UNUSUAL LOCATION OF CYSTIC LYMPHANGIOMA OF THE PANCREAS: ABOUT A CASE AND REVIEW OF THE LITERATURE

ABTRACT

Cystic pancreatic lymphangiomas are rare benign lesions that arise from the lymphatic vessels, representing only 1% of abdominal lymphangiomas and less than 0.5% of all cystic pancreatic lesions, generally affecting women.

Its clinical presentation is polymorphic, its positive diagnosis is now possible. However, the definitive diagnosis is histological.

The curative treatment is surgical, complete resection eliminates the risk of recurrence.

In this work, we report the case of a 69-year-old patient who presented to the emergency room with a picture of upper digestive hemorrhage where a fibroscopy, CT scan and abdominal MRI revealed a duodeno-pancreatic cystic mass circumscribing the head and the isthmus of the pancreas. A complete resection was impossible given the risks associated with total duodenopancreatectomy. Pathological examination of the surgical specimen confirmed the lymphangiomatous nature of the tumor.

The objective in this work is to study the diagnostic and therapeutic features of cystic lymphangioma of the pancreas.

KEYWORDS: Cystic lymphangioma, pancreas, cystic tumor.

INTRODUCTION

Lymphangiomas are dysplasias developed at the expense of lymphatic vessels [1].

The first case was described by Redenbacher in 1828, which subsequently allowed them to be classified into three types: capillary, cavernous and cystic [2].

These hamartomatous malformations are most often found in the cervico-facial region and rarely involve the abdomen [3]. Cystic mesenteric lymphangiomas represent 5 to 6% of all pediatric benign tumors [14].

Pancreatic involvement remains exceptional, presentation in adulthood is extremely rare.

Cystic lymphangiomas of the pancreas represent only 1% of abdominal lymphangiomas and less than 0.5% of all cystic lesions of the pancreas [4].

In fact, no more than 100 cases are reported in the literature [1].

These tumors result from lymphangiectasia, a consequence of a blockage of lymphatic flow. This may be associated with congenital malformations or obstructions resulting from an acquired process [1].

It is a benign condition for which the potential for malignancy has never been demonstrated [4].

Diagnosis is difficult preoperatively. Indeed, although imaging studies can suggest the diagnosis, a histopathological examination is necessary to establish the definitive diagnosis [5].

Curative treatment is surgical; total excision remains the only option for a more favorable prognosis [5,15,16,17,18].

In our work, we will report the case of a 69-year-old patient suffering from cystic lymphangioma of the head of the pancreas treated at the Rabat military hospital in the visceral surgery department II.

The rarity of this pathology due to its frequency and especially its location motivates the present thesis, the objective of which is to study the diagnostic and therapeutic particularities of cystic lymphangioma of the pancreas.

Case presentation :

This is a 69-year-old weaned alcoholic and tobacco patient with a history of urothelial carcinoma treated in 2019, pulmonary tuberculosis treated in 2020 and followed by gastroenterology for isolated melena admitted for treatment of melena complicated by a severe anemic syndrome in which the clinical examination finds a pale asthenic patient stable on the hemodynamic and respiratory level.

The biological analyzes showed hypochromic microcytic anemia (hemoglobin at 9 g/dl), all other analyzes as well as the dosage of tumor markers (CA-125, CA19-9 and ACE) were unremarkable.

Abdominal CT shows a solid, multilocular, retroduodenopancreatic cystic mass measuring 99x66 mm extended over 107mm associated with infiltration of peri-lesional fat and ADP of the lesser omentum (Figure 1).

This mass circumscribes the head and isthmus of the pancreas and maintains a thin line of separation with the superior mesenteric artery.

No soft tissue components and no calcifications were found.

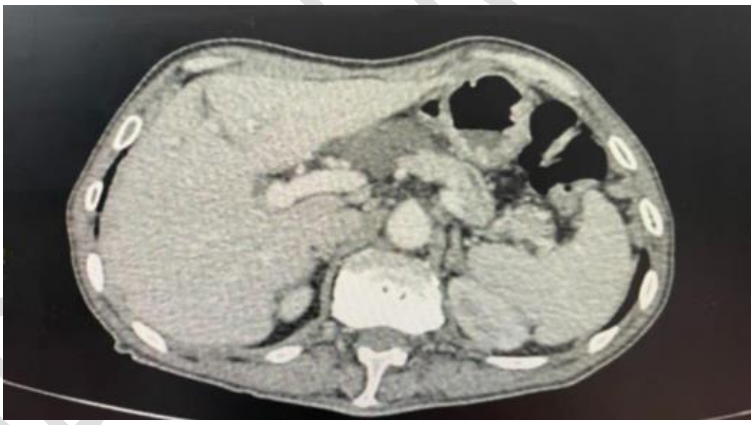

Figure 1: axial section of an abdominal CT scan showing a retroduodenopancreatic mass after injection of contrast product (visceral surgery department II, HMIMV)

Complementary MRI reveals the presence of a finely partitioned duodeno-pancreatic cystic mass (hyposignal on T1 and hypersignal on T2), the partitions of which are discreetly enhanced after injection of contrast product measuring 97x90mm and extending over 107mm (Figure 2).

