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Cystic lymphangioma of the spleen: US-CT-MRI correlation

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Abstract A case of a surgically confirmed cystic lymphangioma of the spleen is presented. Preoperative imaging consisted of US, contrast-enhanced CT and MRI, all showing a multiloculated lesion with small cystic cavities divided by thin septa, corresponding to dilated lymphatic spaces. Preoperative studies correlated well with the pathologic findings. Cystic lymphangioma of the spleen is a very rare condition and is usually solitary and asymptomatic. Large lymphangiomas may be an indication for splenectomy, since the risk of rupture is high even from minor abdominal trauma. Preoperative diagnosis may be achieved with correlated noninvasive imaging.

Key words Spleen · Tumors · Cystic lymphangioma

Introduction

Lymphangiomas are benign congenital malformations of the lymphatic system that can be either solitary or multiple. They principally occur in children or young adults and rarely manifest after the age of 20 years. The most common locations of lymphangioma are the neck (approximately 75% of cases) and the axillary region (20% of cases) [1].

Cystic lymphangioma of the spleen is a very rare condition and is usually solitary and asymptomatic. The prognosis is good but there is a very high risk of rupture even after minor trauma, therefore surgery is indicated as definitive treatment. The percutaneous biopsy is delicate, due to the high risk of rupture. Noninvasive imaging is therefore fundamental for obtaining a preoperative diagnosis. We present here the correlated US, CT and MRI features of a cystic lymphangioma of the spleen.

Case report

A 55-year-old woman was admitted to our institution because of mild pain in the left upper abdominal quadrant, without fever. Red and white blood cell counts were normal and the platelet count was 201,000/ml; other laboratory tests were unremarkable. Abdominal US showed two small angiomas within the liver and an enlarged spleen. Within the splenic parenchyma there was a central mass measuring 5.5 cm in largest diameter. The mass consisted of a larger central cyst surrounded by numerous peripheral cysts of smaller size, divided by thin septa (Fig. 1).

On contrast-enhanced CT the appearance was similar to that on US; the cystic portion showed no contrast enhancement while the septa were slightly hypervascular. There was no evidence of internal soft tissue nodules or spotty calcifications (Fig. 2).

MRI of the upper abdomen was obtained with a 1.5 T unit, using T2*- and T1-weighted sequences. The MR examination confirmed that the mass consisted of a central fluid portion surrounded by multiple smaller cysts(Fig. 3). The walls of the cysts as well as the septa between the cysts were very thin. There was no evidence of extracapsular extension or invasion of surrounding organs. The preoperative diagnosis was that of a multicystic lesion of the spleen, probably a lymphangioma. The patient underwent elective splenectomy, which was carried out without complications. Gross pathologic examination revealed a cystic tumor composed of various-sized multiloculated cysts. Histologically, the tumor was com-

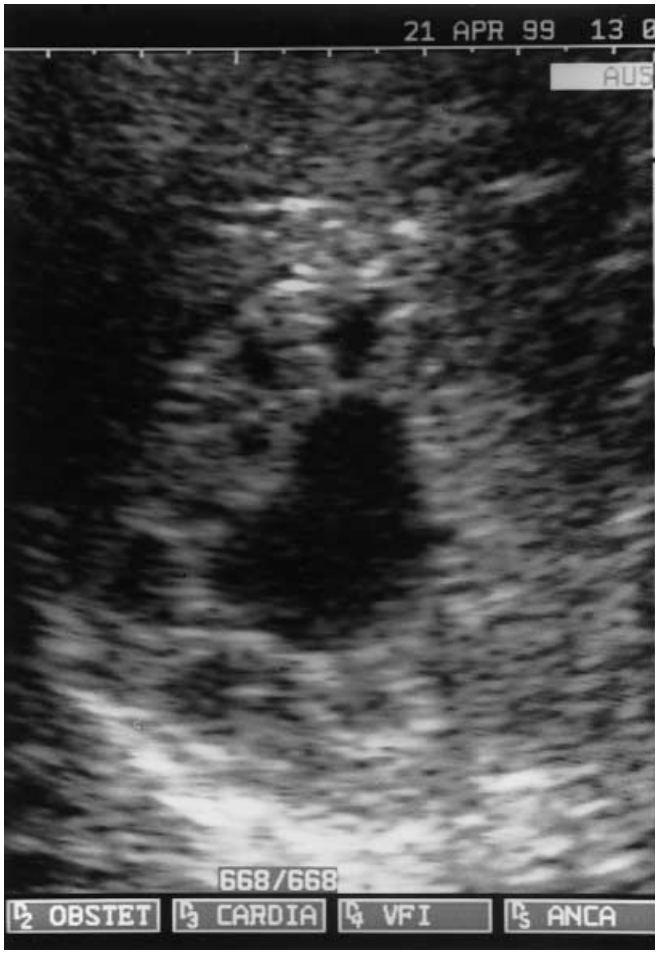

Fig. 1 Coronal US image of the spleen (enlarged view). There is a 5.5 cm mass with a large central cystic area, surrounded by numerous smaller cysts. Some of the smaller cysts are below the power of resolution of the US technique and give a homogeneously hyperechoic appearance to certain areas of the mass

