

Case Report

Lymphangioma of the Ileocecal Valve Clinically Masquerading as a Submucosal Small Intestinal GIST: Report of a Case and Literature Review

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ABSTRACT

Lymphangiomas are rare tumors affecting the gastrointestinal tract, and may be seen in the bowel, gall bladder, and pancreas. They resemble hemangiomas, but consist of spaces of variable sizes containing lymph. In this report, we describe the case of a 53-year-old male who presented with abdominal pain and constipation. Computerized tomography (CT) scan showed a polypoidal lesion at the ileocecal valve which was thought to be a gastrointestinal stromal tumor. Resected specimen did, however, show a lymphangioma. We also describe the clinicopathologic features of gastrointestinal lymphangiomas with a literature review.

Key Words: Gastrointestinal stromal tumors, laparoscopic surgery, lymphangioma

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Lymphangioma is a benign tumor commonly found in children. It usually occurs in the head, neck, and axillary region.^[1] Many lymphangiomas are congenital malformations of the lymphatic system and are considered to be lymphatic hamartomas.^[2] They are masses consisting of flat endothelial cells and supporting connective tissue. Lymphocytes that are surrounded by islands of fat cells are also usually present.^[3] Colorectal lymphangioma was first reported by Chisholm and Hillkowitz in 1932.^[4] A case of lymphangioma of the ileocecal valve clinically masquerading as small submucosal nodule of the ileocecal valve and mimicking gastrointestinal stromal tumors (GISTs) was encountered in our hospital.

CASE REPORT

A 56-year-old Saudi male was admitted to King Khalid University Hospital (KKUH) with the chief complaint of epigastric pain for 3 months associated with feeling

of fullness, heartburn, and constipation. He had a past medical history of a surgical operation for renal stones and tympanoplasty, but with no significant family history. The vital signs recorded upon admission were as follows: Temperature 36.8°C, blood pressure 147/76 mmHg, pulse rate 72 beats/min, respiratory rate 20 breaths/min, and oxygen saturation 97%. His weight was 64 kg and height was 162 cm. Upon examination, the patient's abdomen was soft and lax with no tenderness, rigidity, or guarding noted.

Laboratory tests revealed an increase in white blood cells (WBC) and bilirubin with decreased red blood cells (RBC), hematocrit (HCT), and sodium level. Upper and lower endoscopic procedures were done, and colonoscopic findings were non-significant up to terminal ileum while gastroesophageal endoscopy showed a moderate hiatus hernia with peptic esophagitis Grade A and thickening of the mucosal folds at the pylorus. Ultrasound of the upper abdomen showed small cysts in both kidneys. Abdominal computerized tomography (CT) showed a small polypoidal lesion at the ileocecal valve [Figure 1]. The patient was later on discharged with home medication of Pantoprazole.

Laparoscopic right hemicolectomy was later performed. Histopathology of the resected specimen showed dilated lymphatic vessels consistent with a lymphangioma. Excess fat and congested blood vessels were also seen near the described

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lymphangioma. There was no evidence of malignancy and the excision appeared complete [Figures 1 and 2]. This finding confirms the lymphatic nature of the vessels shown in Figure 2.

Patient was discharged without complications and he has been keeping well until this date.

DISCUSSION

Lymphangiomas probably represent vascular malformations rather than tumors. They may occur in the skin, subcutaneous tissue, and deep soft tissues or in visceral organs. These lesions exist in three forms: Capillary, cavernous, and cystic (cystic hygroma) variants. Intra-abdominal lymphangiomas are rare, and 60% of the tumors are present in patients under the age of 5 years, but a significant percentage of them does not manifest until adult life.^[1]

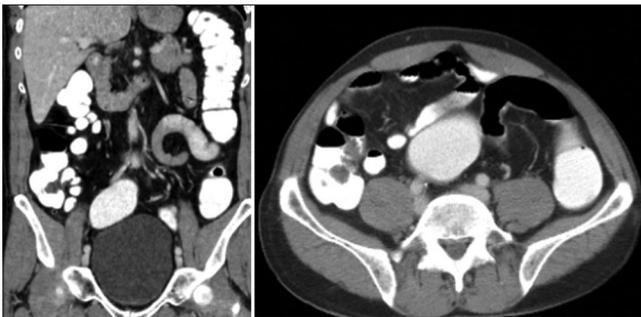


Figure 1: Axial and coronal CT with IV and oral contrast. CT abdomen shows small well-defined intracecal polypoidal mass lesion, related postero-superior to the ileocecal valve

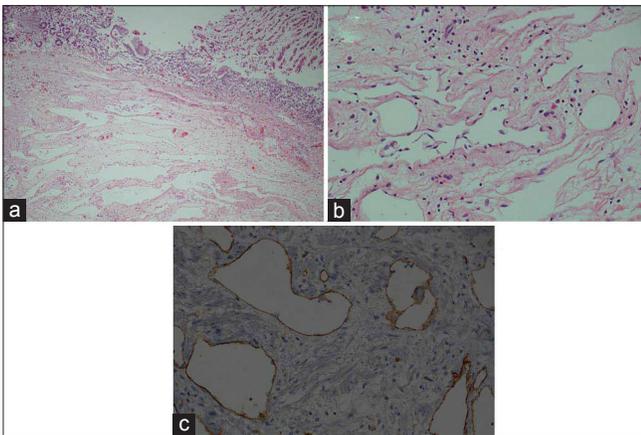


Figure 2: Microscopic findings of lymphangioma of the ileocecal valve. (a) Colonic mucosa with underlying proliferating and irregular lymphatic channels consistent with lymphangioma [hematoxylin and eosin (H and E) stain, $\times 100$]. (b) Lymphangioma high-power view showing lymphatic channels with few scattered lymphocytes in their wall (H and E stain, $\times 200$). (c) Lymphangioma - D2-40 immunohistochemical (IHC) stain for podoplanin (D2-40) showing positive staining of the lining cells (IHC, D240, $\times 100$)

The most common location is in the mesentery, followed by the omentum, mesocolon, and retroperitoneum. In addition to a palpable mass, patients often develop symptoms of an acute condition in the abdomen caused by the common complications of intestinal obstruction, volvulus, and infarction. In fact, a provisional diagnosis of acute appendicitis is frequently entertained because of the common occurrence of right lower quadrant pain. In contrast, retroperitoneal tumors produce few acute symptoms, but ultimately are diagnosed by virtue of a large palpable mass causing displacement of one or more organs.

In the past, an abdominal lymphangioma was seldom diagnosed preoperatively. Currently, the diagnosis can usually be suspected with a combination of radiologic studies.^[5-8]

Ultrasonography is useful for localizing and determining the cystic nature of the tumors. On CT scans, the tumors appear as homogeneous, non-enhancing lesions with variable attenuation values depending on whether the fluid is chylous or serous.

GISTs may arise in the small intestine, colon, or anorectum. These tumors may develop within any portion of the gut wall, but most are centered within the submucosa or the muscularis propria. Most GISTs appear to be well circumscribed, although some may be multinodular. Most GISTs are asymptomatic, but some may present with abdominal pain and bleeding.

Our case represents a rare and perhaps unique example of a submucosal lymphangioma of the ileocecal region which mimicks GISTs both clinically and radiologically.

Furthermore, it distinctly differs from other adult intestinal lymphangiomas of the intestine which usually present with intussusceptions.^[9]

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