Cystic pancreatic lymphangioma

Nazim Gureş, Ercument Gurluler, Altan Alim, Ibrahim Berber, Alihan Gurkan

Acıbadem University, International Hospital General Surgery Department, Istanbul, Turkey

Abstract

Lymphangioma of the pancreas is a rare benign tumor of lymphatic origin. Retroperitoneal lymphangiomas account for 1% of all lymphangiomas. Herein, we report a case of cystic pancreatic lymphangioma diagnosed in 34 year-old female patient who was hospitalized for a slight pain in the epigastrium and vomiting. Radiological imaging revealed a large multiloculated cystic abdominal mass with enhancing septations involving the upper retroperitoneum. During the laparoscopic surgery, a well circumscribed polycystic tumor was completely excised preserving the pancreatic duct. The patient made a complete recovery and is disease-free 12 months postoperatively.

Introduction

Lymphangioma is a well-known benign tumor and its cystic abnormalities of the lymphatic vessels are predominantly congenital. Cystic lymphangioma usually occurs in the neck, axillary region, and rarely in the mediastinum, which frequently occurs in children and young adults.[1] The etiology of these tumors is thought to be congenital lymphatic malformations causing obstruction of the lymphatic flow leading to lymphangiectasis.[2] The abdominal organs are uncommon sites of origin.[3] Of all the lymphangiomas in the peritoneal cavity, about 70% have been found in the mesentery of small intestine.[4]

Several cases of lymphangioma in abdominal organs were reported, however, the pancreas is one of the rarest origins.[3] Other cystic lesions in the pancreas include pseudocysts, simple cysts, serous cystadenomas, mucinous cystic neoplasms and intraductal papillary mucinous neoplasms.[5] Cystic pancreatic lymphangiomas may clinically present as a palpable abdominal mass and cause diagnostic dilemmas with other retroperitoneal cystic tumors, including those arising from the liver, kidney and pancreas. They may manifest with clinical symptoms such as abdominal pain, fever, fatigue, weight loss, and hematuria, due to their size, and occasionally might be complicated by intracystic hemorrhage, cyst rupture, volvolus and/or infection.[6]

Although abdominal sonography (US) or abdominal computed tomography (CT) scan can also incidentally reveal lymphangioma, preoperative diagnosis is difficult as conventional imaging studies cannot characterize this lesion.[7] A larger or symptomatic lymphangioma is treated by total resection to prevent recurrence, infection, torsion and enlargement. Although lymphangioma rarely becomes malignant, its prognosis is generally good.[8]

An interesting and rare case of a retroperitoneal cystic lymphangioma in a 34-year-old female patient is described here.

Case Report

A 34 year-old woman presenting with nausea and vomiting was referred to our hospital. She had no remarkable past medical and previous abdominal surgery history. Upon physical examination, there was no palpable mass and she was found to be slightly tender at the epigastrium. US (Figure 1A) and CT (Figure 1B) revealed a well circumscribed, 6×10.5 cm cystic lesion with multiple septations located in the upper ventral pancreatic area neighboring posterior stomach and left lobe of the liver. Laboratory screening results were as follows: C-reactive protein: 0.14 mg/dL, white blood cell count: 6900/mm^3, hemoglobin: 11.1 g/dL, platelet count: 338.000/mm^3, amylase: 64 U/L, CA 19-9: 0.78 U/mL. With US-guided biopsy yellowish serous fluid sample was obtained and histopathologic examination showed dense mature lymphocyte groups and histiocytes scarcely which was thought to be lymphangioma (Figure 1C). By laparoscopic exploration, a cystic mass adherent to the upper border of pancreatic corpus was detected. The cyst was resected with clear margins with the pancreatic capsule preserving the hepatic artery and the pancreatic tissue. The surgical specimen was extracted with the assistance of an endobag. The tumor, measuring 8×4.5×3 cm, had nodular, red - brown surface and was surrounded by normal pancreatic tissue (Figure 1E). By sectioning, it was seen that the tumor had multiple cystic dilatations, of 0.4-1.5 cm, filled with yellowish hemorrhagic fluid. Upon microscopic examination, dilated cystic lymphatic vessels lined with cuboidal epithelium had significant smooth muscle layer showed patchy lymphoid groups due to the chronic lymphoid cell infiltration (Figure 1D). The final pathological diagnosis was reported as cystic lymphangioma by the Pathology Department.

Correspondence: Nazim Gureş, International Hospital, General Surgery Department, Cad. No: 82 Yesilkoy, 34149 Istanbul, Turkey
Tel. +902124684238 - Fax: +902126620795
E-mail:guresnazim@yahoo.com

Key words: lymphangioma, pancreas, laparoscopic.

Conflict of interests: the authors report no conflicts of interests.

Received for publication: 26 February 2012.
Revision received: 16 March 2012.
Accepted for publication: 22 April 2012.

