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Retroperitoneal Cystic Lymphangioma : case report

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ABSTRACT

Introduction

Lymphangiomas, cystic hygroma or macrocystic lymphatic malformation are a rare and benign congential malformations of the lymphatic system. Adults are affected rarely and less than children, and the abdominal location even less.

Presentation of case

Here, we discuss the case of a 56years old woman admitted and operated for a 7cm cystic mass of the tail of the pancreas, whom per operative exploration showed a retroperitoneal polycystic mass and histopathology concluded a lymphangioma.

Discussion

Less than 1% affect the retroperitoneum, they typically occur in spaces surrounded by loose connective tissue, the are difficult to differentiate from a mucinous cystic neoplasm of the pancreas by imaging since both have the appearance of multilocular cysts. Ultrasonography is very sensitive and relatively specific for evaluation of abdominal cystic masses. Laparoscopic removal has certain advantages over classical laparotomy, complete excision is recommended to protect the patient from subsequent recurrences even though cystic lymphangiomas are low-grade malignancies.

Conclusion

The diagnosis of CL often is facilitated by means of modern imaging. Differential diagnosis include pancreatic pseudocysts, mucinous and serous cystadenomas, other congenital cysts and pancreatic ductal carcinoma with cystic degeneration. If symptomatic lesions or complications arise, complete surgical excision is a must.

Key Words: Lymphangioma, retroperitoneal cyst, surgical cyst resection

INTRODUCTION

Lymphangiomas are rare, also called cystic hygroma (CH) or better macrocystic lymphatic malformation (LM), benign and congential malformations of the lymphatic system. This disease is most common in children under 2 years of age (80–90%) with a 1.2–2.8 per 100 000 incidence rate [3]. Adults are instead rarely affected [4]. primarily found in the neck and axilla (95%), [1] The abdominal cavity is a rare site of origin, and most cases that have been reported were developed in the retroperitoneum or mesentery. [2]

AIM OF THE ARTICLE:

The aim of this study is to describe our experience with a retroperitoneal cystic lymphangioma in a female adult patient, to highlight the role of this rare entity in the differential diagnosis and to present a review of the litterature.

PRESENTATION OF CASE

We present a 56-year-old female patient, followed for an invasive breast carcinoma of the right breast, having benefited from a mastectomy with lymph node surgery followed by radio-chemotherapy, admitted in our structure for a heavines-like pain straddling between the epigastrium and the left hypochondria, without any other associated functional signs.

The clinical examination found a patient with a performance status at 0, soft abdomen, no exquisite sensitivity, pelvic examination was unremarkable.

An abdominal CT-scan showed a cystic mass encompassing the tail of the pancreas, of rather well limited polylobate contours, not enhanced after contrast injection measuring 78x77.5mm.

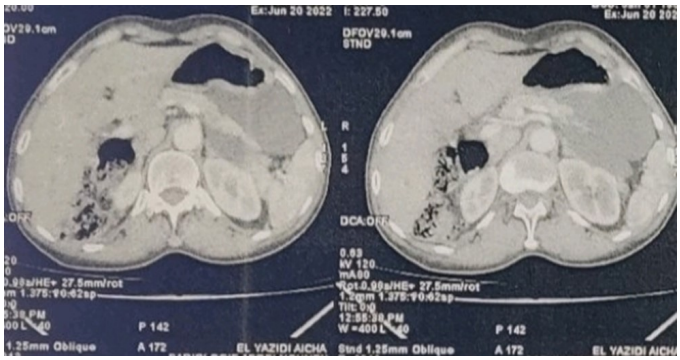

Figure1: a CT-scan slide showing the retroperitoneal cystic mass.

An abdominal MRI was performed, objectifying a retroperitoneal cyst measuring 66.5x97.5mm.

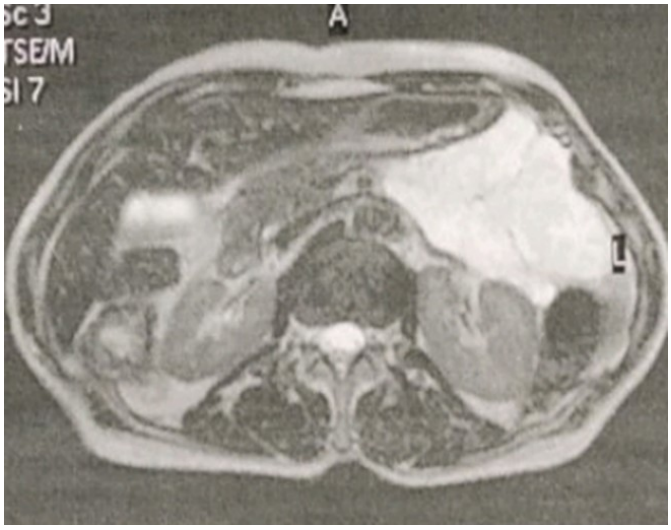

Figure2: an MRI slide showing the retroperitoneal cystic mass.

