CASE REPORT

Differential Diagnosis of Cystic Lymphangioma of the Pancreas Based on Imaging Features

Ting-Kai Leung, Chi-Ming Lee, Li-Kuo Shen, Ya-Yen Chen*

Lymphangioma is a benign tumor, which is a consequence of lymphatic malformation with blockage of lymphatic flow. Most lymphangiomas occur in the neck and axillary region, and < 1% occur in the mesentery or retroperitoneum. Lymphangiomas arising from the pancreas are extremely rare. We report the case of a 34-year-old woman with cystic lymphangioma of the pancreas without major symptoms or signs. A 6 × 6 cm intra-abdominal cystic mass was incidentally revealed by sonography during a health examination. It is always a challenge to differentiate the lesion from other possible cystic-like pancreatic neoplasms. Differential diagnosis of cystic lymphangioma from other cystic-like tumors of the pancreas can be performed based on their imaging characteristics, including presence of septa, cystic or wall calcification, soft tissue, wall thickness, single or multiple loculation, and dilatation of the pancreatic duct. Post-gadolinium magnetic resonance imaging is excellent in defining the origin of intra-abdominal cystic mass and intracystic septa. [*J Formos Med Assoc* 2006;105(6):512–517]

Key Words: cystic-like neoplasm, cystic lymphangioma, pancreas

The availability of medical imaging modalities such as sonography, computed tomography (CT) and magnetic resonance imaging (MRI) has led to the increasing discovery of cystic lesions of the pancreas in asymptomatic patients. Although only a minority of pancreatic cystic tumors are true neoplasms, proper diagnosis and accurate staging is important for early treatment and to increase the length of survival in patients.[1] In the majority of cases of cystic pancreatic tumor, the differential diagnosis includes non-neoplastic cystic lesions such as pseudocysts,[2] and neoplastic cystic lesions such as serous cystadenomas, mucinous cystic neoplasms, nonfunctioning islet cell tumors with cystic degeneration, and cystic solid and papillary epithelial neoplasms. Malignant cystic tumors are rare and comprise only about 1% of all pancreatic malignancies.[3] They are sometimes potentially curable by surgery. Many benign pancreatic cystic neoplasms are considered premalignant and should be resected. Pseudocysts do not always require resection, and many may be treated with observation, internal drainage or other methods. Correct diagnosis of these lesions is essential to determining appropriate treatment. We report a case of cystic pancreatic lymphangioma, including its preoperative diagnosis and treatment.

Case Report

A 34-year-old woman with no history of systemic disease was referred to our hospital because of the incidental sonographic finding of an intra-abdominal cyst during a health check-up. According to the admission note, there was no abdomi-

Department of Diagnostic Radiology, Taipei Medical University Hospital, Taipei, Taiwan.

Received: March 17, 2005
Revised: April 27, 2005
Accepted: September 13, 2005

***Correspondence to:** Dr. Chi-Ming Lee, Department of Diagnostic Radiology, Taipei Medical University Hospital, 252, Wu Hsing Street, Taipei 110, Taiwan.
E-mail: yayen0220@yahoo.com.tw

nal fullness, tenderness or jaundice. Serum levels of carcinoembryonic antigen (CEA), CA19-9 and amylase were all normal. Follow-up spiral CT scan revealed a 6 × 6 cm non-enhanced thin encapsulated intraperitoneal cyst with homogeneous hypodensity, the major border of which was continuous with the pancreatic tail (Figure 1A). No soft tissue component and no calcification were found. The cyst exhibited obvious mass effect on adjacent organs, with anterior and inferior displacement of the stomach and spleen, respectively. The pancreatic parenchyma showed homogeneous enhancement without dilatation of the pancreatic duct. No mesenteric infiltration or enlarged lymph nodes could be identified. Based on CT findings, pancreatic cyst was suspected, but the possibility of a duplication cyst, mesenteric lymphangioma or mesenteric cyst could not be ruled out. MRI (1.5-T; GE Medical Systems, Milwaukee, WI, USA) study further confirmed a cystic pancreatic mass with characteristic appearance of low signal intensity on T1WI and high signal intensity on T2WI without post-gadolinium enhancement. Unlike the CT study, MRI in the coronal view of T2WI could distinguish a clear border from the stomach. MRI also revealed some thin curvilinear septations separating cystic areas on T2W1 and post-gadolinium T1W1 (Figure 1B,C). Since both the imaging and laboratory studies did not favor a pseudocyst or advanced malignancy, surgical exploration was arranged. Intraoperatively, the lesion was found to originate from the pancreatic tail and to be composed of multiloculated cystic spaces and thin transparent walls. Some clear fluid content was found within the cystic spaces (Figure 1D). Microscopically, the resected cyst showed dilated lymphatics with flattened lining cells associated with lymph-like fluid content, focal lymphocytic infiltration and supporting fibrous stroma (Figure 2). Immunohistochemical studies of the lining cells

