Case report

Cystic Lymphangioma of the Pancreas: About A Case and Literature Review

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Summary

Cystic lymphangioma is a rare malformative benign tumour of the lymph vessels, its histogenesis is still hypothetical. This tumour is mainly found in child’s neck and axillary, the pancreatic location remains exceptional and represents less than 1% of whole lymphangiomas, and occurs more frequently in women and especially in the left pancreas.

Its clinical presentation is polymorphic; however, the diagnosis can’t be specifically evoked by imaging and requires therefore histological confirmation. Surgery remains the only curative treatment.

We report in this work the case of an incidentally discovered cystic lymphangioma of the pancreatic tail in a 58-year-old woman. The histological study of the surgical specimen concluded on a benign cystic lymphangioma.

On later surgical outcomes, the patient developed a pseudokyst of the remaining pancreas and today she has not had any recurrences.

The aim of this article is to share our experience in the management of this case and to review the clinical and therapeutic aspects of this rare pathology.

Keywords: Cystic Lymphangioma, Pancreatic cystic tumours

Introduction

Cystic lymphangiomas are rare tumours that are usually benign but can be locally invasive and can be found in all anatomical sites. They most often occur in the neck, axillary hollow and mediastin, especially in children. Intra-abdominal locations remain very rare and pancreatic cystic lymphangiomas remains exceptional and accounts for only 1% of all lymphangiomas. The diagnosis of cystic lymphangioma of the pancreas is most often carried in an intraoperative and after histological study of the resected specimen due to the lack of clinical, biological and radiological specificity, thus the diagnosis of cystic lymphangioma of the pancreas remains above all a diagnosis of elimination. Only surgical resection of the tumour is associated with prolonged survival and a favorable prognosis. We report in our work the case of a 58-year-old patient with an incidental cystic lymphangioma of the pancreas.

Case report

A 58-year-old patient with a history of recently discovered diabetes under oral anti-diabetics, rheumatoid arthritis initially on corticosteroid and immunosuppressants and then treated by biotherapy. The patient being asymptomatic, the tumour was discovered incidentally at the end of a pre-therapeutic assessment during which a thoracic CT scan (Scanner) revealed an hypodense pancreatic mass presenting lobulated contours, central calcification and measuring 11 cm of its large axis. Subsequently, the patient was referred to the visceral surgery department of HMIMV (Mohamed V Military Training Hospital) for therapeutic management. On physical examination, the abdomen was supple, painless and mass free.

The abdominal CT scan, with contrast injection, revealed a body and tail pancreatic mass. This well-limited and encapsulated fleshy and polylobated mass was heterogeneous and containing small irregular micro calcifications. It seems intimately linked to the posterior edge of the stomach, although it respects the left bowel, the pre-renal fat, the portal and celiac vessels. The intra-hepatic bile ducts and the main bile duct are not dilated. (Figures 1.2 and 3)
The patient also benefited from a pancreatic MRI (Magnetic Reasoning Imaging) that revealed a large confluent polycystic mass of the pancreatic tail, showing polylobate contours and measuring approximately 113x93 mm. This mass was presenting hyposignal in T1 (Time one) sequence and hypersignal with an hyposignal central area in T2 (Time two) sequence. After Gadolinium injection, heterogeneity was accentuated with radial contrast and sharp individualization of the late-rise central fibrous area. This process pushed back the splenic vein. The pancreatic pathways were free, the peri-pancreatic fat was respected and there were no dilation of extra-hepatic bile duct and no peritoneal effusion (Figure 4 and 5).

The biological finding (blood formula count, liver and pancreatic balance) was normal and the tumour markers (CA-125, CA19-9 and ACE) were normal.

A complete resection of the mass was achieved by a caudal spleenpancreatectomy.

The pathological study of the specimen showed on the macroscopic examination, a 14 cm large cyst with a multi-cystic appearance presenting clear content. Microscopic examination showed a benign cystic tumour proliferation of dilated and cystic vascular structures bordered by non-atypical regular endothelial
cells (Figure 6 and 7). The immunohistochemical study was not used though the diagnosis was obvious.