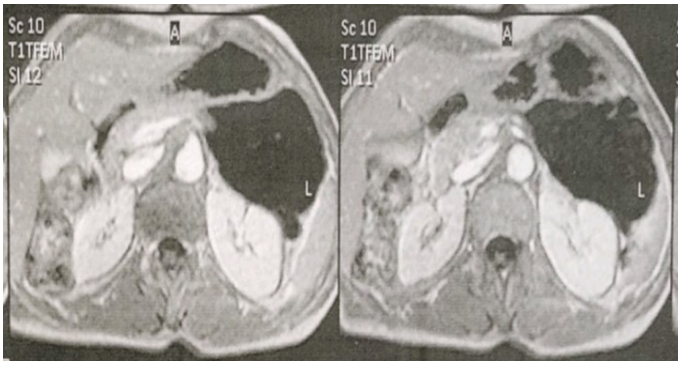

Figure3: an MRI slide showing the retroperitoneal cystic mass.

The patient was operated having benefited from a resection of retroperitoneal polycystic mass in which the exploration found a polycystic mass of 20cm of diameter at retroperitoneum adhering to the tail of the pancreas and to the transverse mesocolon with the presence of a plane of cleavage.

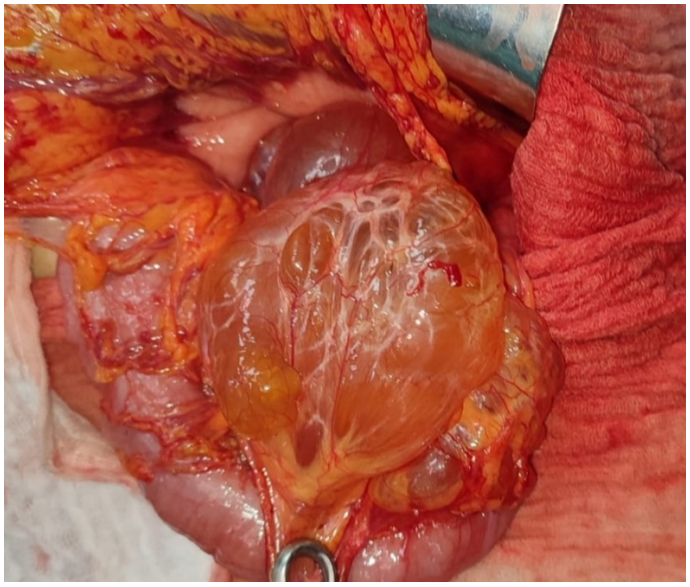

Figure4: Per-operatoire image of the cystic lymphangioma.

Histopathological analysis found a cystic lymphangioma.

Post-operative follow-up was unremarked, with the reported patient exiting at 05 post-operative days.

DISCUSSION:

Cystic lymphangiomas can be located in any part of the body except the brain[5]. They are most frequently single, but multiple lymphangiomatous cysts can affect a single organ -[10], a single region such as the abdominal or retroperitoneal cavity or both[11], or the disease can be generalized [12] .Only a small proportion of cystic lymphangiomas are multilocular, and most contain a single cavity [13,15]. The tumors are usually located in the neck (75%, also called cystic hygroma), in the axilla (20%)[10,16,17,18,19,20,21,22,23,24]. Less than 1% affect the mesentery, greater omentum, and retroperitoneum [5,6,8,13,25]. Among all intra-abdominal lymphangiomas, 10% involve the mesocolon and 5% the retroperitoneum; the small bowel mesentery is affected more frequently [6,9,26] .

Pathologically, lymphangiomas are subdivided into three main types: capillary, cavernous and cystic. The first two are predominantly cutaneous lesions; cystic lymphangiomas are generally found in the abdomen and retroperitoneum, that was the case for our patient.