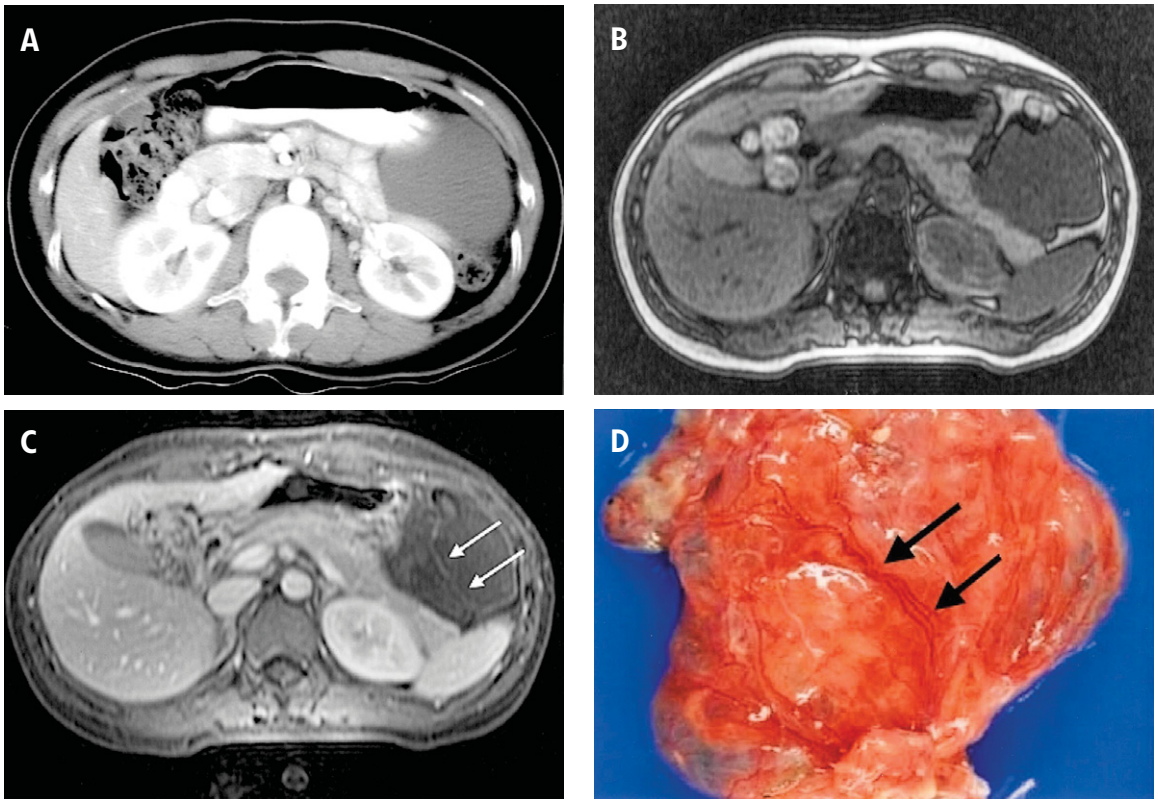

Figure 1. (A) Enhanced computed tomography reveals a thin encapsulated intraperitoneal cyst continuous with pancreatic tail. (B) T1-weighted (TR = 120 ms, TE = 2.2 ms) axial magnetic resonance imaging (MRI) reinforces our imaging diagnosis of a pancreatic tail cystic mass (homogeneous hypointense appearance). (C) Gadolinium-enhanced axial MRI shows a hyperintense thin septum (arrows) within the cystic mass. (D) Grossly, the major cyst is composed of multiloculated cystic spaces with transparent thin wall and thin septum (arrows), which corresponds to the findings in (C).

