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Hepatic perivascular epithelioid cell tumor: A case report

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ABSTRACT

Perivascular epithelioid cell tumor (PEComa) of liver is extremely rare hepatic neoplasm with only 30 cases reported in the literature. These lesions are found mainly in young females and may present a potential pitfall in the characterisation of focal liver lesions. The biological behavior of PEComa varies from generally benign to rarely malignant and metastatic disease. We report a case of a patient with hepatic PEComa with the corresponding imaging findings on the ultrasound, contrast-enhanced ultrasound (CEUS) and hepatospecific MRI. After failed attempt to characterize the lesion by percutaneous biopsy, surgical resection was conducted and the final diagnosis was achieved.

1. Introduction

Perivascular epithelioid cell tumor (PEComa) is a mesenchymal neoplasm, predominantly affecting young female adults. The predominant site of origin for PEComa is a uterus, but the tumor may be found in various locations in the body. Cases in the liver are extremely rare [1] and up to this date, only 30 cases of liver PEComas were reported in the literature and only two of them included imaging findings on contrast-enhanced ultrasound (CEUS). The biological behavior of PEComa varies in different cases from generally benign to rarely malignant and metastatic disease [2].

2. Case report

A 24 - year old previously healthy female was referred to a gastroenterologist for unspecific pain in the lower abdominal region. The physical examination was normal and the levels of laboratory tests were within reference ranges. An ultrasound examination of the abdomen revealed normal sized liver with normal echotexture of liver parenchyma. A well-defined 20 mm hypoechogenic lesion with mass effect was identified in the segment IV of the liver. Colour Doppler analysis demonstrated hyperemia in the lesion in comparison with normal hepatic tissue (Fig. 1). No pathologic lymph nodes or other pathologic findings were noted at the examination. CEUS was performed for characterization of the liver lesion. A 1.8 ml of second-generation ultrasound contrast media SonoVue (Bracco, Italy) was used for the examination. The lesion showed homogenous hyperechogenic enhancement in the arterial phase (20–40 s post injection) (Fig. 2) and stayed iso- to hyperechogenic in comparison to surrounding liver parenchyma in portal venous (60–120 s post injection) and late phase (2–4 min post injection). On the basis of enhancement pattern and absence of diffuse liver disease, the differential diagnosis of focal nodular hyperplasia (FNH) or hepatic adenoma was made according to EFSUMB Guidelines for Contrast Enhanced Ultrasound in the Liver [3]. The patient was scheduled for US follow-up exam. This was performed after six months and it revealed an increase in the size of the lesion (25 mm) with the same pattern of enhancement. The growth of the lesion was the indication for the referral of the patient to the MRI examination of the liver.

MRI with the hepatobiliary-specific contrast agent demonstrated a 25 mm liver lesion that was hypointense on T1 and hyperintense on T2 weighted sequences and showed no notable signal drop on GRE opposed-phase sequences (Fig. 3). After the injection of the contrast medium lesion demonstrated a homogeneous hyperintense enhancement in the arterial phase and washout was noted in portal venous phase (70 s post injection) and in the late phase (2 min post injection). The lesion was completely hypointense in hepatospecific phase (Fig. 4), a feature not specific for FNH and ultrasound guided histologic puncture was subsequently indicated. The result of histologic puncture was inconclusive and the decision for surgical treatments was made.

