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Case Report

Splenic cystic lymphangioma in adult: a case observed in the Visceral Surgery Department II of Mohamed V Military Hospital

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ABSTRACT

Cyctic lymphangioma is a primary benign tumor due to congenital malformations of the lymphatic vessels. It is more often seen in pediatric patients and the usual site for lymphangioma is the cervical and axillary region. However, it is exceptionally seen in adults and involving rarely the spleen. The discovery of the splenic lymphangioma is generally accidental during imaging performed for another pathology. Indeed, being most often asymptomatic, the clinical presentation of cystic lymphangioma is polymorphic and it is related generally to mass effects. In this paper, we report the case of a 43 years old patient in whom the discovery was fortuitous during imaging for a non-specific symptomatology. The physical examination was normal and magnetic resonance imaging strongly evoked the diagnosis of cystic lymphangioma. A total splenectomy was performed and the histological examination has confirmed the positive diagnosis of splenic lymphangioma. Finally, this case confirmes that the diagnostic of splenic lymphangioma should be evoked in front of splenic cystic lesions and the confirmation and the differential diagnosis are made by histopathology and immunohistochemistry studies.

Keywords: Cystic lymphangioma, Splenic, Adult, Splenectomy

INTRODUCTION

Cystic lymphangioma is a primary benign tumor whose pathogenesis is mainly due to congenital malformations of the lymphatic channels. It is frequently encountered in children and exceptionally in adult. The splenic localization is rare.[1] Through this work, a case of splenic lymphangioma is reported, observed and treated in the Visceral Surgery Department II of Mohammed V Military Instruction Hospital in Rabat.

CASE REPORT

The patient referred to as Mrs. B.S., a 43 years old woman, who has as a history of three cesarean deliveries. She had chronic pain in the left upper quadrant associated with nausea which dates back to more than a year. The patient consulted and benefited from an abdominal ultrasound which objectified hypo echogenic splenic cystic lesions. The patient was referred to us in November 2019 for specialized care.

The clinical examination found a eupneic, apyretic patient with normo-colored conjunctivas. The abdominal examination showed a pfannenstiel scar, a supple abdomen with no palpable mass in the left hypochondrium. The rest of the clinical examination was unremarkable. Biological tests were normal and the hydatid serology was negative.

Abdominal magnetic resonance imaging (MRI) in axial section on T1-weighted sequence demonstrated a multiple multilocular formations in hypo signal, not enhanced after

gadolinium injection (Figure 1). In coronal section on T2-weighted sequence, these formations were in hyper signal and crossed by thin septations which were in hypo signal (Figure 2).

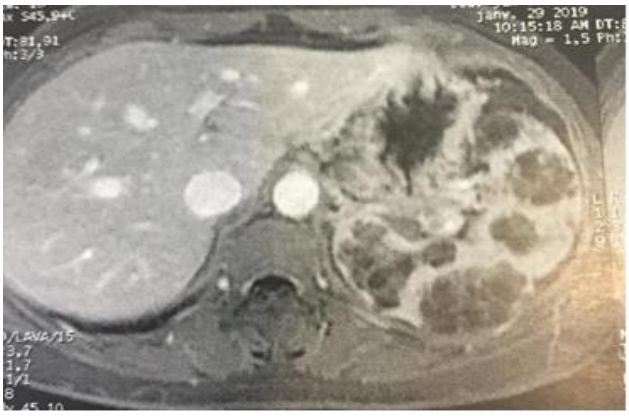

Figure 1: T1-weighted sequence in axial section.

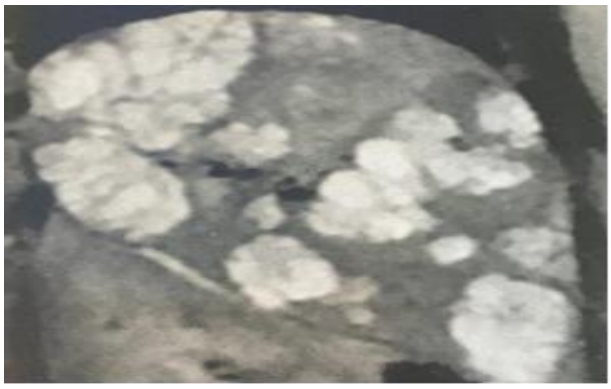

Figure 2: T2-weighted sequence in coronal section.

After preoperative preparation and vaccination, the patient had benefited from a middle supra umbilical laparotomy. On exploration, we found a splenomegaly with a multi-lobulated appearance and the presence of cystic formations taking all the splenic parenchyma. The decision was to perform a total splenectomy (Figure 3). The spleen measured 17 cm×12 cm×3 cm.

