Cystic lymphangioma of the pancreas : a rare observation

A. Ouazzani[1], P. Gruselle[2], B. Ramdani[2], M. Ceuterick[1], M. Idrissi[1], W. Boudaka[1]

Departments of (1) Digestive surgery and (2) Gastroenterology, Charleroi University Hospital, Charleroi, Belgium.

Abstract

Cystic lymphangioma of the pancreas is a rare benign and a slow growing lesion. The diagnosis remains a challenge for even the most experienced clinicians. We report a case of a pancreatic lymphangioma in a 31-year-old female complaining of epigastric pain, nausea, constipation and urticaria several weeks ago ; and we discuss diagnosis, anatomopathological findings and therapeutic attitude. (Acta gastroenterol. belg., **2006**, 69, **330-331**).

Keys words : pancreas, cystic lymphangioma.

Dear Sir,

Lymphangioma is a rare benign tumour which occurs more frequently in paediatric age at the head, neck and axilla. Pancreatic localisation accounts for less than 1% of all cases (1) and appears predominantly in women (2). The diagnosis of Pancreatic Lymphangioma (PL) remains a challenge for even the most experienced clinicians.

Case report

A 31-year-old female patient, Algerian, complained of epigastric pain, non-related to meals and without any radiation in the back, nausea, constipation and urticaria several weeks ago. Physical examination was normal. Upper gastrointestinal endoscopic evaluation was also normal. Abdominal ultrasound (US) showed a multilocular retroperitoneal cyst while the abdominal computerized tomography (CT) revealed an 86 mm diameter unilocular cyst adjacent to the pancreas and compressing the left kidney. Haemoglobin level was 13.2 g/dl (12-16 g/dl), white blood cell count was $6.4*10^9$/l (4.5-$10*10^9$/l), eosinophilic count was $0.23*10^9$/l (0-$0.60*10^9$/l) and the carcinoembryonic antigen (CEA) was 8.1 U/ml (< 35 U/ml). The other blood tests, serum amylase and lipase level were all within the normal limits. Echinococcal serological tests were negative. Echoendoscopy described a large cystic multilocular retropancreatic lesion, containing homogeneous fluid. In order to delineate the exact relationship between the cyst and the abdominal organs, a Magnetic Resonance Imaging (MRI) was performed, confirming the presence of a multiloculated cyst with internal septa bulging anteriorly the pancreas and the splenic vein, without any connections with both the liver and the kidney (Fig. 1).

At this point, the patient underwent surgery for either neoplastic or hydatid cyst of the pancreas.

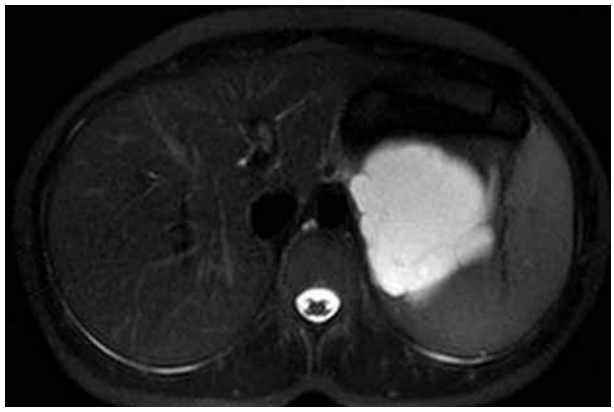

Fig. 1. — Abdominal MRI revealed a cystic lesion with many internal septa, independent of the left kidney and liver.

At laparotomy, a large tumour was palpated in the epigastric level. After dissection of the gastrocolic ligament and omental bursa, the cyst was clearly visualized. Because of the technical difficulties to unblock this lesion due to its close adherence to the pancreas, a peroperative puncture was done. On the possibility of being faced with hydatid cyst, hypertonic serum was introduced in the abdominal cavity. The lesion was found to contain surprisingly a chylous fluid. A complete resection of the cyst was then performed and the abdominal wall was closed. The postoperative course was uneventful.

Histological sections of the cyst wall revealed macroscopically a multilocular cyst and microscopically a dense collagenous tissue with some fascicles of smooth muscle lined by flat endothelial cells. Large lymphatic channels were also observed. In addition, abundant aggregates of lymphocytes were found on lymphoid follicle in many localisations. On immunohistochemistry, strongly endothelial cell positivity for CD34 (Fig. 2) and CD31 was observed whereas calretinin was negative. The diagnosis was cystic lymphangioma of the pancreas. No adjuvant therapy was administered and the patient has been followed without any recurrence for one and a half year.

Corresponding author : Aous Ouazzani, Route de Gozée, 706, BP 91, 6110 Montigny-le-Tilleul, Belgium. E-mail : aouazzan@ulb.ac.be.

Submission date : 30.06.2006.
Acceptance date : 09.07.2006.