Rare Tumors 2012; 4:e27
doi:10.4081/rt.2012.e27

Discussion

Cystic lymphangiomas are multiloculated soft cystic masses, composed of a combination of variably sized, dilated lymphatic channels, divided by thin septae. It is believed that they develop from lymphatic vessels which dilate progressively because of insufficient drainage, due to atresia or inadequate efferent channels.[7] The vast majority of them (90%) are discovered before the age two, and half of them at birth. They most commonly occur in the neck, axilla and mediastinum in the pediatric age group.[9] Pancreatic lymphangioma is a very rare entity. In 1913, Koch had reported the first case.[3] Most reports in the literature describe the body and tail of the pancreas as the most common regions involved. Tumor size may vary between 3 and 20 cm in diameter (average 12 cm).[10] Clinically, these tumors may mimic pancreatic neoplasms. The patients are mostly asymptomatic. When complicated, anemia, pain, hemorrhage, nausea, hydronephrosis and/or infection may also be present.[2] US typically shows a polycystic tumor and calcifications which are typical for cystadenomas of the pancreas. On CT, the tumor is a well-circumscribed, encapsulated, water- isodense, polycystic tumor with thin septa similar in appearance to the cystadenomas, which occur far more frequently.[11] The histopathological diagnosis of pancreatic lymphangioma has traditionally been established after surgery. Endoscopic Ultrasound-Guided Cyst Aspiration is recommended to establish a definite preoperative diagnosis.[7]

Although not all pancreatic cystic lesions need aspiration, our patient was symptomatic,

with a large pancreatic cyst, in an easily accessible site. Therefore, we decided to aspirate the cyst preoperatively because of its unclear morphology, and also because our patient wanted a positive diagnosis before undergoing a laparotomy. Surgical resection is the treatment of choice. Complete resection should be performed whenever possible. After complete resection, recurrences can also occur, but the rate is much lower around 7%.[9] The laparoscopic approach is reported to be a safe and effective alternative for the treatment of retroperitoneal cystic lymphangioma.[12,13] Our patient presented with complaints of upper abdominal pain, nausea and vomiting. US and CT imaging revealed a large septated cyst saddled on the pancreatic body neighboring posterior stomach and the left lobe of the liver. The patient has no previous pancreatitis history and no evidence of pancreatic pseudocysts. The tumor markers were negative before the operation. The cystic aspiration fluid showed benign findings in the favor of lymphangioma. We performed diagnostic laparoscopy initially and ruled out the possible signs of invasive metastasis and evaluated the resectability of the lesion. Afterwards, having continued laparoscopically, we resected the lesion with clear margins from the upper pancreatic body. Pathologic examination confirmed the diagnosis of cystic lymphangioma of the pancreas.

Conclusions

Cystic pancreatic lymphangiomas are rarely seen tumors. Although lymphangioma is pathologically benign, surgical removal should be the first choice for symptomatic lymphangiomas. Complete surgical resection is curative. Laparoscopic excision should be considered as a therapeutic option to treat pancreatic cystic lymphangioma for uncomplicated cases.