This formation molds the adjacent structures in particular: the inferior vena cava, the liver and the lesser omentum and is associated with an infiltration of the posterior duodenal wall and the posterior surface of the pancreas with absence of dilation of the intra and extrahepatic bile ducts.

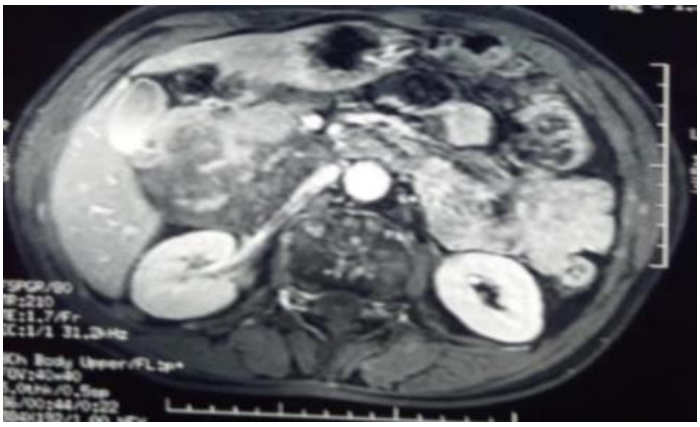

Figure 2: Axial T1 abdominal MRI showing the heterogeneous mass associated with infiltration of the posterior duodenal wall and the posterior surface of the pancreas. (Visceral surgery II, HMIMV)

Based on the CT and MRI findings a cystic lymphangioma of the pancreas was suspected.

Consequently, due to the impossibility of establishing a definitive diagnosis and not being able to exclude with certainty a malignant origin: we decided to submit the patient to laparotomy surgery through a left subcostal approach extended towards the right side .

Exploration during laparotomy confirmed the presence of a retroperitoneal polycystic mass infiltrating the entire posterior surface of the pancreas, the posterior duodenal wall as well as the root of the mesentery (Figure 3).

Complete resection was impossible without mutilating surgery with total pancreaticoduodenectomy. Considering the burden of radical surgery (R0), its high morbidity and mortality, we performed an incomplete tumor resection (R2) with preservation of the pancreas (infiltrated over its entire posterior surface).

The area of duodenal invasion causing the bleeding was resected with macroscopically healthy margins.

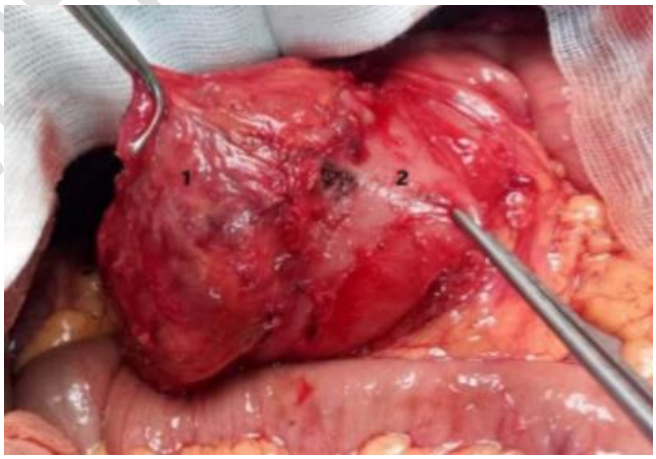

Figure 3: Intraoperative view showing a large pancreatic mass with duodenal infiltration.

1- The tumor, 2- second portion of the duodenum

Histological examination of the surgical specimen showed a lesion measuring 9x6x2.5 cm with a heterogeneous multi-cystic appearance with benign cystic tumor proliferation consisting of dilated vascular structures lined with flattened and regular endothelial cells of variable sizes with eosinophilic and acellular content including walls contain an inflammatory infiltrate organizing into a lymphoid follicle with absence of signs of malignancy thus concluding the definitive diagnosis of cystic lymphangioma of the pancreas (Figure 4).

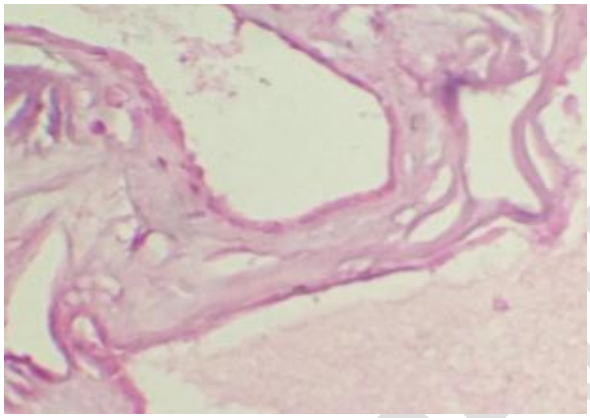

Figure 4: Benign cystic tumor proliferation consisting of dilated vascular structures lined with flattened endothelial cells of variable sizes with eosinophilic and acellular anhist content (HE, Gx400)

The postoperative course was marked by a pancreatic fistula which dried up spontaneously after 17 days.

