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case of the month

Malignant Lymphomas

Lymphangioma of the spleen in an elderly patient

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ABSTRACT

Splenic cystic lymphangioma is a very rare condition, and is classified among cystic proliferations of the spleen. It is considered to be the result of a developmental malformation of the lymphatic system and can involve the spleen alone or be a part of multiorgan disease. It is usually seen in children, often found incidentally. We describe a case of cystic lymphangioma of the spleen in an elderly woman putting emphasis on the rarity of the case in old age, and on the problems of differential diagnosis with the other cystic proliferations of the spleen, in particular hydatid disease, in the absence of histologic information.

Key words: lymphangioma, spleen

Parasitic cysts are the most common cystic proliferations of the spleen. Non-parasitic cysts, classified as primary or true cysts, are very rare and include epidermoid and dermoid cysts, cystic hemangiomas and cystic lymphangiomas, and pseudocysts.[1]

Splenic cystic lymphangioma is thought to be extremely rare, with very few cases having been reported. It is usually found in children in whom it is often an incidental imaging finding. We report the case of an elderly woman with a cystic lymphangioma of the spleen who was admitted as an emergency with acute abdominal pain. We present this case with emphasis on the problem of differential diagnosis of cystic and neoplastic lesions of the spleen, due to limitation of imaging techniques, and hence on the difficulties of diagnostic certainty in the absence of histologic features.[2]

Case report

An 84-year old woman was admitted for evaluation of upper left quadrant pain with nausea, vomiting and fever. She reported the presence of pruritus and significant weight loss (30 kg) in the preceding months and mild diffuse continuous abdominal discomfort in the week before admission. In addition the patient's spleen was enlarged to the transverse umbilical line and she had diminished bowel sounds. She was submitted to an ultrasound of the abdomen. The spleen had a bipolar diameter of 16 cm and at the bottom there was a round hyperechoic and solid lesion of 6 cm in size, containing a fluid; within the spleen, beneath this lesion there were a lot of cystic lesions, the largest of which was 33 mm long. These findings were compared with those of eighteen months previously when the spleen had already been enlarged with disruption of morphologic and ultrasound structure due to a large round hyperechoic lesion. This was a 7 cm lesion of the lower pole with a round anechoic lesion of 19 mm in size in the middle. Under this there was another round anechoic lesion. In addition, the patient had undergone a CT examination of the brain that had revealed five calcified homogeneous lesions in the supratentorium, considered as being the result of a parasitic process.

A fine needle aspiration of the splenic lesion was attempted, but was not diagnostic.

The patient's biochemical and hematologic tests are reported in Table 1. Pruritus associated with splenomegaly, mild anemia and a high level of β_2 microglobulin led us to suspect a lymphoproliferative disease. The high plasma β_2 microglobulin levels (6,402 mg/L: normal range < 3 mg/L) could not be related to chronic mild renal failure; in addition the levels decreased after splenectomy, being 3,238 mg/L after six months. The patient, therefore, underwent a bone marrow biopsy. This showed anomalies of morphology of all phases of maturation of erythropoiesis with diserythropoiesis. High levels of granulopoietic and megakaryocytic cells were present with infiltration of plasma cells and small lymphocytic elements (15%). These findings added further weight to the suspicion of a lymphoproliferative disorder. In addition, there was no evidence of micro-organisms such as bacteria and protozoa in the marrow, and serologic screening for hydatid disease and amebiasis, leishmaniasis and cysticercosis resulted negative.

An abdominal CT examination confirmed the spleen lesions shown by sonography, but they appeared larger and more numerous, without hemorrhagic areas in the middle. This kind of description suggested a parasitic process, in contrast with the first suspicion (Figure 1). In addition, CT scans of the brain detected intraparenchymal calcifications.

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Table 1. The patient's biochemical and hematologic parameters.

