Case Report

Peripancreatic cystic lymphangioma with secondary hemorrhage: A rare case report

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ABSTRACT

Lymphangiomas are thought to be true neoplasms, hamartomas or lymphangiectasias. Intra-abdominal lymphangiomas are rare and occur most frequently in children. This is a case report of a 27-year-old female with recurrent episodes of the left-sided upper abdominal pain of 2 years duration. She gives a history of intermittent nausea and vomiting. Liver function test and complete blood count with differential were normal. During the initial stages of illness, the serum lipase was elevated; the serum amylase level was normal all through the entire period. The upper gastrointestinal endoscopy suggested hiatal hernia and mild duodenitis. Two computed tomography scans done at 5 months interval showed a hypodense lesion in the distal tail of pancreas with irregular margins. The size of lesions had decreased from 15 mm × 14 mm to 13.5 mm × 10 mm during this period. Endoscopic ultrasound showed ill-defined area in the distal tail of pancreas and pseudocyst was suspected. Magnetic resonance cholangiopancreatography after 2 months showed a cystic lesion in the tail of pancreas of size 11 mm × 10 mm. Due to increasing severity of pain and fainting spells, the patient was taken up for a distal pancreatectomy. The histopathologic examination confirmed a diagnosis of peripancreatic cystic lymphangioma with secondary hemorrhage. During the postoperative period, the drain amylase was high suggestive of grade A pancreatic fistula. Gradually, the levels decreased, the patient became stable and discharged after pneumococcal vaccination.

Key words: Lymphangioma, pancreas, pseudopancreatic cyst

INTRODUCTION

Many consider lymphangiomas as malformations that arise from sequestrations of lymphatic tissue which fail to communicate normally with the lymphatic system. The most common site of lymphangioma is the neck and axillary regions of children (95%).[1] Intra-abdominal tumors are rare. Galifer *et al*.[2] studied 139 cases of intra-abdominal lymphangiomas. Mesentery was the most common location, followed by the omentum, mesocolon, and retroperitoneum. Other sites of occurrence are the liver, spleen, mediastinum, lung, colon, pancreas, pericardium, pleura, ureter, kidneys, bone, cervix, scrotum, and penis.[1] Tanimu *et al*. have recently reported a single case report of peripancreatic cystic lymphangioma.[3] The occurrence of lymphangioma is uncommon in the peripancreatic location and hence reported.

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CASE REPORT

A 27-year-old female presented with recurrent episodes of left-sided upper abdominal pain. The aching pain radiated to the back and was associated with a history of intermittent nausea and vomiting.

Clinical examination showed mild tenderness in the left hypochondrium. Liver function test and complete blood count with differential were normal during the entire period. The serum lipase level estimated during the initial episode of pain was 730 U/L (normal 73–393 U/L).

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The levels became normal thereafter. The serum amylase level during the entire period showed normal values. The upper gastrointestinal endoscopy suggested hiatal hernia and mild duodenitis. Initial plain and contrast computed tomography (CT) scan showed an enhanced lesion with irregular margins of size 15 mm × 14 mm in the tail of pancreas. A suspicion of mass lesion/focal pancreatitis was raised. The rest of pancreas was normal. A repeat CT scan done 5 months later showed a hypodense lesion in the tail of pancreas measuring 13.5 mm × 10 mm with irregular outline and mild enhancement on contrast. The possible sequelae of pancreatitis and the remote possibility of underlying malignancy were considered. Magnetic resonance cholangiopancreatography done 2 months later showed a small cystic lesion in the tail of pancreas measuring 11 mm × 10 mm in maximum axial dimension. The possibility of residual pseudocyst was considered. The possibility of benign indolent tumor though unlikely could not be ruled out.

The episodes of pain increased in frequency. The patient was taken up for surgery because of the development of associated fainting spells. Initial diagnostic laparoscopy was done to rule out pancreatitis followed by laparotomy. Intraoperative ultrasound could not detect any lesion in the pancreas. A distal pancreatectomy was done. During the postoperative period, the patient was on octreotide and antibiotics. The drain amylase was high suggestive of grade A pancreatic fistula. Gradually, the drain output reduced. The patient was given pneumococcal vaccination and on attaining stability was discharged.