After 3 weeks of hospitalization and given the favorable evolution, the patient left the hospital.

Six-month follow-up with abdominal CT showed tumor recurrence at the head of the pancreas measuring 58x40x78 mm.

Today, two years after the operation, the patient is clinically asymptomatic, two control abdominal CT scans were carried out and all came back in favor of lesion stability.

Annual monitoring is recommended.

DESCUSSION

Cystic lymphangiomas are rare, slow-growing benign tumors most often found in children under 2 years of age [6].

These are conditions that were first described by Redenbacher in 1828 [2].

The most frequently affected areas are the cervico-facial region with a frequency estimated at 75%, then the axillary region with a frequency estimated at 15% [6].

Its presentation in the pancreas is very rare, representing less than 1% of all lymphangiomas and only 0.5% of all pancreatic lesions.

It affects all age groups but is frequently found in women (ratio 2:1) explained by the use of oral contraceptives, hyperprogesteronemia and pregnancy which have positive effects promoting the growth of lymphangiomas [5, 7].

The precise etiology of cystic lymphangiomas of the pancreas is not yet well defined.

In the majority of cases described in the literature, most patients with LKP were asymptomatic, their lesions were discovered incidentally during radiological examinations or during treatments for other unrelated diseases.

However, depending on their size, they can become symptomatic, manifesting as non-specific gastrointestinal symptoms such as abdominal pain sometimes associated with nausea and vomiting [9].

Although rarely, they may manifest as acute abdomen due to complications such as cyst rupture, volvulus, infection, or intracystic hemorrhage [8].

In general, biological parameters are not specific and are of little interest in diagnosis.

This lack of biological specificity is one of the particularities of this pathology [10].

In our case, the assessments carried out showed no disturbance of the biological parameters.

CT and MRI help determine the location of the tumor, its size, its relationships with neighboring organs and also detect complications.

Ultrasound endoscopy is an optimal way to reveal the internal details of pancreatic lesions. LKPs have a variable appearance, they can be uniocular or multilocular with the possibility of finding macro or micro cysts in the same lesion [1].

Therapeutically, the treatment of choice for LKP is almost exclusively surgical in the absence of contraindications.

Complete excision of the tumor is curative for cystic lymphangiomas of the pancreas. However, when it is incomplete, there is a risk of recurrence [5].

Depending on the size and location of the tumor, complete resection of the tumor can range from a simple excision of the mass to a pancreatectomy associated with ablation of adjacent organs when the latter are invaded.

Until now, no consensus has established a therapeutic reference.

However, in this chapter we will report the possible surgical alternatives, namely:

Cephalic duodenopancreatectomy, central pancreatectomy, caudal pancreatectomy

and tumor enucleation.

Non-surgical alternative:

Aspiration of cystic contents: It consists of pricking into the cyst and emptying it by sucking up the liquid with a syringe.

It is subject to certain conditions (compliance with aseptic conditions and under endoscopic ultrasound).

This technique is used for symptomatic, easily accessible and unresectable lesions [10].

Percutaneous sclerotherapy: It constitutes a therapeutic alternative [11,12], it is a percutaneous injection of a sclerosing product which will cause a giganto-cellular type inflammatory reaction, which

subsequently transforms into sclerosis or fibrosis responsible for a stabilization of the evolution then the virtual disappearance of the tumor [13].

Given that LKP are benign tumors, their prognosis is generally favorable [1,4].

CONCLUSION

Cystic lymphangiomas of the pancreas are rare benign lesions that arise from the lymphatic vessels, accounting for less than 0.5% of all pancreatic cysts.

Generally it is asymptomatic, but can present atypically.

Its diagnosis used to be late and difficult preoperatively; the discovery occurred incidentally during imaging examinations for non-pancreatic diseases.

Currently the development of endoscopic ultrasound coupled with suction puncture makes it possible to identify cysts.

The combination of ultrasound morphological characteristics, macroscopic aspects of the aspirated fluid, biochemical and cytological evaluation of the sample make preoperative diagnosis possible.

However, the definitive and certain diagnosis is made by surgery after resection of the tumor tissue and its histological study.

To date, surgery remains the only curative therapy in the treatment of cystic lymphangiomas of the pancreas.

Complete excision eliminates the risk of recurrence but this alternative is not always possible.

Consent

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

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1.

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