Test	*Before surgery*	*After six months*	*Normal range*
Hb, g/L	110	134	120–155
Hct, L/L	0.34	0.42	0.35–0.45
RBC $\times 10^{12}$/L	3.96	4.3	4.7–6.1
MCV fL	86.1	85.3	80–100
WBC, $\times 10^9$/L	7.96	6.18	3.4–10
Plt, $\times 10^9$/L	254	285	150–450
Iron, µg/dL	27	35	50–175
Ferritin, µg/L	29.52	31.3	4-161
ESR, mm/h	20	16	< 15
CRP, mg/dL	0.8	0.3	< 0.5
β_2-M, mg/L	6.402	3.238	< 2
Creatinine, µmoli/L	114.1	103.3	50 – 100
BUN, µmoli/L	21.6	12.3	2.9-7.1
Total bilirubin µmoli/L	15.9	10.1	2–21
Direct bilirubin, µmoli/L	4.61	2.45	< 7
ALT, µkat/L	0.18	0.15	0–0.58
AST, µkat/L	0.26	0.19	0–0.58
γGT, µkat/L	0.22	0.16	0.15–1.42
Alkaline phosphatase, µkat/L	4.32	3.1	0.7–2.2
LDH, µkat/L	6.12	3.1	1.46–3.82

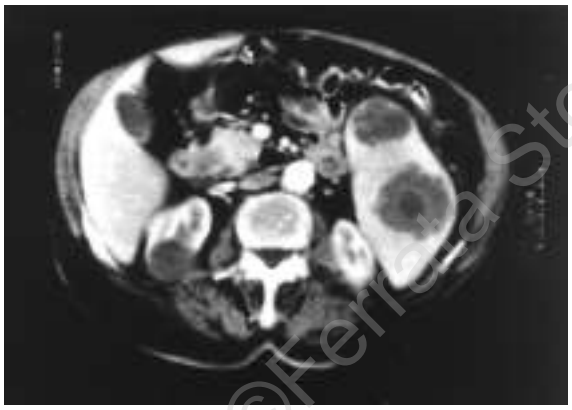

Figure 1. CT after i.v. contrast medium administration showing an enlarged spleen with multiple parenchymal focal lesions. There were hypodense, heterogeneous, with irregular margins. No calcifications or hemorrhagic foci were detected.

These were found in the temporal and occipital regions on the right and in the anterior parietal region on the left, and were considered to be of inflammatory parasitic cause.

To elucidate whether the splenomegaly was related to parasitic echinococcal infiltration or non Hodgkin's lymphoma, the patient underwent a surgical splenectomy. The spleen was 17×9.5×5.5 cm and weighed 580 g; sectioning it revealed multiple nodules (Figure 2a). Histologic examination showed that the nodular lesions consisted of vascular spaces

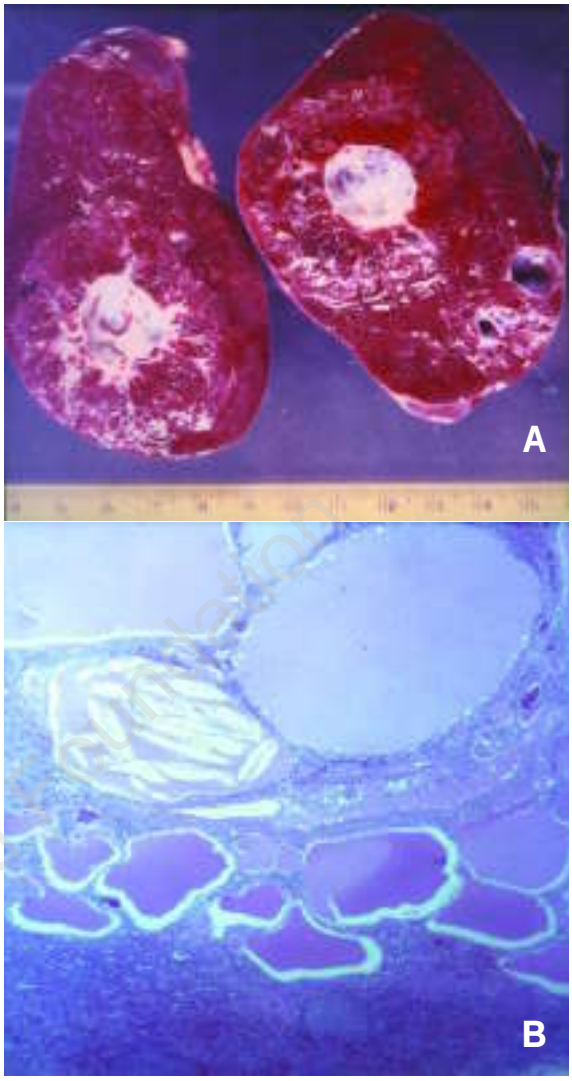

Figure 2. A. The spleen (weight: 580 g) showed multiple nodular lesions (arrows). (H&E). B. The nodular lesions consisted of numerous vascular spaces filled with proteinaceous fluid. (H&E 10×).

associated with fibrous trabeculae; the spaces were lined by flat endothelial cells and were filled with proteinaceous fluid (Figure 2b).