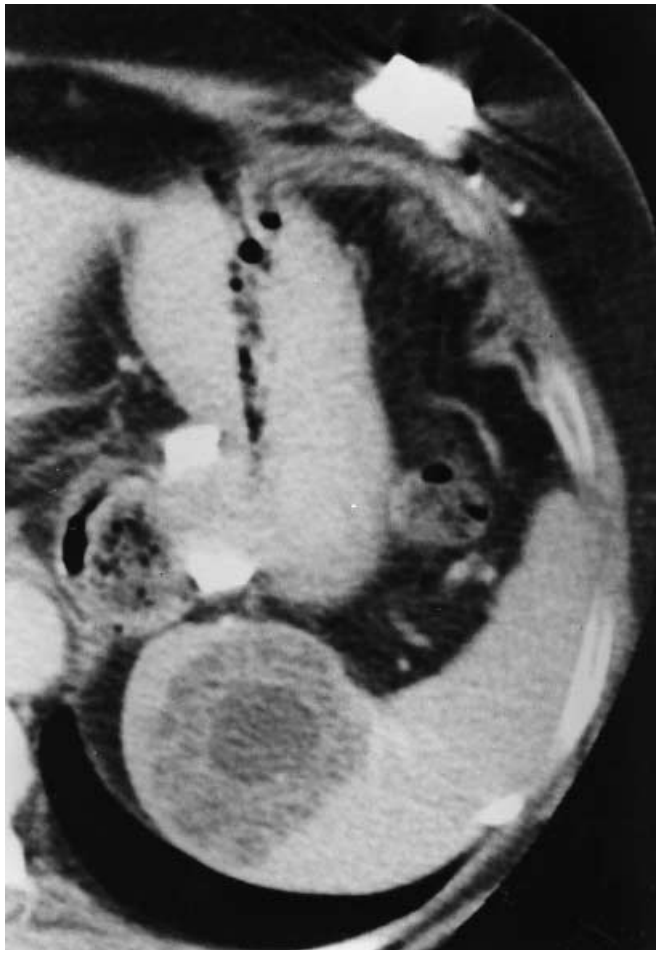

Fig. 2 Contrast-enhanced CT scan. The study confirms the presence of a large central cyst surrounded by smaller cysts. The septa between the cysts are barely visible and enhance to the same extent of the splenic parenchyma. No calcifications are seen. There is a hyperdense ring around the stomach due to a previous gastric banding procedure

posed of dilated lymphatic vessels, and contained homogeneous eosinophilic material. The rest of the spleen was unremarkable. The final diagnosis was cystic lymphangioma of the spleen.

Discussion

Lymphangiomas are usually described as benign neoplasms but should probably be considered vasoformative malformations made of abnormally dilated lymphatic vessels [2]. Lymphangiomas are usually divided into three types – capillary, cavernous and cystic – depending on the size of the dilated lymphatic channels. Capillary lymphangioma occurs more frequently on the skin of the face, the neck, and the axillary region. Cavernous lymphangiomas are most commonly seen in the skin and subcutaneous tissue, but can also occur in the lips and tongue. The cystic type is most often located in the regions of the neck and axilla.

Involvement of other regions such as the mesentery or the intra-abdominal organs is less frequent [2, 3]. Multisystem involvement of visceral organs, kidney, bones, and soft tissues is a different and extremely rare disease entity, and is frequently termed systemic cystic angiomatosis [4]. In contrast to single lymphangiomas, this form has been reported to be progressive and to have a poor prognosis [5, 6].

Lymphangiomas of the spleen are rare and can be of any of the three above-mentioned types [7, 8]. Splenic cystic lymphangioma can present as a single cystic cavity, but more often consists of multiple cysts of different sizes separated by internal septations. The ‘cysts’ are abnormally dilated lymphatic vessels which contain