The distal pancreatectomy specimen received in the histopathology laboratory showed a multicystic lesion measuring 40 mm × 10 mm in the peripancreatic fat. It extended from the distal tail end of the pancreas to the spleen [Figure 1]. The cystic spaces were filled with mucoid, brown to black material. The histopathological examination showed medium to large cystically dilated lymphatic channels lined by flattened endothelium. The dilated channels were filled with pink fluid-like material as well as blood. Lymphocytes were seen within the lumen and also in the stroma [Figure 2]. The pancreas and spleen [Figure 3] were free of lesion. The histopathological examination thus confirmed a diagnosis of peripancreatic cystic lymphangioma. The patient has an uneventful 6 months follow-up period.

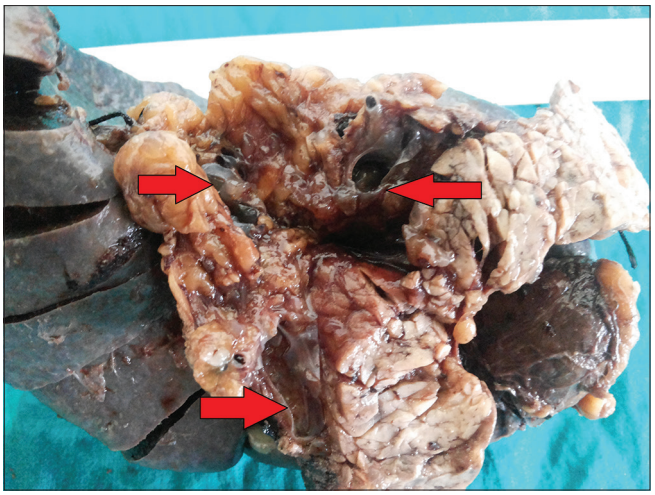

Figure 1: Multiloculated cystic lesion in the peripancreatic fat extending from the distal tail end of pancreas to spleen (red arrows pointing to the cystic areas)

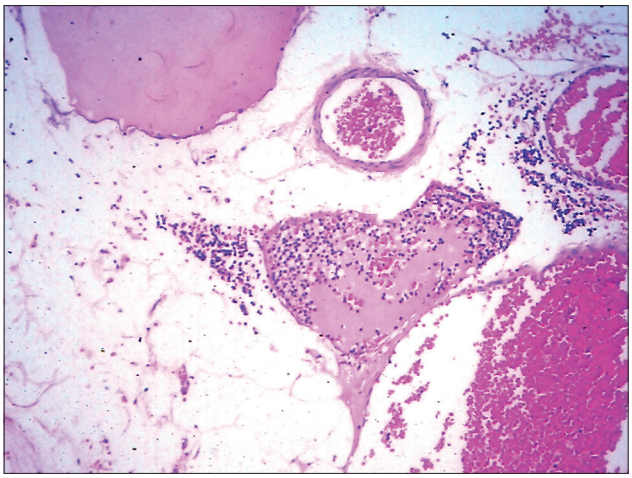

Figure 2: Dilated lymphatic channels filled with pink fluid-like material and blood. Lymphocytes were seen within the lumen and also in the stroma

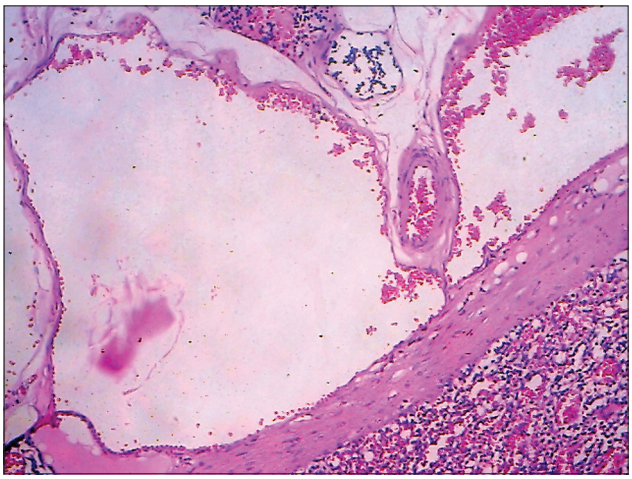

Figure 3: Splenic tissue with adjacent dilated lymphatic channels. Note the splenic capsule separating spleen from the lesion