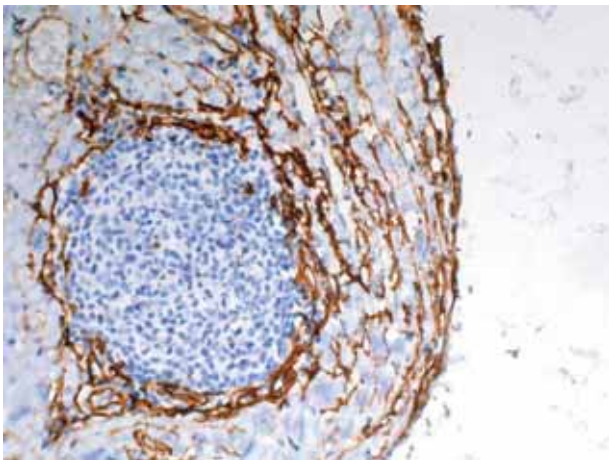

Fig. 2. — Immunohistochemical study of the endothelial cells lining the lymphatic channels showed positivity for CD34.

Discussion

First described by Koch in 1913 (3), PL is an uncommon slow growing mass with uncertain histogenesis. Embryologic, traumatic or inherited origins were proposed to explain the occurrence of this lesion. At the moment, the most accepted theories suggest that PL arises from lymphatic congenital malformations during the second and third months of foetal life (2,3,4). Pancreatic origin is considered, if the lymphangioma arises from the pancreatic parenchyma, connected by pedicle or adjacent to the pancreas (2). The clinical presentation is variable ; abdominal pain, vomiting and nausea can be the principal symptoms. Torsion of the pedicle, intracystic haemorrhage and cystic rupture are responsible of an acute abdominal symptomatology.

Several differential diagnoses of PL including begin and malignant lesions must be considered (pseudocyst, adenoma, cystadenocarcinoma, mucinous cyst and hydatid cyst for instance). Although, cystic pancreatic lesions may easily be found by US and CT, but the diagnosis of PL remains difficult to establish preoperatively.

US showed classically a cystic mass with internal septations or scattered internal echoes ; and CT determines cystic walls boundaries and homogeneous unenhancing low-density (5,6). In the present report, intracystic septa were visualized by US and MRI. According to the authors (7,8), CT can show intracystic septa surrounded by regular or calcified walls. MRI is an interesting tool to complete anatomical study of the cystic morphologies and its relationship with the other abdominal organs.

Recently, PL was diagnosed following a cystic fluid studies (chylous compound with high triglyceride level) after a fine needle puncture by endoscopic US, avoiding surgery in asymptomatic patient (9). However, fluid analysis can not always provide a precise diagnosis (3), and because of the risks of disseminating neoplastic cells or parasites and bleeding, this approach will be performed only in some selected cases under antibiotic prophylaxis to minimize infectious complications.

The mainstay treatment is a complete surgical excision of the cyst (1,2,3,10). Definitive diagnosis of PL is established after pathological study of the resected specimen. Usually, the wall contains abundant collagenous tissue lined by flattened endothelial cells, irregular fibres of smooth muscle, lymphatic channels and irregular aggregates of lymphoid cells. Immunohistochemical studies (2,3) demonstrate positivity of the endothelial cells for factor VIII-related antigen and CD31 while the CD34 can be negative.

References

1. KOENIG T.R., LOYER E.M., WHITMAN G.J., RAYMOND A.K., CHARNSANGAVEI C. Cystic lymphangioma of the pancreas. *AJR*, 2001, **177** :1090.
2. PAAL E., THOMPSON L.D., HEFFESS C.S. A clinicopathologic and immunohistochemical study of ten pancreatic lymphangiomas and a review of the literature. *Cancer*, 1998, **82** : 2150-58.
3. CASADEI R., MINNI F., SELVA S., MARRANO N., MARRANO D. Cystic lymphangioma of the pancreas : anatomoclinical, diagnostic and therapeutic considerations regarding three personal observations and review of the literature. *Hepato-Gastroenterology*, 2003, **50** : 1681-86.
4. ABE H., KUBOTA K., NOIE T., BANDAI Y., MAKUUCHI M. Cystic lymphangioma of the pancreas : a case report with special reference to embryological development. *AJG*, 1997, **92** : 1566-67.
5. CUTILLO D.P., SWAYNE L.C., CUCCO J., DOUGAN H. CT and MR Imaging in cystic abdominal lymphangiomatosis. *J. Comput. Assist. Tomogr.*, 1989, **13** : 534-36.
6. GOH B.K., TAN Y.M., OOI L.L. Hepatobiliary and pancreatic : Cystic lymphangioma of the pancreas. *J. Gastroenterol. Hepatol*., 2006, **21** : 618.
7. FERROZZI F., CUSMANO F., ZUCCOLI G., TOGNINI G., BASSI S., GABRIELLI M. Mesenchymal tumors of the pancreas : computed tomography patterns. *Radiol. Med. (Torino)*, 1999, **98** : 295-99.
8. GRAY G., FRIED K., IRACI J. Cystic lymphangioma of the pancreas : CT and pathologic findings. *Abdom. Imaging*, 1998, **23** : 78-80.
9. JATHAL A., ARSENESCU R., CROWE G., MOVVA R., SHAMOUN DK. Diagnosis of pancreatic cystic lymphangioma with EUS-guided FNA : report of a case. *Gastrointest. Endosc.*, 2005, **61** : 920-22.
10. DE PERROT M., ROSTAN O., MOREL P., LE COULTRE C. Abdominal lymphangioma in adults and children. *Br. J. Surg.*, 1998, **85** : 395-97.