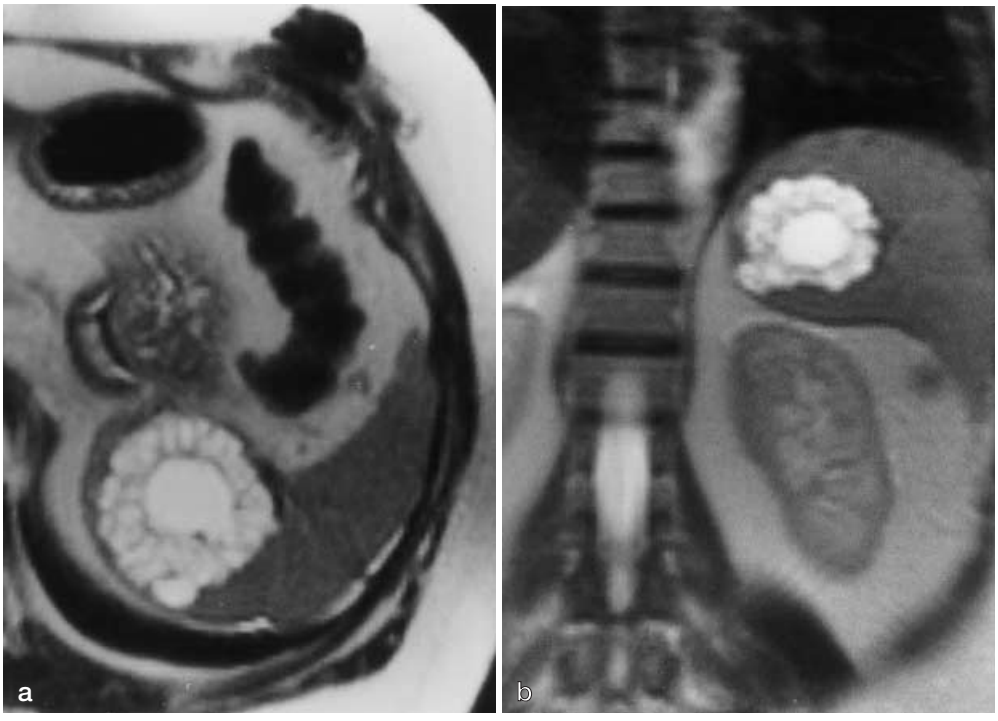

Fig. 3 T2*-weighted MR scan in axial (**a**) and coronal (**b**) planes. The septa between the cysts are better demonstrated due to the contrast with the hyperintense cystic spaces. There is no evidence of endocystic nodules of soft tissue signal intensity

eosinophilic fluid [2, 9], while the septations are made of fibrous connective tissue with rare vessels. The connective tissue of the septa may calcify.

At ultrasonography the cysts are anechoic or hypoechoic with internal debris. The septa are hyperechoic and may present detectable vasculature at color Doppler study [10]. Calcifications may be detected if they cause acoustic shadowing. In our case the intraseptal vasculature was not detectable at color Doppler. In some cases the whole spleen may be replaced by cysts with no evidence of residual splenic parenchyma. If the cystic spaces are small and below the resolution of the US technique, then portions of the mass may appear hyperechoic due to the existence of numerous reflecting interfaces, as in our case.

On CT, the appearance is that of a mass made up of multiple low-density thin-walled sharply marginated cysts which may contain mural calcifications. After intravenous injection of iodinated contrast the cystic fluid does not enhance, while the septations present moderate enhancement.

MRI usually shows a multiloculated hyperintense mass, indicating a cystic lesion, on T2-weighted images. The septa are distinctly demonstrated as hypointense bands compared with the splenic parenchyma. On T1-weighted images the cyst are usually hypointense, but may rarely be hyperintense owing to the proteinaceous nature of the fluid or internal hemorrhage [11, 12, 13]. It is not possible to reliably establish the presence of septal calcification with MRI.

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 paramagnetic contrast injection [14].

The differential diagnosis of cystic lymphangioma includes true splenic cysts, mesothelial cysts, old hematomas and echinococcosis, even in a serologically negative patient, and the extremely rare cystic hamartoma [1]. True splenic cysts, with true epithelial lining, usually show no wall enhancement. They may present septations but usually the multicystic cavities are larger than in our case. False cysts are thought to result from unrecognized splenic hematoma and may not be differentiated from true cysts; false cysts less frequently present septation [15]. Hematomas, if recent, may demonstrate MR signals typical of blood products, corresponding to their age; the wall may calcify. Echinococcal cysts, in the early phase of development, usually present as single cysts with regular margins. When they grow, they become multiloculated with septations and may be indistinguishable from any other multicystic mass; therefore they may resemble multicystic lymphangioma. Any patient with a cystic splenic mass should have a serologic test for echinococcosis. Cystic hamartomas are very rare in the spleen, and occur more frequently in the liver in the pediatric age group [16].

The normal evolution of splenic lymphangioma is variable. Small lesions are often incidentally detected. Larger lesions may increase in size and cause the com-

pression 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 [17].

Although splenic lymphangiomas are benign, Feigenberg et al. reported one patient with a malignant lymphangioma [18]. The malignant degeneration was suggested by the increased proportion of solid areas. In their patient, the endothelial cells had formed papillary excrescences within some of the cystic spaces, and solid foci in other regions of the mass. In such cases, MR imaging may facilitate the detection of solid areas by the high-contrast resolution and by the enhancement after gadolinium injection, and contribute to diagnosis of the malignant growth.

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