Lymphoepithelial cyst of the pancreas : case report and review of the literature

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Abstract

Background and study aims : **Lymphoepithelial cyst of the pancreas (LCP) is a rare, benign cyst mimicking pseudocyst or cystic neoplasm. Literature describing LCP is limited to case or brief series reports, and the natural history of this condition is largely unknown. A literature review was carried out in order to elucidate the clinical, pathological and biochemical features of LCP. The aim of this study was to define diagnostic criteria and treatment.**

Methods : **A Medline and Pubmed search was conducted by using the key-words "lymphoepithelial cyst" and "pancreas". The articles found were accurately examined and all details regarding clinical and pathological features were included in a data-base. Furthermore, a case recently observed in our unit was added to the review.**

Results : **Ninety-two cases of LCP were found in the worldwide literature, including the case that we observed. LCP occurs more frequently in males (M:F = 5.5:1), its preferred site is the tail of the pancreas, and its size ranges between 2 and 10 centimetres. Histologically, it is a true cyst delineated by a keratinizing squamous epithelium surrounded by lymphoid tissue. LCP is asymptomatic in the majority of cases and preoperative diagnosis is complicated by a lack of specific radiological features of the disease. An accurate preoperative diagnosis can only be made by obtaining cytological specimens and placing them in the hands of a pathologist who is familiar with the cytological appearances of the disease.**

Conclusions : **LCP is a rare lesion worldwide, without any prevalence in different countries or in different ethnic groups. Understanding the features of LCP, making an accurate diagnosis and differentiating it from cystic neoplasm preoperatively is vital, as when it is diagnosed certainly, a conservative treatment is justified. Otherwise, radical surgery in the form of pancreatic resection is required to exclude the diagnosis of pancreatic cystic neoplasm.** (Acta gastroenterol. belg., **2011**, 74, **343-346**).

Key words : lymphoepithelial cyst, pancreas, pancreatic cyst, pancreatic surgery.

Introduction

Lymphoepithelial cyst of the pancreas (LCP) is a rare, benign cyst mimicking pseudocyst or cystic neoplasm of the pancreas. It was first described in 1987 by Truong (1) and he also coined the term LCP. Macroscopically, it is a true cyst filled with pasty, keratinous material and histologically, the wall of the cyst consists of a keratinizing squamous epithelium surrounded by lymphoid tissue. The differential diagnosis of LCP is a pancreatic cystic mucinous neoplasm (PCMN) and this is of clinical interest, because LCP can be managed conservatively, whereas the management of PCMN entails radical surgery. The literature on LCP is limited to reports of single or small numbers of cases (2,3) and its natural history is largely unknown. However, there has never been a report of the transformation of LCP into a malignancy or local recurrence after resection. We reviewed the literature to assess clinical and pathological characteristics of this uncommon pancreatic disease. A case of LCP, recently presenting and treated as a PCMN at our unit, was added to the review.

Methods

We conducted Medline and Pubmed searches for all articles using the key-words "lymphoepithelial cyst" and "pancreas". All articles found were closely examined ; papers without detailed pathological or clinical data were considered unsuitable for this research and were excluded. For each article, the following details were recorded and included into the data base :

- Country of the Author/s
- Number of case/s reported ;
- Age and sex of patient/s ;
- Size of LCP ;
- Site of the lesion (head, body or tail of the pancreas) ;
- Presenting symptoms ;
- Relevant laboratory findings (tumour markers) ;
- Diagnostic means used and preoperative diagnosis ;
- Performed treatment and outcome.

Case report

The following case was managed in our unit and has been included in the analysis :

A 62-year-old man without any significant past medical history, underwent occasional abdominal ultrasound scan (US). This revealed a solid/cystic mass occupying the body and tail of the pancreas. There were no stones in the gallbladder and he had no history of pancreatitis. A computerized thomography (CT) scan was performed to further characterize the lesion, and a 10 cm cystic mass with an enhancing thin wall and an internal daughter cyst was found. Laboratory tests revealed a Ca 19-9 of 74 U/mL (reference range < 37) and a diagnosis of PCMN was made on the basis of the

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Submission date : 13/09/2009
Acceptance date : 07/09/2010