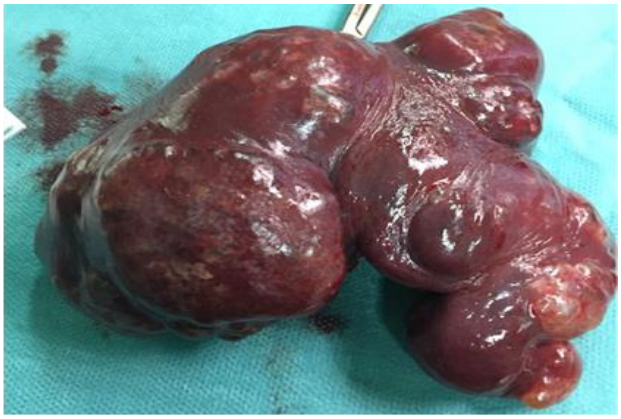

Figure 3: Operative piece of total splenectomy.

DISCUSSION

Cystic lymphangioma is a benign tumor caused by congenital malformations of the lymphatic system. Generally observed during childhood and rarely in adulthood with a clear predominance in young women.[2] It can affect all organs and is usually located in the cervical and axillary region. In the abdomen, the lesions preferentially reach the mesentery, the omentum, the retro peritoneum and rarely the spleen.[3] In our observation, it's about a young woman with a splenic localization of the lesions.

Being most often asymptomatic, the discovery of the splenic lymphangioma is generally accidental during imaging performed for another pathology.[4] However, the symptomatology, when present, remains non-specific. The clinical signs encountered are linked to the large volume of the spleen, which manifests as pain in the left hypochondrium or signs of compression of neighboring organs such as anorexia, nausea, vomiting and transit disorders.[5] Complications have been also described and are linked to portal hypertension. Clinical examination finds a palpable mass of the left upper quadrant.[6] In our case, the clinical signs were chronic pain in the left hypochondrium associated with nausea.

The positive diagnosis of splenic lymphangioma is evoked by medical imagery.[7] Abdominal ultrasound remains the most accessible, least expensive and non-invasive mean. It confirms the splenic site and the nature of the lesions that appears hypoechoic, partitioned and avascular on Doppler, whit calcifications and a heterogeneous appearance if intra cystic hemorrhage.[8,9,10] In our observation, the ultrasound evoked the diagnosis in the face of multi-partitioned, hypoechoic and avascular splenic lesions. Abdominal computerized tomography remains the radiological examination of choice, it shows hypodense, homogeneous, compartmentalized, and non-enhanced sub-capsular images after the injection of the contrast product. It provides information on the loco regional extension of the tumor and the presence of other locations, which define cystic lymphangiomatosis with a darker prognosis. Regarding magnetic resonance imaging, lesions appear in hypo signal on T1 and in hyper signal or iso-signal on T2 without enhancement after Gadolinium injection. For our patient, the MRI was the radiological examination carried out after the ultrasound that evoked the diagnosis of lymphangioma.

Biology has little interest in the diagnosis of splenic lymphangioma. However, it is necessary for the preoperative biological work up, searching for complications and the differential diagnosis, notably hydatid serology.

The anatomopathological examination is the only examination which confirms the diagnosis of splenic lymphangioma. Microscopically, it shows dilated lymphatic vessels, bordered by flattened endothelial cells

without signs of atypia, with the presence of abundant lymphoid tissue. Smooth muscle cells and foam cells containing lipid material can be seen at the wall of these vessels.

Immunohistochemistry currently contributes to the positive and differential diagnosis of splenic lymphangioma. Selective markers such as podoplanin (D2-40) can be used to distinguish lymphangiomas from hemangiomas because they selectively stain the lymphatic endothelium. Other techniques such as cluster of differentiation 31 and 34 (CD31, CD34) and factor VIII can also be used.[11]

Differential diagnosis of splenic lymphangioma includes solid and cystic lesions of the spleen such as hemangiomas, mesothelial and parasitic cysts, lymphoma and metastasis.[12]

Conservative treatment of splenic lymphangiomas with interferon alpha has been carried out in a child by Reinhardt et al. with success and with good tolerance. The duration of treatment and its dosage remain the only obstacles of this therapy.[13] The treatment of choice is a laparotomy total splenectomy. Laparoscopy, for its part, can be used in the absence of contraindications, in particular a splenomegaly or lesions with inaccessible localization. Some authors prefer conservative treatment by partial splenectomy with the drawback of a recurrence rate around 10%.[3]

CONCLUSION

Cystic lymphangioma of the spleen is a rare disease in adults. The clinical presentation is very polymorphic and the diagnosis is evoked by medical imagery and confirmed by the histology of the operative specimen of splenectomy. Laparoscopic treatment remains very promising.

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