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Splenic Cystic Lymphangioma : Focus On Two Clinical Cases

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Summary:
We report in this article a rare entity in terms of its occurrence and location: cystic lymphangioma of the spleen, through two clinical cases in the visceral surgery department of the MOHAMED V military hospital in Rabat, operated on within ten years.

Key words: *Cystic lymphangioma, Spleen, Splenectomy, Diagnosis.*

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I. INTRODUCTION:

Cystic lymphangioma is a benign tumor that develops at the expense of the lymphatic vessels. It occurs in both adults and children.

The clinical presentation is variable, the diagnosis is made by postoperative histology, the surgical treatment is the reference, and its prognosis is often good.

We will study and discuss through two clinical cases of splenic cystic lymphangioma, epidemiological data, clinical aspects and diagnosis in the literature.

II. Case presentation:

Case N°1

Mr A.H, a Moroccan 46-year-old admitted, operated at the age of 41 for peritonitis by ulcer perforation. Three months earlier, he presented epigastric and left hypochondrium pain associated **with a weight loss of 10kg**. The physical examination is normal except an inconspicuous splenomegaly. Hydatid serology and oeso-gastro-duodenal fibroscopy were normal. Abdominal ultrasound showed a splenomegaly containing a heterogeneous liquid mass without deep lymphadenopathy. Abdominal CT scan with contrast showed a 50 mm splenic lesion which has a cystic appearance, raised in the periphery after injection of the contrast, associated with abnormal thickening of the gastric wall (fig.1). A malignant pathology has been suspected, such as lymphoma or splenic metastasis. ACE and CA 19-9 were negative. A laparotomy was performed. Abdominal exploration found a homogeneous splenomegaly with no other abnormalities: splenectomy was realized (Fig.2) and the diagnosis of splenic cystic lymphangioma was made on histological exam of the surgical specimen. The evolution was good for 6 years on half-yearly and then annual check-ups.

Case No. 2:

Miss N.B, Moroccan 22 years old, without antecedents, admitted for non-specific abdominal pain, without digestive signs and without any repercussion on the general state. The physical examination on admission was strictly normal. An abdominal ultrasound revealed a very limited multi-partitioned cystic lesion of the spleen measuring about 60 mm. The complementary abdominal CT scan confirmed the presence of the splenic lesion with a second retroperitoneal cystic lesion of 40 mm opposite the lower pole of the left kidney and a third cystic lesion of 20 mm on the left ovary. Hydatid serology, CA19-9, ACE and CA125 were normal. The patient was operated on laparoscopically (fig.3): a splenectomy was performed. The retroperitoneal lesion was resected by breaking the capsule, measuring about 50 mm in diameter with a chylous content and a very thin and fragile wall. The ovarian cyst was simple with a thin, homogeneous wall with no partitions and was about 20 mm long; it was completely removed. The postoperative follow-up was simple and the patient was discharged from the hospital on D3 of the operation. The results of hystological exam was:

- Spleen: splenectomy specimen measuring 12x6x2cm, with a cystic-looking section with haemorrhagic reshaping, and a whitish lesion measuring 1cm located 1mm from the capsule.
- Left pararenal cyst: presence of several fragments of 0.4 to 1.8 cm.

- Pelvic cyst: a fragment of 3x2x2 cm, with a cystic-looking section, presence of a very fine capsule.

The morphological appearance is identical at the three sites corresponding to cystic lymphangiomas, with no histological sign of malignancy.

III. Discussion:

Lymphangioma was first described by REDENBACHER in 1828 but its relationship with the lymphatic system was re-established half a century later by KOESTER. Splenic localization of KL is very uncommon. The first case of splenic non-parasitic cystic lymphangioma was described by ANDRAL in 1829, and PEAN initiated splenectomy for this condition in 1867(1).

LK is a tumour that can affect all organs except the central nervous system, which is devoid of lymphatics. They are located in the subcutaneous tissues of the face and neck (60%), extremities (20%), trunk (10%) and armpits. Deep abdominal (2-10%), mediastinal or thoracic (5%) locations are less common (3). Splenic localization is rare. Approximately 90 cases have been reported in the literature. The discovery of spleen localisation in adults is exceptional and often affects the female sex (4). Our work clearly demonstrates the rarity of this localization, as we found only two cases over a period from 2006 to 2016.

Three theories have been put forward to explain the physiopathogeny of EKL: the mechanical theory is beginning to be abandoned, based on the obstruction of the lymphatic canalicules by an exogenous obstacle or an inflammatory factor and upstream cystic formation. The more modern and more adopted congenital theory, defended by the frequency of LK before the 2nd year of life and explained by faulty connections between the lymphatic ducts and the lymphatic sacs in the embryonic age.the 3rd theory not yet proven stipulates a runaway angiogenesis(5)

Splenic KL is often asymptomatic, discovered incidentally by imaging, however it can give symptoms such as: pain or adherent renal mass in the deep left hypochondrium, splenomegaly, and signs of compression (vomiting, hiccups, transit disorder, pleurisy, and proteinuria). The occurrence of splenic infarction or splenic vein thrombosis with segmental portal hypertension has been described (6).

The typical ultrasound appearance of the tumour is that of a hypoechoic and heterogeneous lesion, sometimes cystic and partitioned. Doppler shows the absence of flow within the malformation.

CT scan remains the best radiological examination for the exploration of cystic lymphangiomas of the spleen. It is performed without and with contrast material. The same ultrasound images are observed; it presents as a homogeneous hypodense liquid lesion crossed by fine partitions with no enhancement after injection of contrast product.

MRI remains a second-line examination to assess good mapping of the lesion; the LK of the spleen appears as a hyposignal T1 and a hypersignal T2.

The combination of ultrasound and CT scan or even MRI allows the diagnosis to be supported, but it remains difficult in the case of bulky cystic lymphangioma or in the event of complications. (7)

Definitive proof of the diagnosis of cystic lymphangioma is provided by pathological examination of a biopsy or surgical specimen. The macroscopic appearance is whitish or translucent, unilocular or polycystic with or without communication channels with serous or chylous contents. Microscopically: it is a cystic formation bordered by an endothelial lining with connective tissue partitions whose thickness is related to the age of the cyst (8).

Splenectomy is the classical treatment of spleen CF by laparotomy or laparoscopy with preparation of the patient (vaccine and antibiotic prophylaxis). Partial splenectomy or cystectomy is possible (9).

The prognosis is good after surgical treatment, recurrence is the main complication if the excision is partial, estimated at 10 to 15 % (10).

IV. Conclusion:

Cystic lymphangioma of the spleen is a rare benign tumor, usually asymptomatic and of incidental discovery. Its diagnosis can be directed by ultrasound and CT scan, confirmed by pathological examination of the surgical specimen. If left untreated, complications may occur including intracystic haemorrhage, infection and rupture of the cyst, while malignant transformation is exceptional.

The treatment of choice is surgical, consisting of complete removal of the lesion. The prognosis is excellent, however recurrences may occur in the event of incomplete resection of the cyst. This requires regular monitoring of these patients.

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