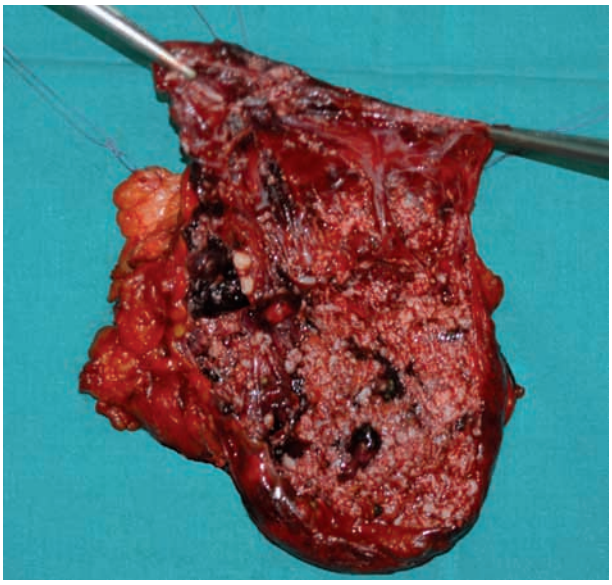

Fig. 1. — Surgical specimen (spleen-preserving left pancreatectomy) : the cystic lesion is opened ; it contains a pasty, keratinous material.

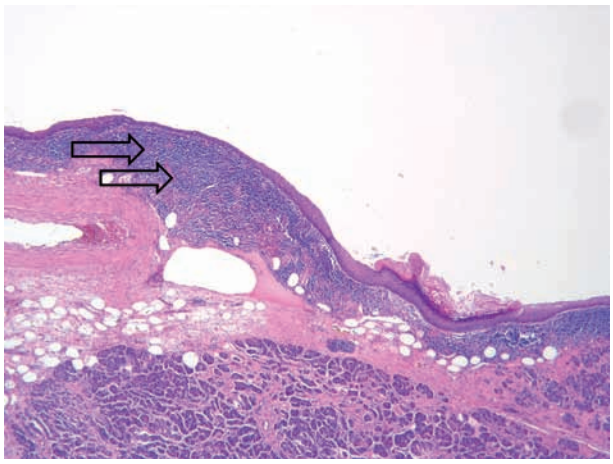

Fig. 2. — Section of the cyst : keratyn-filled cyst lined by mature keratinizing epithelium overlying a layer of lymphoid tissue (arrows) (H&E, original magnification ×40).

radiological findings. The patient subsequently underwent left pancreatectomy with preservation of the spleen. Gross inspection of the surgical specimen revealed an approximately 10 × 10 cm cyst protruding from the tail of the pancreas and filled with white, pasty material (Fig. 1). Microscopically, the cyst was lined by a mature keratinising squamous epithelium overlying a layer of lymphoid tissue with occasional germinal centers (Fig. 2). On the basis of the histological examination, the cystic lesion was diagnosed as a LCP.

Results

Between November 1987 and June 2009, sixty-six papers concerning LCP were published in the Literature. 24 reports were from American Authors, 24 from Asiatic Authors and 18 from European ones. Single cases were reported in 58 papers : 2 cases were reported in 4 papers (4,5,6,7) ; 4 cases (8), 9 cases (9) and 12 cases (10) were reported, respectively, in a single paper. Ninety-two cases were reported in total, including the case described in this article.

Among the 92 patients with LCP, 78 were males and 14 females with a 5.5:1 males/females ratio. The average age of patients at the time of diagnosis was 53.6 years (range from 20 to 74 years) ; in the majority of cases the disease occurred between the fifth and seventh decade of life (Table 1). The most common site was the tail of the pancreas (61.95%) followed by the head (19.56%) and the body (18.47%). The average dimension of the lesion was 4.32 cm (range from 2 to 10 cm).

Among the 92 patients, 49 (53.26%) were asymptomatic and LCP was incidentally diagnosed during an US or a CT performed for other reasons. In several cases, the diagnosis was made postmortem (11,12). The range of

Table 1. — **Distribution for age of the reported cases of LCP**

Age (years)	Number of cases reported
0-10	–
11-20	1
21-30	–
31-40	4
41-50	22
51-60	30
61-70	29
71-80	6
Total	92

presenting symptoms included : abdominal pain (30.43%), back pain (7.6%), diarrhoea (4.34%), vomiting (2.71%) and weight loss (2.17%). In all cases reported, US, CT and more recently, Magnetic Resonance (MR) scans were used in the diagnostic work-up. The radiological appearances of LCP at US and CT scan are different. At US, LCP is usually demonstrated as a uni- or multi-locular hypoecoic mass containing granular hyperecoic areas. At CT, LCP appears as a cystic mass with an enhanced thin wall and containing internal septa, daughter cysts or papillary projections (13). Differentiating between LCP and pancreatic cystic neoplasm on the basis of radiology is extremely difficult, because their radiological features are similar. This means that radiological investigation alone cannot be relied upon in making an accurate diagnosis of LCP preoperatively. In some cases, high serum levels of the tumour markers CEA and Ca 19-9 were reported. In one case, CEA levels were greater than 5.000 nanograms/millilitre (ng/ml) (14) ; in another case, Ca 19-9 levels were 8.100 International Unit per litre (IU/L) (15). High values of CEA and Ca 19-9 were also reported in the intracystic fluid (8,15). A confident diagnosis, before surgery, was obtained only from fine needle aspiration (FNA) cytology. Among the 92 patients, this analysis