Discussion

Cystic lesions of the spleen include parasitic and non-parasitic cysts. Parasitic cysts, almost exclusively caused by echinococcal disease, represent 50 to 80% of splenic cysts, although splenic echinococcosis represents only 3.5% of echinococcal diseases.[1] In addition, encephalic localization of echinococcosis has been described.[3] Non-parasitic cysts are classified as primary or true cysts, which have an epithelial or endothelial lining, and pseudocysts which may be classified as post-traumatic, degenerative or inflammatory. Endothelial true cysts may be lymphan-

giomas or hemangiomas.[1]

Cystic lymphangioma is a very rare condition. It is usually seen in children in whom it is frequently discovered incidentally.[4] These tumors occur more frequently in females and 80 to 90% are detected before the end of the second year of life.[5]

The neck (75%) and axillary regions (20%) are the most common locations of lymphangioma, but it can occur in the retroperitoneum, mediastinum, mesentery, omentum, colon, pelvis, groin, bone, skin, scrotum and spleen.[6] Lymphangioma of the spleen can involve the spleen alone, or it can be a part of multivisceral involvement; when diffuse it is termed systemic cystic angiomatosis.[7]

Patients with splenic lymphangioma may be asymptomatic or symptomatic at diagnosis. Upper left quadrant pain is the most common symptom, frequently followed by fever, nausea, vomiting, and weight loss, i.e. the symptoms reported by our patient, hypertension, hypersplenism and consumptive coagulation disorders. Because of the similarity of symptoms and signs, it is often confused with hydatid disease which a negative *E. granulosus* agglutination test result cannot always exclude. The right diagnosis depends on histopathologic examination after removal of the spleen.[8,9]

Ultrasonography and computed tomography are the most helpful imaging techniques in the diagnosis of splenic cystic lesions.[1,10,11] Usually, in ultrasound images, a lymphangioma appears as cystic, multiseptate masses with lobules that may be anechoic or contain internal echoes or sedimentation with fluid-fluid levels caused by debris. Nowadays computed tomography helps pre-operative diagnosis. On CT scans, lymphangiomas appear as cystic masses, with Hounsfield units ranging from the attenuation of water (if the contents are serous) to that of fat (if the contents are chylous). Notwithstanding this, because of the similarity of ultrasound features of cystic proliferation of lymphangiomatous origin of splenic endothelium, the diagnosis of splenic cystic lymphangioma must be made in the light of the histologic findings of biopsy specimens. In fact, the contents of the cyst may, rarely, be hemorrhagic and show high attenuation on CT scans.[12,13]

A fine needle aspiration biopsy can be useful in differentiating between benign and malignant lesions.[14,15] In our case it was not diagnostic so surgical excision was planned.

Lymphangioma is a benign tumor and is considered to be a developmental malformation in which obstruction or agenesis of lymphatic tissue results in lymphangiectasia secondary to lack of normal communication of the lymphatic system.[6] Splenic lymphangioma may involve the spleen in the form of solitary nodules, multiple nodules or diffuse growth. Its classification has been based primarily on histologic findings of three types: simple, cavernous and cystic.[16]

Histologic examination of the nodules promptly excluded the clinical suspicion of parasitic cysts and established the vascular origin of the lesions. The differential diagnoses included both malignant (i.e. angiosarcoma) and benign (i.e. lymphangioma, splenic hemangioma, and peliosis of spleen) splenic vascular proliferations. Angiosarcoma was promptly excluded because of the absence of significant cytologic atypia of the endothelial cells; among the benign lesions, the presence of a typically flat endothelium and an eosinophilic proteinaceous material filling the endothelium-lined spaces favored the diagnosis of splenic lymphangioma instead of hemangioma, in which the endothelium-lined spaces are filled by blood. Cyto-architectural features also excluded peliosis of the spleen since blood spaces in this condition are haphazardly scattered in the red pulp instead of forming a discrete tumor.[17]

Although benign, abdominal lymphangioma may become locally invasive, so treatment remains surgical and splenectomy seems to be the best choice.[9,10,18]

The prognosis is good.[19]

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MV: writing of the report; MV with EO, MM and AM care of the patient; ALF: radiologic examinations; MP, RR: histologic diagnosis.

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