DISCUSSION

Abdominal lymphangiomas are rare and constitute <1% of all lymphangiomas, they can occur in any age group but are more frequent in female adults.[4] Lymphangiomas in the mesentery, omentum, and mesocolon can present as palpable masses or with symptoms of an acute abdomen either due to intestinal obstruction, volvulus, and infarction.

Retroperitoneal tumors produce a few acute symptoms. A single case report of peripancreatic cystic lymphangioma in a 73-year-old man with abdominal discomfort, intermittent nausea, vomiting, and 17-pound weight loss of 5 months duration is described in the literature.[3] Our patient had in addition to the above symptoms two episode of fainting attacks possibly due to the secondary hemorrhage within the tumor.

Grossly, lymphangiomas may vary from well-circumscribed multiloculated lesions made up of one or more large interconnecting cysts to ill-defined, sponge-like lesions which are composed of microscopic cysts. The cysts usually contain milky fluid. The lymphatic spaces are lined by attenuated endothelium. Small lymphatic spaces usually have inconspicuous adventitial coat, and large lymphatic spaces may have fascicles of poorly developed smooth muscle. Proteinaceous fluid with lymphocytes and occasionally erythrocytes are present in the spaces. The stroma shows small lymphoid aggregates. In the present study, the cystically dilated spaces contained mucoid brown to black material and not milky fluid due to the secondary hemorrhage within the tumor.

Histologically, lymphangiomas with secondary hemorrhage may be confused with cavernous hemangiomas. The presence of lymphoid aggregates in the stroma and irregular lumens with widely spaced nuclei are features consistent with a diagnosis of lymphangioma over hemangioma on routine staining. In the present study, both the features were present on light microscopy for the histopathological diagnosis of lymphangioma. When in doubt immunohistochemistry for lymphatic lineage markers mainly VEGFR3, D2-40 may also assist in this distinction.[5] However, studies done by Bhawan *et al*. on cutaneous lymphangiomas concluded that there was no advantage in performing immunohistochemical staining in differentiating lymphangioma from hemangioma.[6]

Lymphangiomas can be preoperatively diagnosed by radiological modalities such as abdominal ultrasonography (USG), CT, magnetic resonance imaging (MRI), and angiography. Septated cysts can be visualized in USG. CT and MRI may give additional information regarding the capsule, the septation of the cyst, and their thickness. Preoperative fine-needle aspiration cytology (FNAC) may be done, and the biochemical characteristics and cytological features of the fluid may be studied. The role of FNAC is still controversial as it might cause hemorrhage, rupture, or tumor implantation in malignant cases.[7] A definitive diagnosis may be possible only by a histopathologic examination of the resected lesion.[8]

Complete resection of lymphangiomas is advised since there are chances of recurrence. In the present case, a distal pancreatectomy was done due the presence of a palpable firm cystic lesion extending from the distal tail of pancreas and attaching to the spleen intraoperatively.

Akwei *et al*. have reported a case report of benign mesenteric lymphangioma with previous history of an inguinal hernia repair and a hiatus hernia.[9] In Dubowitz syndrome, subcutaneous lymphangiomas, hiatal hernia, and umbilical hernia are described.[10] Our patient had associated hiatal hernia. The association of lymphangiomas with hernia may require further studies.

In summary, we would like to conclude that peripancreatic cystic lymphangiomas are rare and difficult to diagnose preoperatively. Peripancreatic cystic lymphangiomas may be considered in the differential diagnosis of pancreatic cystic lesions, even though they are rare. The importance of diagnosing lymphangiomas is that total resection of the tumor is mandatory to prevent recurrence. The hiatal hernia associated with lymphangioma may require further studies.

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Conflicts of interest
There are no conflicts of interest.

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