Table 2. — **FNA cytology performed for LCP**

Author	N. patients submitted to FNA	Correct diagnoses
Mitchell (16)	1	
Cappellari (17)	1	
Rino (18)	1	
Mandavilli (19)	1	1
Liu (7)	2	2
Zou (20)	1	1
Bolis (21)	1	1
Renou (22)	1	
Policarpio-Nicolas (8)	4	1
Jian (23)	3	2
Nasr (9)	9	6
Total	25	14 (56%)

was performed in 25 cases (Table 2) and an accurate diagnosis was obtained in 14 patients. The typical cytological appearance includes a mixture of squamous cells and small, mature lymphocytes in a background of keratinaceous debris, anucleated squamous cells and multinucleated histiocytes.

In the absence of a correct diagnosis, most of the patients with LCP were treated surgically. Sixty patients underwent surgical resection : 34 left pancreasectomies (in 9 cases with spleen preservation), 22 local excisions and 4 Whipple's procedures, were performed.

Discussion

During recent years, cystic lesions of the pancreas have been diagnosed increasingly because of the widespread use of imaging techniques (US, CT, MR). Nevertheless, making the distinction between benign or malignant pancreatic cysts based on radiological findings is not often possible. LCP represents a rare lesion worldwide without any prevalence in different countries or in different ethnic groups. Fully understanding the features of this disease is essential in achieving an accurate diagnosis and consequently, avoiding unnecessary pancreatic resection. LCP is usually localized to the tail of the pancreas, but it can also be found in the head or in the body of the pancreas, with its size ranging from 2 to 10 cm. It usually occurs between 41 and 70 years of age with a males/females ratio of 5.5:1. LCP is asymptomatic in the majority of cases, but it can occasionally cause non-specific abdominal pain and other symptoms such as diarrhoea, weight loss and back pain. Currently, cystic lesions of the pancreas are diagnosed by US or CT scans and are either found incidentally, or when the scans are done because of the occurrence of symptoms. The radiological features of LCP and PCMN lesions are similar and they cannot be reliably differentiated from each other based on radiological findings. Levels of tumour markers found in the serum and cystic fluid are also unhelpful in aiding the diagnosis. Differentiating between these two conditions is important as it can avoid the need for unnecessary radical surgery. Conversely, obtaining cytological specimens using endoscopic US (EUS)-guided or CT-guided FNA allows a correct diagnosis especially when the pathologist has expertise in the cytological features of the disease. Liu (7) reported two cases in which a pre-operative cytopathological diagnosis of LCP, obtained by CT-guided or EUS-guided FNA, was subsequently confirmed following surgery. He concluded that preoperative cytological diagnosis of LCP, could allow a conservative surgical management strategy. Policarpio-Nicolas (8) reported four cases of suspected LCP submitted to an EUS-guided FNA ; one patient only had cytological findings consistent with LCP and was treated conservatively. More recently, Nasr (9) reported 9 patients who underwent EUS-FNA : six patients avoided surgery on the basis of cytologic results. The differential diagnosis includes some benign lesions such as dermoid cyst, or malignant lesions, such as adenosquamous carcinoma and metastatic squamous carcinoma. However, these lesions show cytologic features of malignancy and differential diagnosis should not be difficult. In our opinion, in all cystic pancreatic lesions it is mandatory to obtain an EUS-guided sample of the cyst with analysis of the fluid for amylases, CEA, mucus and cytology. Familiarity with the cytological features of LCP allows a correct diagnosis and permits a conservative management (8,9).

In our patient, diagnosis of PCMN was made after radiological examination and, consequently, the patient underwent a pancreatic resection. Subsequently the diagnosis of LCP was made postoperatively on the basis of histological findings. If a certain diagnosis of LCP can be achieved preoperatively, the condition can safely be managed conservatively, as there has never been a report of the transformation of LCP into a malignancy, or local recurrence following resection. If the diagnosis remains uncertain, a pancreatic resection is justified to exclude the diagnosis of PCMN.

Acknowledgment

The authors thank Miss Sarah Louise Schofield MD for the English editing of this manuscript.

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