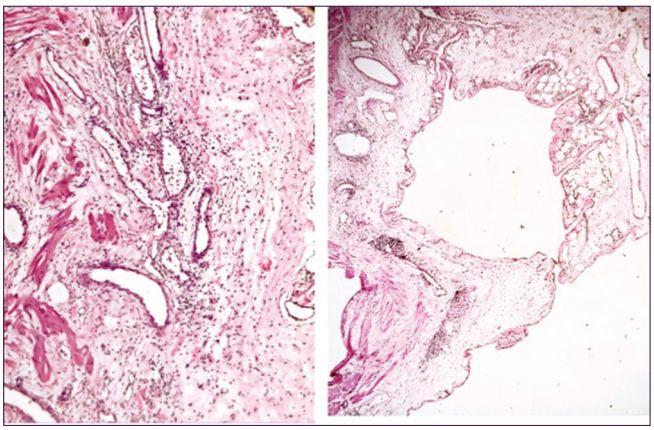

Figure 4: Photomicrograph depicting multiple dilated spaces lined by flattened epithelium with underlying lymphocytic infiltrate

bleeding, and protein losing enteropathy.[10] The huge multiseptate cystic lesion was seen encroaching on the right ureter causing mild hydroureter and hydronephrosis. The accurate pre-operative diagnosis of intra-abdominal lymphangioma is uncommon, particularly in adult patients. It has been stated that diagnosis is possible on careful colonoscopy.[8] On CT, these masses show densitometric characteristics of the fluid type, regular margins, and only a peripheral contrast enhancement. The demerit of CT imaging is that lesions <2 cm cannot be identified.[11] Pre-operative biopsy is helpful to confirm the diagnosis. However, it has been stated that endoscopic biopsy can un-roof the thin wall of these cysts and lead to infection. The diagnosis of lymphangioma is further complicated by histologic overlap with lymphangiectasia of the GI mucosa.[6] The use of endoscopic USG for definite diagnosis of cystic lymphangioma of colon has shown promising results, overcoming the need for resection for biopsy.[12] Magnetic resonance imaging is the most useful pre-operative radiological tool for diagnosis and in surgical planning.[13,14] Complete surgical excision remains the gold standard treatment to prevent recurrence. Pedunculated or semi-pedunculated colonic lymphangioma of about 2 cm or smaller can be managed by polypectomy as reported by Yildiz *et al.* Other treatment options include de-roofing the lymphangioma lesion with cyst drainage and aspiration injection of sclerosant agents.[10] However, these are not recommended due to the high recurrence rates.

Accurate anatomic localization and definition of the lesions are important in pre-operative planning because lymphangiomas have an insinuating nature that makes complete surgical excision difficult as in our case. Although these are rare lesions showing no malignancy, with many cases asymptomatic which were discovered accidentally due to increasing use of endoscopy, they may often lead to life-threatening conditions, such as infection, volvulus, obstruction, or bleeding into the lumen of a cyst that is usually very difficult to manage. They should be treated surgically as they have the potential to grow resulting in recurrence.

In our case, large size of the lesion, critical location, extensive spread within the peritoneum, compression of the surrounding structures, and possibility of malignant nature of the lesion guided our choice for surgical resection.

Pre-operative diagnosis of abdominal cystic lymphangioma is usually difficult due to its variable misleading clinical presentations, rarity of the disease, and its resemblance to many other intra-abdominal cysts, and hence, anterior cruciate ligament should be kept in the list of differentials when the patient is encountered with intra-abdomen multiseptate cystic lesion.

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