Splenic cystic lymphangioma in a young woman : case report and literature review

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Abstract

Splenic cystic lymphangioma is extremely rare, with very few cases reported until now. Here, we report a case of cystic lymphangioma of the spleen in a young woman who was admitted for evaluation of abdominal pain and a mass lasting for two years. We present this case with emphasis on the problem of differential diagnosis and the difficulties of diagnostic certainty in the absence of histologic features. (Acta gastroenterol. belg., **2011**, 74, **334-336**).

Introduction

Splenic cystic lymphangioma is thought to be extremely rare, with very few cases reported until now. It is usually accidentally found in children through imaging examination, and only few cases have been reported in adult patients (1-10). Splenic lymphangiomas have a wide spectrum of clinical and laboratory findings from an incidental finding to a large, symptomatic mass (11). The prognosis is good, however, there is a remarkable high risk of spleen rupture and surgery is always the preferable definitive treatment (12). The clinical data concerning adult splenic lymphangiomas are insufficient. Here, we report a case of cystic lymphangioma of the spleen in a young woman who was admitted for evaluation of abdominal pain and a mass lasting for two years. We present this case with emphasis on the problem of differential diagnosis and the difficulties of diagnostic certainty in the absence of histologic features.

Case report

A 28-year-old woman was admitted for evaluation of upper left quadrant pain and a mass lasting for two years. She had no history of alcohol consumption or smoking. The patient's body temperature, blood pressure, pulse rate and respiratory were unremarkable. A physical examination demonstrated tenderness and enlargement of the spleen which was felt 5 cm below the left costal margin. Abdominal ultrasonography showed a significantly enlarged and deformed spleen with a lot of cystic lesions of 2 mm to 30 mm in size. These cystic lesions contained fluid and were anechoic and well separated, with fine light spots distributed in the anechoic area. The cystic wall was very thin, with splenic tissue located between these cystic lesions. Blood vessels in the hilum of the spleen were significantly thickened. Nothing abnormal was detected in the liver, kidneys and ovaries. A clinical diagnosis was made as multicystic megalosplenia. Surgical splenectomy was performed, and during surgery, no other organs were found to be involved. The excised spleen weighed 1100 g and was 19 × 16 × 10 cm. The surface was firm, uneven and nodular, and multiple yellowish or white nodes could be seen protruding onto the surface (Fig. 1A). Cross section of the spleen revealed a cellular texture with many cysts containing aqueous liquid (Fig. 1B). Microscopy demonstrated hyperplastic lymphatic vessels within the spleen filled with proteinaceous fluid (Fig. 2A and B). Pathologically, it was diagnosed as cystic lymphangioma.

Discussion

Lymphangioma is a rare disease generally regarded as a congenital malformation of the lymphatic system, in which obstruction or agenesis of lymphatic tissue results in lymphangiectasia due to lack of normal communication of the lymphatic system (1-10). Lymphangiomas occur most commonly in the neck (75%) and axillary region (20%) (13). They sometimes, though rarely, involve the mediastinum, retroperitoneum, mesentery, omentum, and parenchymal organs such as the lung, gastrointestinal tract, spleen, liver, pancreas, adrenal gland, gallbladder, and bone (1-4,6-23). When lymphangioma affects the bone, soft tissue, or parenchymal organs in a diffuse or multifocal manner, it is called lymphangiomatosis. Despite its benign origin, the prognosis of diffuse lymphangiomatosis depends on the organs involved and the extent of involvement. Extensive liver, lung, or bone involvement can be fatal.

Lymphangiomas have been classified as simple, cavernous and cystic depending on the size of the dilated lymphatics (24). Splenic lymphangiomas, which are usually of the cystic type, are rare tumors, usually seen in children and with a female predominance (6,25).

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Submission date : 03/06/2009
Acceptance date : 01/09/2010

