Solitary cavernous lymphangioma of the duodenum : a case report

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Abstract

Duodenal lymphangioma is an extremely rare benign tumor of the gastrointestinal tract. In this case report, we describe the case of a 39-year-old Lebanese female with cystic lymphangioma of the duodenum diagnosed by exploratory laparotomy and immunohistochemical analysis. Herein our findings are described. (Acta gastroenterol. belg., **2015**, 78, **60-61**).

Case report

A 39-year-old previously healthy female was referred to the American University of Beirut Medical Center with sudden onset epigastric fullness and pain. She had suffered from abdominal discomfort and had noted unintentional weight loss in the last 3 months. She denied any fever, chills, nausea, vomiting, diarrhea, hematemesis, melena, constipation, or jaundice. Her blood pressure was 125/80 mm Hg, and her body temperature was 36.8°C. On physical exam, epigastric tenderness was noted. Workup was initiated including an ultrasound of the abdomen, which showed an epigastric mass with mixed solid and cystic component next to the head of the pancreas. Consequently, a triple phase CT scan of the abdomen and pelvis was performed that showed a porta hepatis cystic mass, measuring 6.4 × 5.3 × 6.8 cm, abutting the liver laterally and anteriorly, portal vein superiorly, and pancreatic head medially, suggestive of a differential including an enlarged choledochal cyst, a tumor of the head of the pancreas, or a duodenal gastrointestinal stromal tumor (GIST) (Fig. 1) . Upper GI endoscopy did not reveal any endoluminal lesion or intramural ulceration with a normal ampulla of Vater. Endoscopic ultrasound showed a well-defined lesion with irregular border invading the duodenal wall, in favor of an exophytic duodenal GIST. Patient underwent surgical resection through a right subcostal incision. Intra-operatively, a multicystic lesion was noted to be arising from the first part of the duodenum without invading any of the surrounding organs. It was dissected all around, followed by kocherization of the duodenum in order to facilitate local incision and primary closure. The tumor was excised with a rim of 1.5 to 2 cm of normal duodenal wall. After completion of the resection, the duodenum was closed primarily a long its transverse diameter. On macroscopic examination, the tumor is cystic, measuring 7.5 cm × 6.5 cm with attached 4 cm × 2.5 cm duodenal mucosa (Fig. 2). The

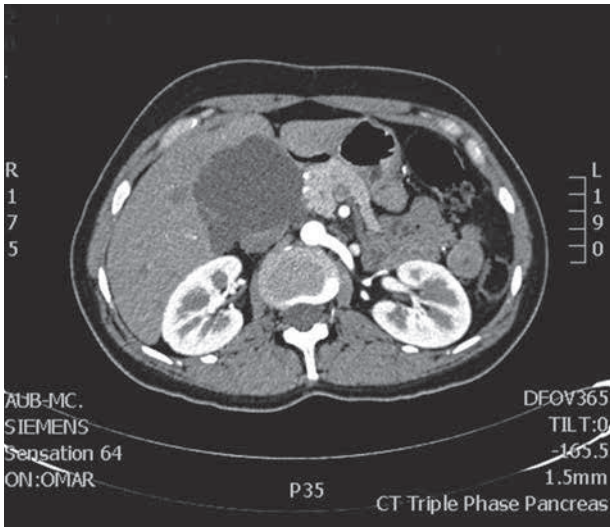

Fig. 1. — CT of the abdomen showing a right sub hepatic lesion at the time of operation.

lumen contained brown fluid and was focally clavicular. Immunohistochemical stains performed with adequate controls showed that the cells lining the cystic spaces express CD34, which confirms their endothelial nature and the diagnosis of a cystic lymphangioma. There was no evidence of malignancy. Her postoperative course was uneventful. No drain was inserted. Following the procedure, Gastrografin was given to the patient and passed to the small intestine with no evidence of leakage. No immediate complications were encountered.

Discussion

Benign tumors of the small intestine are uncommon (1). Moreover, the majority of lymphangiomas arise in the neck and axilla (2) . Thus, the gastrointestinal tract

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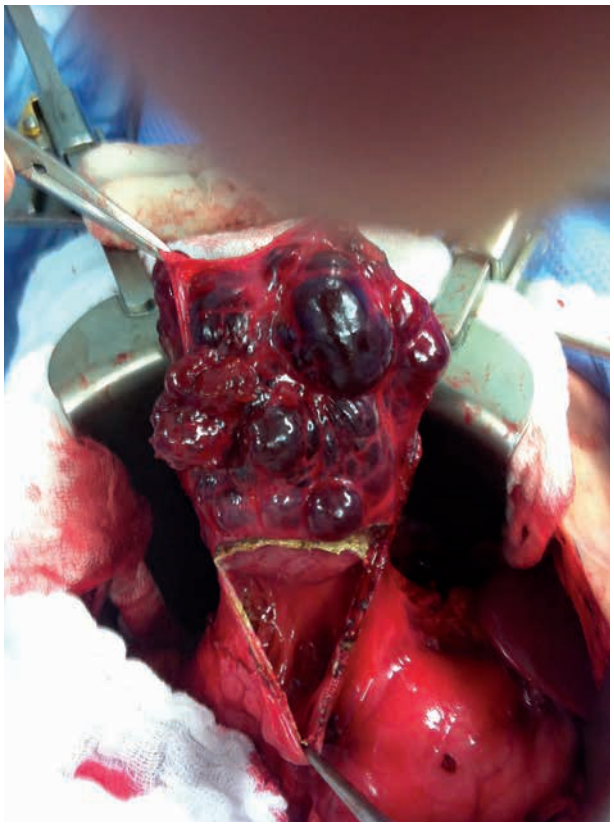

Fig. 2. — Surgical excision through the duodenal wall that reveals spared duodenal mucosa.

is seldom affected with lymphangioma of the duodenum being extremely rare (3). Lymphangioma is generally thought to originate as an obstruction of lymphatic tissue that fails to communicate with the systemic/normal lymphatic system (2,4). Lymphangiomas are pathologically distinguished by dilated lymphatics in the submucosa and can be classified into three types : capillary, cavernous, and cystic lymphangioma (2) . Categorization is based on pathological findings. In the present case, a multi-cystic duodenal lesion was observed and immunohistochemical analysis of the biopsy sample showed CD34 expression. As a result, the patient was diagnosed with cystic lymphangioma of the duodenum.

Although very few have been reported in the literature, the majority of duodenal lymphangiomas are incidentally discovered in asymptomatic individuals (5). Nevertheless, our patient presented with acute epigastric pain and involuntary weight loss, suggesting that the lesion was large enough to cause obstruction of the small bowel. Similar presentations have been previously reported in symptomatic patients diagnosed with lymphangioma of the small bowel (5). Moreover, in the present case it was important to differentiate lymphangioma from lymphatic cysts or lymphangiectasia. Lymphatic cysts are always 1 cm or less in diameter and asymptomatic (6), thus ruling out this possibility. Furthermore, the absence of steatorrhea, chyluria, chylous ascites, and lymphedema rules out a diagnosis of lymphangiectasia.

In conclusion, we describe a rare case of duodenal lymphangioma at the American University of Beirut Medical Center. A benign submucosal, multicystic tumor with brown exudate was observed during exploratory laparotomy and was successfully resected with no postoperative complications. We consider these findings to be key features for the diagnosis of duodenal lymphangioma.

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