Figure 6: Histological Cup: Benign Cystic Tumor Proliferation of Dilated Vascular Structures (HE, GEX50) (HMIMV)

Figure 7: Histological image: Endothelial cells lining these structures are flattened regularly and without atypies (HE, GEX400) (HMIMV)

Immediate surgical follow-up was simple, the patient resumed feeding at day one and drains were removed on the third day. 15 days after the patient presented an angina type abdominal pain. An abdominal CT scan revealed a false cyst of the remaining pancreas and an extensive mesenteric venous thrombosis. Heparin was started at curative dose and given the good clinical evolution; the patient went home on oral anticoagulant treatment. Three-month check-up, by an abdominal CT scan, showed a free mesenteric vein and the regression of the false cyst.

Discussion

Cystic lymphangioma is a benign rare tumour of the lymphatic vessels. Its histogenesis is still hypothetical [3] and the most probable hypothesis of the development of a cystic lymphangioma is a congenital malformative origin [2].

Mainly found in children in the neck and axillary region; the pancreatic location remains exceptional and represents less than 1% of lymphangiomas [2]. In adults, it occurs more frequently in women and especially in the left pancreas (body and tail) [3].

The clinical presentation is polymorphic and nonspecific [4]; Cystic lymphangioma of the pancreas is generally symptomatic in 92.2% of the cases described [5], according to published articles and most often manifests itself by abdominal pain, epigastric and nonspecific seat, ranging from a simple abdominal discomfort to a more intense and chronic pain. Other clinical signs also remain no less specific and are often due to digestive compression; nausea and vomiting have been reported in several patients. Cystic lymphangioma of the pancreas can also be revealed by jaundice secondary to bile duct compression [4]. Rarely, complications such as: a rupture, an haemorrhage [7], a twist or cyst infection; reveals the diagnosis [8].

As other pancreatic tail process; Diabetes can be revealing. A recently discovered diabetes in our case was an important sign that leads us to the diagnosis. In some cases the tumour may remain asymptomatic and be discovered incidentally during a radiological examination [9]. The imaging (ultrasound, abdominal CT scan and MRI) is not specific but allows a diagnosis orientation, the diagnosis therefore requires histological confirmation [10,11]. The natural evolution of cystic lymphangiomas of the pancreas is towards the persistence of the tumour. Although they are considered benign these tumours are invasive, they can increase in size to a diameter of 20 cm [11] and infiltrate adjacent organs thus increasing the risk of complications: rupture, twisting, haemorrhage, cyst infection or vascular, digestive or bile duct compression [11]. No cases of malignant transformation have been reported [1]. The treatment of choice is surgical. Complete exeresis of the tumour is curative for cystic lymphangiomas of the pancreas. Only incomplete exeresis is associated with a risk of reoffending [12]. The surgical exploration of the pancreatic tail, guaranteed a complete resection of the cystic lymphangioma. Out comes are simples for light surgical procedures, and there is no risk of recurrence if the resection is radical with healthy resection margins, no cases of tumour recurrence have been reported in all cases described, however if tumoral resection is not complete the patient remains exposed to recurrences [5,11,12,13,14].

Conclusion

Cystic lymphangioma of the pancreas is an extremely rare benign malformative tumour developing at the expense of the lymphatic system. Its pancreatic location is exceptional and diagnosis is often late and difficult to establish in preoperative period because of its polymorphic clinical expression. Therefore the diagnosis is always established after surgical resection and histological study.

Inaugural diabetes in adult should lead to a morphological exploration of the pancreas as not to miss a cystic tumour of the asymptomatic pancreas. Cystic lymphangioma of the pancreas should therefore be considered and evoked in front of any cystic mass of the pancreas especially in women. The prognosis of this tumour is often favourable with a cure after radical surgical treatment.

Ethics approval and consent to participate

For our article, we had the approval of an ethics committee and the written and signed consent of the patient.

We are sending you this signed consent as an attachment.

List of abbreviations

CT Scan: Scanner
HMIMV: Mohamed V Military Instruction Hospital
MRI: Magnetic Reasoning Imaging
CA-125: Cancer antigen 125
CA-19.9: Cancer antigen 19-9
ACE: Carcinoembryonic antigen

Data Availability

The data used for this work are available in the database stored at the Rabat Military Hospital. These data can be accessed upon request.

Conflicts of Interest

No conflict of interest was reported

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Authors’ contributions

AK reviewed the literature and wrote the final document. AM reported the clinical case and improved the final document. BM, NM, KH and BA made final corrections. All authors read and approved the final manuscript.

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