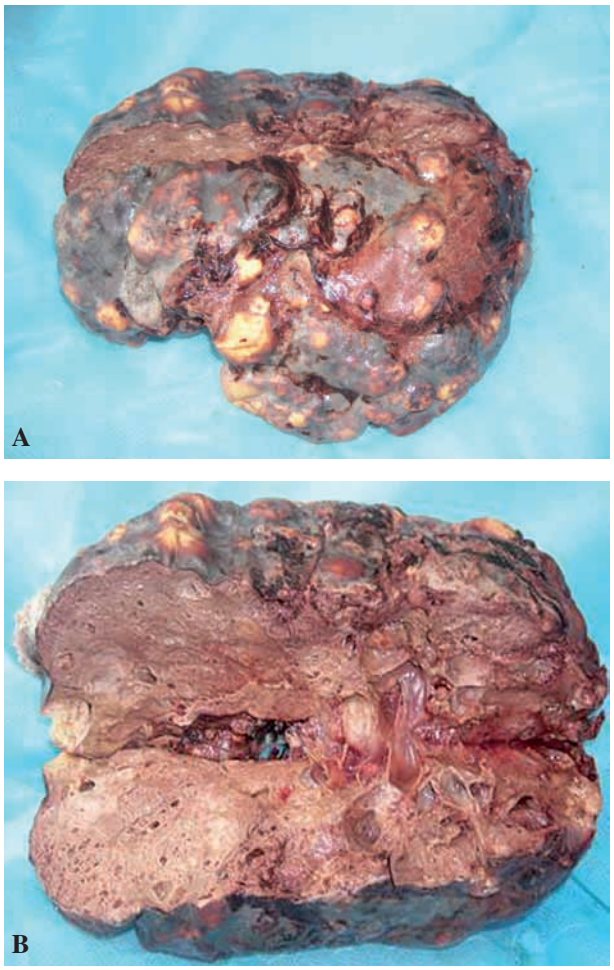

Fig. 1. — A&B. Gross appearance of the excised spleen

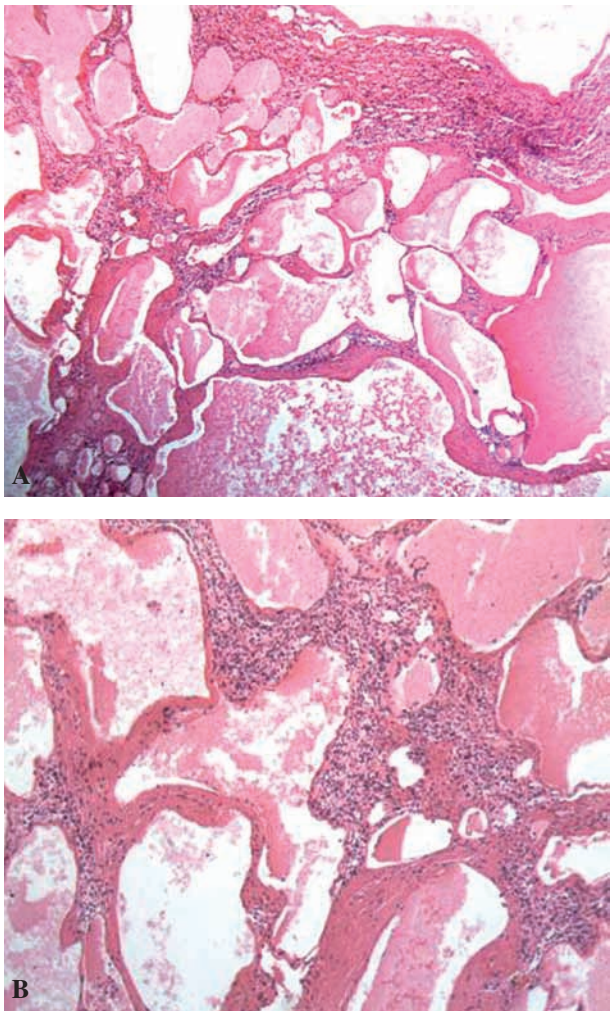

Fig. 2. — A&B. Microscopic examination of pymphangioma spleen. A. HE staining 40× and B HE staining 400×.

Lymphangiomas of the spleen may present as a single cyst, as multiple cysts of different sizes separated by internal septations or as diffuse growth (6,26). They are thought to be benign, although one case with malignant transformation has been reported (27). Lymphangioma of the spleen can involve the spleen alone or multiple organs. At gross examination, lymphangiomas may appear as unilocular or multilocular cystic masses (Fig. 1A&B) ; they may be solitary or may involve the entire spleen (lymphangiomatosis). The contents are serous to chylous. At microscopic examination, lymphangiomas are seen to be composed of multiple vascular channels that are lined with a single layer of endothelium and filled with proteinaceous fluid (lymph) (Fig. 2A&B) (17,28). Small areas of hemorrhage may be present within the lymphangiomas. Patients with simple splenic lymphangioma may be asymptomatic or symptomatic at diagnosis. Upper left quadrant pain is the most common symptom, frequently followed by fever, nausea, vomiting, weight loss, hypertension, hypersplenism and consumptive coagulation disorders (11).

Ultrasonography, computed tomography (CT) and magnetic resonance imaging (MRI) are the most helpful imaging techniques in the diagnosis of splenic cystic lesions (3,6,9,12,28). 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. CT features include splenomegaly, with single or multiple areas of low attenuation. Lymphangiomas are sharply marginated and do not enhanced after contrast administration (28). However, small, marginal and linear calcifications may be present on CT scans. The MRI appearance of the lesions is very similar to that of cysts, with homogeneously low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. Areas of high signal intensity may be shown on T1-weighted images if internal hemorrhage is present or if the lesions contain a large amount of protcinaceous fluid. The advantage of MRI is that it is multiplanar and multiparametric ; this is important for the identification of possible areas of malignant degeneration, through the

variation in the signal and especially with the enhancement after paramagnetc contrast injection (3). However, because of the similarity of imaging features of a cystic lesion in the abdomen, the differential diagnosis is extensive, including true splenic cysts, mesothelial cysts, old hematomas and echinococcosis, even in a serologically negative patient, and the extremely rare cystic hamartoma (26). In patients whose lesions were not biopsied preoperatively, lymphangioma was not detained as a likely diagnosis. In a series of lymphangioma, the diagnosis of lymphangioma after imaging studies was suspected in less than 25% of cases (29). The exact diagnosis of splenic cystic lymphangioma can only be made in the light of the histological findings on biopsy specimens.

The normal evolution of splenic lymphangioma is variable. Small lesions are often incidentally detected, and large lesions may increase in size and cause the compression of other organs or the rupture of the splenic capsule. For large splenic cysts, splenectomy is advantageous, as the risk of rupture is very high, even from minor abdominal injury. Symptomatic splenomegaly is also a valid indication for surgery (11). In cases of infected lymphangioma, the patient should be stabilized if possible with supportive treatment and antibiotics before resection.

In conclusion, splenic lymphangiomas are rare tumors that present a difficult diagnostic challenge, and the correct diagnosis often remains elusive until tissue is obtained. In young patients, however, it should be outlined that the possibility of the rare but usually of poor prognosis multiorgan lymphangiomatosis must always be excluded.

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