Solid lymphangiomas are occasionally found in the abdominal cavity. [27] Cystic lymphangiomas typically occur in spaces surrounded by loose connective tissue such as the mesentery and retroperitoneum.[5,6]

Based on their specific histology, cystic lymphangiomas are classified as hamartomas.[13,14] The cystic spaces are lined with a single layer of endothelium; there are small lymphoid aggregates in the cyst wall that aid in distinguishing lymphangiomas from simple cysts of the mesentery.[5] If there is secondary bleeding into the cavity, lymphangiomas might be difficult to distinguish from hemangioma; the diagnosis can be established immunohistochemically.[28,29]

A cystic lymphangioma of the pancreas is difficult to differentiate from a mucinous cystic neoplasm of the pancreas by imaging since both have the appearance of multilocular cysts. Moreover, lymphangiomas and cystic pancreatic neoplasms are both rare and slow-growing, occur in similar age groups, and remain localized for a long time, making the differential diagnosis difficult. CT typically shows lymphangiomas as well-circumscribed, homogene-

ous, and unenhanced lesions with low density, while the ultrasonographic appearance is usually a cystic or multicystic mass. Cystic mesothelioma, lymphangiosarcoma, and myxoid degeneration of lymphangioma must be further included in the differential diagnosis.[30,31,32]

CL usually contain serous, serosanguinous, or chylous fluid.[5,6] The variability in fluid content might be explained by different degrees of lymph stasis, varying numbers of communicating channels within the lymphatic system, and the cyst fluid's protein content.[5,33] Serosanguinous fluid represents cyst hemorrhage; occasionally, the cyst fluid can be purulent because the cyst cavity is infected. [7]

Most patients are initially asymptomatic, with vague and obscure abdominal symptoms emerging early or late, depending on the cyst's size and location. The clinical symptoms are protean and include pain, nausea, vomiting, or alterations in bowel habits.[34] Retroperitoneal cystic lymphangiomas are discovered late. [5,7,13,15,36] A prominent and highly unusual feature of the present case is the adult presentation with a rapidly growing abdominal mass and severe abdominal pain.

Other acute complications include traumatic rupture,[35] intracavitary or intraabdominal bleeding, intestinal obstruction with necrosis, intestinal gangrene due to volvulus, and cyst infarction. All these result in an acute abdomen requiring emergency laparotomy. [7,14,36,37,38,39,40,41] Nearly 50% of patients present with ascites, in some patients, the CL can grow large enough to resemble ascites at physical exam.[34,42]

Ultrasonography is very sensitive and relatively specific for evaluation of abdominal cystic masses; the lesions appear as sharply defined cystic or multicystic formations, often with internal septations. The fluid can be anechoic, or it can have scattered internal echoes representative of cyst infection. Ultrasonography is also the modality of choice for postoperative followup.

CT and MRI can add important preoperative information regarding anatomical location, organ involvement, cyst size, and complications; they can better ascertain the attenuation coefficient of the cystic fluid, and can there by differentiate chylous fluid, blood, or pus, supporting the clinical findings. [36,43,45] CT and MRI best display solid components in the cyst, which aids the preoperative diagnosis and surgical planning. [44,45,47]

In our case, the CT exam showed a cystic mass encompassing the tail of the pancreas, of rather well limited polylobate contours, not enhanced after contrast.

Lymphangiography and radionuclide scintigraphy yield additional diagnostic information, but their use is restricted to elective evaluation of lymphangiomas. Diagnosis must be made promptly in emergency cases.[33] Though, our case patient did not benefit those additional exams.

The treatment of CL is surgical. Although the lesions have a generally benign course, they tend to recur and invade neighboring structures. Total removal of a lesion that invades potentially resectable intraabdominal structures such as bowel, spleen, or pancreas is possible, but adhesions to vital structures can make resection hazardous or even impossible. [13,34]

Laparoscopic removal has certain advantages over classical laparotomy, including a more precise dissection around the vessels, minimal trauma to the abdominal structures, less pain, and a shorter hospital stay. More experience is needed to demonstrate the superiority of this method over conventional techniques. [47]

Thus, complete excision is recommended since an incomplete resection may lead to subsequent recurrence even though cystic lymphangiomas are low-grade malignancies.

CONCLUSION:

The diagnosis of CL often is facilitated by means of modern imaging; however, other diagnosis may be discussed. Differential diagnoses include pancreatic pseudocysts, mucinous and serous cystadenomas, other congenital cysts and pancreatic ductal carcinoma with cystic degeneration. The final diagnosis is histological. If symptomatic lesions or complications arise, complete surgical excision, when possible without major sacrifice, seems to be the best therapeutic option.

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