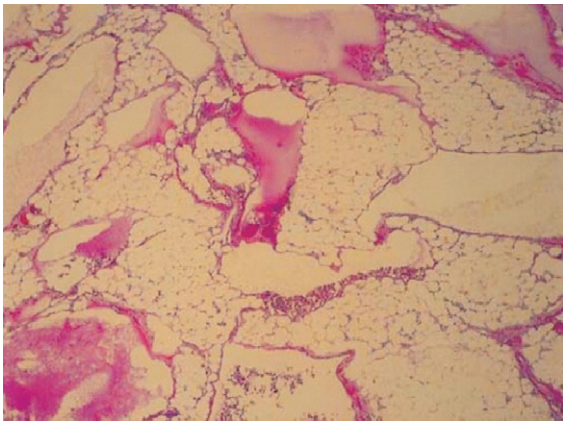

Figure 2. Histologic examination shows dilated lymphatics and lymph-like fluid content (hematoxylin & eosin, × 200).

using a Factor VIII-R antigen test were positive. The pathologic diagnosis was cystic lymphangioma.

Discussion

Lymphangiomas are benign lymphatic malformations that lead to blockage of lymphatic flow and lymphangiectasia. Gui et al described these sequestered lymphatic channels as a developmental abnormality rather than a true neoplasm.[4] Most lymphangiomas occur in the neck and axillary region. Abdominal lymphangiomas are rare, accounting for < 1% of all lymphangiomas, and mainly occur in the mesentery and retroperitoneum.[5–7] Our review of the literature from 1995 to 2005 revealed only a few cases of pancreatic lymphangioma (Table 1). Most reported cases, including the present case, were cystic type lymphangioma. One case of cavernous lymphangioma was reported.[8] Pancreatic lymphangioma occurs predominantly in females, with a mean age of 28.9 years at initial presentation.[2]

The initial clinical symptoms are variable and may include abdominal pain, nausea, vomiting, and a palpable abdominal mass. In some cases, however, the cysts are symptomless and discovered as an incidental finding,[4] as in the present case. No specific or significant laboratory abnormalities have been reported. Previous reports of pancreatic lymphangioma have suggested that imaging studies including CT and MRI could confirm a pancreatic cystic mass but are unable to classify the type of cystic lesions.[9] Our review of the literature suggested that the imaging characteristics of cystic lymphangioma may be useful in the differential diagnosis from other cystic tumors of the pancreas (Table 2). Resembling cystic lymphangioma, serous adenoma has multiple cysts and very thin septa, some of them contain a central stellate scar and sunburst calcification.[10,11] Scott et al reported that about 75% of mucinous cystadenomas are unilocular or multilocular with thin septa.[12] In 66% of mucinous cystadenocarcinomas, the lesions are multilocular with thick walls and solid excrescences. Overall, 62% of mucinous cystic neoplasms of the pancreas exhibit cyst wall calcification, focal thickening of the cyst wall and papillary projections.[12] The imaging characteristics of cystic-like neoplasms of the pancreas are summarized in Table 2.

Differential diagnosis using serum tumor markers may also be useful, because mucinous cystic neoplasms of the pancreas are immunoreactive for CEA and CA19-9. Most cases of nonfunctioning islet cell tumors with cystic degeneration have a thick wall and an irregular inner surface. Over 90% of cystic solid and papillary epithelial neoplasms were associated with dilatation of the pancreatic duct.[8] Patients with pseudocysts often have a history of acute or chronic pancreatitis, while the majority of other cystic tumors may lack such an antecedent factor. The radiographic characteristics of pseudocysts are usually unilocular and there is associated pancreatic duct dilatation. In addition, pseudocysts have a different appearance from cystic lymphangiomas in terms of CT findings, with absence of septa, loculation, solid component, and cyst wall calcification.[13] Initially, medical imaging modalities, sonography or spiral CT may have the advantages of lower cost and shorter acquisition time, in addition to higher sensitivity to cystic calcification. However, this case suggests the advantage of post-gadolinium MRI studies to define the thin septa, which contrast-enhanced CT could not demonstrate. In addition, the coronal and sagittal views of MRI could further distinguish the structural interface with other

Table 1. Imaging characteristics of pancreatic lymphangiomas reported between 1995 and 2005*