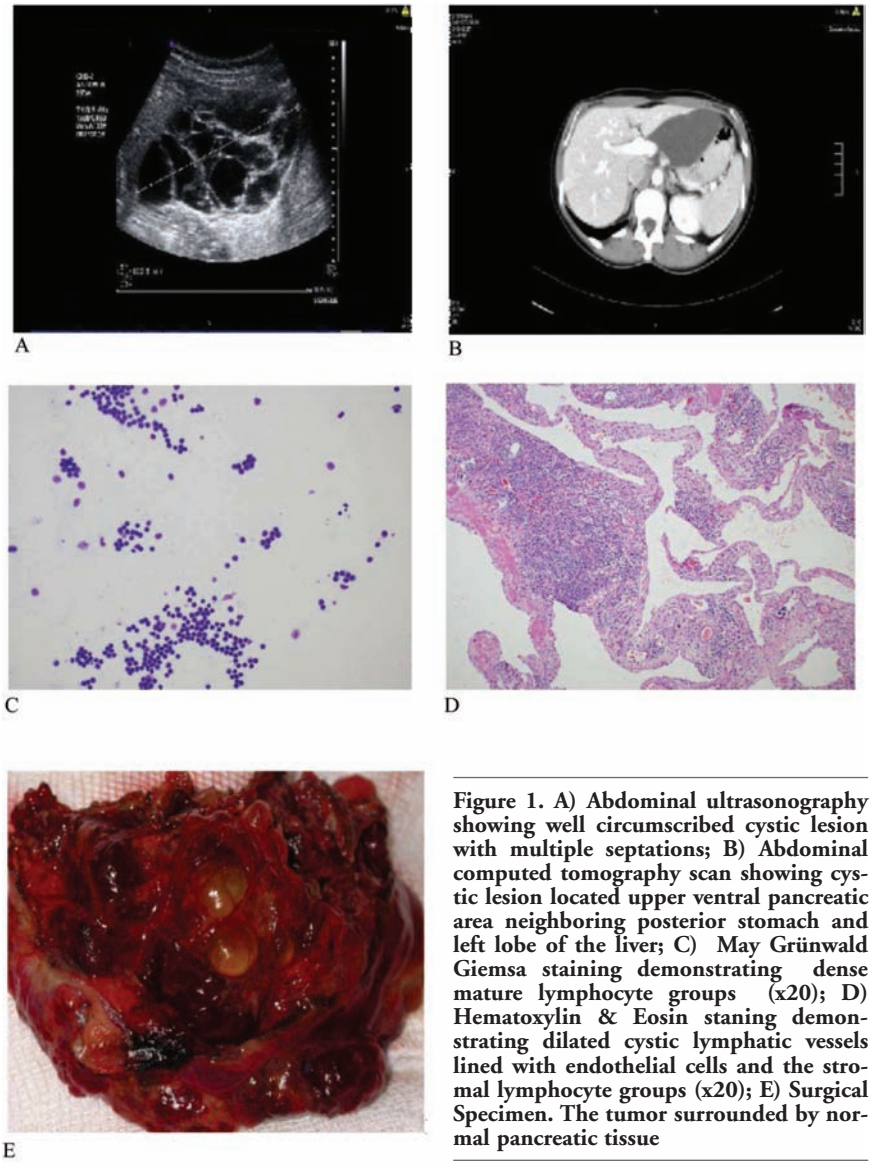

Figure 1. A) Abdominal ultrasonography showing well circumscribed cystic lesion with multiple septations; B) Abdominal computed tomography scan showing cystic lesion located upper ventral pancreatic area neighboring posterior stomach and left lobe of the liver; C) May Grünwald Giemsa staining demonstrating dense mature lymphocyte groups (x20); D) Hematoxylin & Eosin staning demonstrating dilated cystic lymphatic vessels lined with endothelial cells and the stromal lymphocyte groups (x20); E) Surgical Specimen. The tumor surrounded by normal pancreatic tissue

References

1. Kanzaki M, Kikkawa T, Obara T, Onuki T. Successful excision of an isolated mediastinal cystic lymphangioma with bilateral thoracoscopic surgery. Ann Thorac Cardiovasc Surg 2011;17:570-2.
2. Fahimi H, Faridi M, Gholamin S, et al. Cystic lymphangioma of the pancreas: diagnostic and therapeutic challenges. JOP 2010;11:617-9. Erratum in: JOP 2010;11:619.
3. Kawaguchi K, Motoi F, Ohtsuka H, et al. Cystic lymphangioma of the pancreas with spontaneous rupture: report of a case. Case Rep Gastroenterol 2011;5:288-94.
4. Kim JH, Ryu WS, Min BW, et al. Acquired omental cystic lymphangioma after subtotal gastrectomy: a case report. J Korean Med Sci 2009;24:1212-5.
5. Chung JC, Kim HC, Chu CW, et al. Huge cystic lymphangioma of the pancreas. Can J Surg 2009;52:E303-5.
6. Bhavsar T, Saeed-Vafa D, Harbison S, Inniss S. Retroperitoneal cystic lymphangioma in an adult: a case report and review of the literature. World J Gastrointest Pathophysiol 2010;1:171-6.
7. Bhatia V, Rastogi A, Saluja SS, et al. Cystic pancreatic lymphangioma. The first report of a preoperative pathological diagnosis by endoscopic ultrasound-guided cyst aspiration. JOP 2011;12:473-6.
8. Chung SH, Park YS, Jo YJ, et al. Asymptomatic lymphangioma involving the spleen and retroperitoneum in adults. World J Gastroenterol 2009;15:5620-3.
9. Surlin V, Georgescu E, Dumitrescu C, et al. Retropancreatic cystic lymphangioma - considerations upon a case. Rom J Morphol Embryol 2011;52Suppl1:493-6.
10. Ghatak S, Ray S, Sanyal S, et al. An unusual cause of acute abdomen in adults: giant cystic lymphangioma of the pancreatic head. A clinical case and literature review. JOP 2011;12:266-70.
11. Colovic RB, Grubor NM, Micev MT, et al. Cystic lymphangioma of the pancreas. World J Gastroenterol 2008;14:6873-5.
12. Tsukamoto T, Tanaka S, Yamamoto T, et al. Laparoscopic excision of a retroperitoneal cystic lymphangioma: report of a case. Surg Today 2003;33:142-4.
13. Wildhaber BE, Chardot C, Le Coultre C, Genin B. Total laparoscopic excision of retroperitoneal cystic lymphangioma. J Laparoendosc Adv Surg Tech A 2006;16:530-3.