Reference	Year	Sex	Age (yr)	Location in pancreas	Size (cm)	Presence of septa	Imaging modalities positive for septa	Presence of solid part	Loculation	Wall thickness
Daltrey & Johnson[18]	1995	F	14	Body	14	Grossly not prominent	(–)	Present	Multiloculate	NM
Fan et al[19†]	1995	M	34	Head	4	Present, with enhancement	CT(+)	Present	Multiloculate	Thin
Viola et al[20]	1997	F	29	Tail	20	Present	Sono(+)	None	Multiloculate	Thin
Abe et al[21]	1997	F	67	Extralobular connective tissue of pancreas	8	Present	CT(+) MRI(+)	None	Multiloculate	Thin
Gray et al[22]	1998	F	31	Body-tail	12	Present	CT(+)	None	Multiloculate	Thin
Shinozaki et al[17]	2001	M	47	Head	NM	Present, with fat component	CT(+) MRI(+), high SI septum	None	Multiloculate	Thin
Titus et al[23]	2001	M	20	Body-tail	20	Present	CT(+)	None	Multiloculate	Thick, uneven
Schneider et al[24]	2001	F	43	Head (affect duodenum)	8	Present	MRI(+)	None	Multiloculate	Thin
Igarashi et al[25]	2001	F	33	Extralobular connective tissue of pancreas	23	Present, not prominent	CT, MRI not prominent	None	Multiloculate	Very thin (difficult to identify)
Gui et al[4]	2003	F	36	Tail	1.5	Grossly not prominent	CT(–)	Mesenchymal stroma-like	Uniloculate (grossly)	Not visible
Casadei et al[26]	2003	M	44	Tail	15	Present	Sono(+)	None	Multiloculate	Thin
Casadei et al[26]	2003	F	40	Tail	NM	Present	CT(+)	None	Multiloculate	Thin
Casadei et al[26]	2003	F	28	Body to tail	20	Present	CT(+)	None	Multiloculate	Thin
Leung et al (this report)	2006	F	34	Tail	6	Present	CT(–) MRI(+)	None	Multiloculate	Thin

**None of the imaging studies in these cases showed pancreatic duct dilatation and central stellate scar; †only case that showed cystic calcification and that was proven to be cavernous type lymphangioma. NM = not mentioned; CT = computed tomography; Sono = sonography; MRI = magnetic resonance imaging.*

adjacent organs in the cystic lesion, which was too large to define on axial spiral CT. MRI is superior to CT in ruling out communication between cyst and pancreatic duct[14,15] (Table 2). The imaging features of pancreatic lymphangioma can be used systematically to eliminate the other possible cystic-like tumors of the pancreas.

References

1. Goldsmith JD. Cystic neoplasms of the pancreas. *Am J Clin Pathol* 2003;119:S3–16.
2. Kehagias D, Smyrniotis V, Kalovidouris A, et al. Cystic tumors of the pancreas: preoperative imaging, diagnosis, and treatment. *Int Surg* 2002;87:171–4.
3. Gasslander T, Arnelo U, Albiin N, et al. Cystic tumors of the pancreas. *Dig Dis* 2001;19:57–62.
4. Gui L, Bigler SA, Subramony C. Lymphangioma of the pancreas with "ovarian-like" mesenchymal stroma: a case report with emphasis on histogenesis. *Arch Pathol Lab Med* 2003;127:1513–6.
5. Okur H, Kucukaydin M, Ozokutan BH, et al. Mesenteric, omental, and retroperitoneal cysts in children. *Eur J Surg* 1997;163:673–7.
6. Ng IO, Fan ST, Nicholls J, et al. Lymphangioma of the pancreas. *J Gastroenterol Hepatol* 1989;4:299–302.
7. La Greca G, Rapisarda V, Randazzo V, et al. Cystic lymphangioma of the pancreas: clinical case and review of the literature. *G Chir* 2001;22:295–8. [In Italian]

Table 2. Imaging characteristics of other cystic-like neoplasms of the pancreas[14–16]

	Common imaging findings								
	Cystic wall calcification	Calcification or central scar	Focal thickening of cyst wall (> 3 mm)	Dilatation of pancreatic duct	Communication between cyst and pancreatic duct	Soft tissue component or papillary projection	Presence of septa	Loculation	Higher incidence of pancreatic location
Other cystic pancreatic tumor									
Serous adenoma	Less frequent	More frequent	Less frequent	Less frequent	Rare	Less frequent	Non prominent or appear as delicate fibrous septum	Multiloculate (more frequent)	More in body and tail
Mucinous cystic neoplasm	More frequent	Less frequent	More frequent	Less frequent	Less frequent	More (thick septum)	Malignant lesions may have thickened wall or septum	Uniloculate (more frequent)	Predominantly in head
Nonfunctioning islet cell tumor		Frequent	More frequent	Variable	Absent	More frequent	Variable	Variable	Head:body and tail (1:1)
Cystic solid and papillary epithelial neoplasm with cystic degeneration	Not frequent	Not frequent	More frequent	More frequent	May or usually present	More frequent	Less frequent	Variable	Predominantly in tail
Pseudocyst	Less frequent	Less frequent	More frequent	More frequent	Usually present	May mimic by presence of necrotic pancreatic or peripancreatic debris or blood	Less frequent	Uniloculate	Predominantly in body and tail (extra-pancreatic cysts)
Cystic lymphangioma (according to our 14 review cases)	0	1	1	0		3	12	Multiloculate (13) Uniloculate (1)	Head (3) Body-tail (9) Extralobular connective tissue of pancreas (1)

8. Fukukura Y, Fujiyoshi F, Sasaki M, et al. Intraductal papillary mucinous tumors of the pancreas: thin-section helical CT findings. *AJR Am J Roentgenol* 2000;174:441–7.
9. Yamaguchi K, Tanaka M. Radiologic imagings of cystic neoplasms of the pancreas. *Pancreatology* 2001;1:633–6.
10. Chen F, Frankel WL, Wen P. Pathologic quiz case: a multicystic mass of the pancreatic body in a 70-year-old man. Serous microcystic adenoma of the pancreas. *Arch Pathol Lab Med* 2004;128:e83–5.
11. Khurana B, Mortele KJ, Glickman J, et al. Macrocystic serous adenoma of the pancreas: radiologic–pathologic correlation. *AJR Am J Roentgenol* 2003;181:119–23.
12. Scott J, Martin I, Redhead D, et al. Mucinous cystic neoplasms of the pancreas: imaging features and diagnostic difficulties. *Clin Radiol* 2000;55:187–92.
13. Civello IM, Frontera D, Viola G, et al. Cystic neoplasm mistaken for pancreatic pseudocyst. *Hepatogastroenterology* 1996;43:967–70.
14. Brugge WR, Lauwers GY, Sahari D, et al. Cystic neoplasms of the pancreas. *N Engl J Med* 2004;351:1218–26.
15. Brian KP, Tan YM, Tan PH, et al. Mucinous nonneoplastic cyst of the pancreas: a truly novel pathological entity? *World J Gastroenterol* 2005;11:2045–7.
16. Hashimoto M, Watanabe G, Miura Y, et al. Macrocystic type of serous cystadenoma with a communication between the cyst and pancreatic duct. *J Gastroenterol Hepatol* 2001; 16:836–8.
17. Shinozaki K, Yoshimitsu K, Honda H, et al. Abdominal cystic tumors containing small amount of fat in the septa: report of two cases. *Abdom Imaging* 2001;26:315–8.
18. Daltrey IR, Johnson CD. Cystic lymphangioma of the pancreas. *Postgrad Med J* 1996;72:564–6.
19. Fan YC, Shih SL, Yan FS, et al. Cavernous lymphangioma of the pancreas: a case report. *Pancreas* 1995;11:104–5.
20. Viola G, Frontera D, Bellantone R, et al. Lymphangiomas of the pancreas: a case report. *Pancreas* 1997;14:207–10.
21. Abe H, Kubota K, Noie T, et al: Cystic lymphangioma of the pancreas: a case report with special reference to embryological development. *Am J Gastroenterol* 1997;92: 1566–7.
22. Gray G, Fried K, Iraci J. Cystic lymphangioma of the pancreas: CT and pathologic findings. *Abdom Imaging* 1998;23: 78–80.
23. Titus RK, Evelyn ML, Gary JW, et al. Cystic lymphangioma of the pancreas. *AJR Am J Roentgenol* 2001;177:1090.
24. Schneider G, Seidel R, Altmeyer K, et al. Lymphangioma of the pancreas and the duodenal wall: MR imaging findings. *Eur Radiol* 2001;11:2232–5.
25. Igarashi A, Maruo Y, Ito T, et al. Huge cystic lymphangioma of the pancreas: report of a case. *Surg Today* 2001;31: 743–6.
26. Casadei R, Minni F, Selva S, et al. Cystic lymphangioma of the pancreas: anatomoclinical, diagnostic and therapeutic considerations regarding three personal observations and review of the literature. *Hepatogastroenterology* 2003;50: 1681–6.