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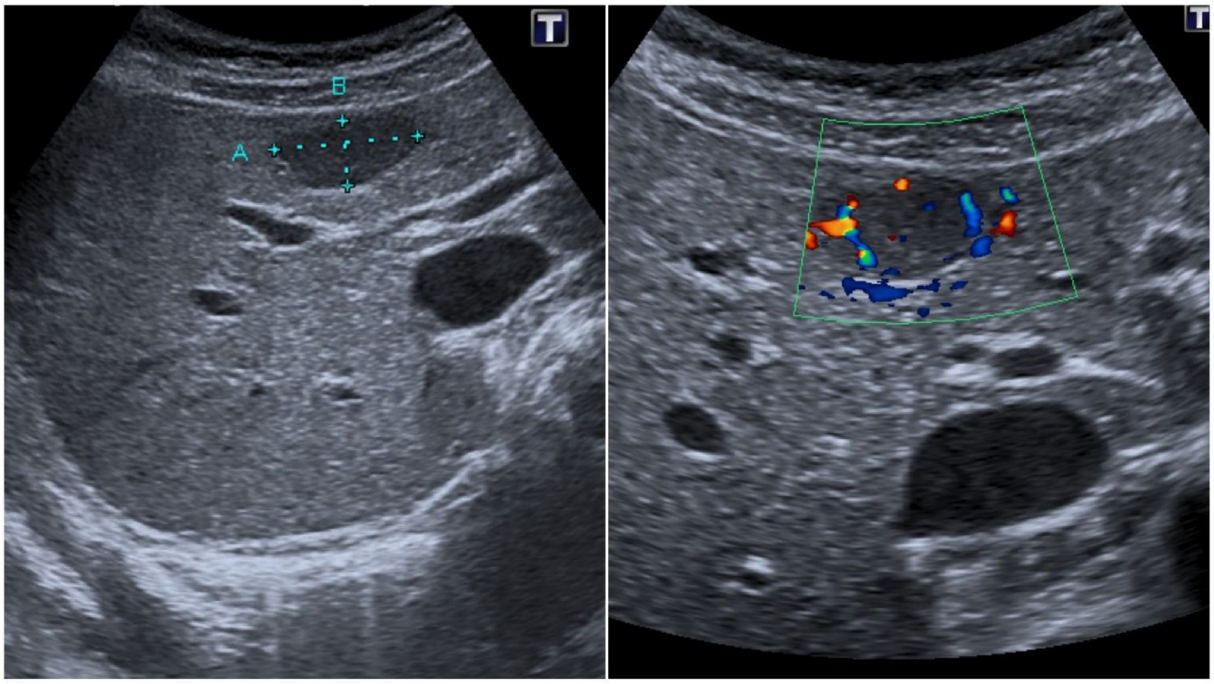

Fig. 1. Ultrasound examination of the abdomen revealing well-defined, hypoechogenic incidental lession in the liver (a). Color dopler analysis demonstrates a hypervascularity of the lession (b).

Non-anatomical resection achieved complete resection of the tumor and histological examination revealed a well-demarcated and unencapsulated tumor with epithelioid cells growing in sheets and displaying perivascular arrangements. The nuclei of tumor cells were bland with occasional regular mitoses (Fig. 5). By immunohistochemistry, tumor cells were diffusely positive for HMB45 and melan A, focally positive for smooth muscle actin and S100, and were negative for desmin. The histological features and the results of immunohistochemical stainings were consistent with PEComa of the liver.

3. Discussion

The term of perivascular epithelioid cell unifies a class of tumors that share the presence of smooth muscle and melanocytic differentiation. PEComas have been described in different organs such as the liver, uterus, vulva, rectum, heart, breast, urinary bladder, abdominal wall and are considered ubiquitous tumors. Clinical presentation of liver PEComas are unspecific and a definite pre-operative diagnosis is difficult to make due to non-specific radiological features. It might be often misdiagnosed with hepatocellular carcinoma, hemangioma, FNH and GIST tumor [4]. The first reported case of hepatic PEComa was in 2000 by Yamasaki [5]. We have performed a literature search using MEDLINE and found altogether 30 cases of hepatic PEComas to this date and only two cases of PEComas where the CEUS imaging was performed.

Tumors were usually found in healthy livers and the right lobe of the liver was the most common site. Size varied from 0.8 to 23 cm in greatest dimension (mean 8 cm). The usual ultrasonographic appearance was described as a well-defined round lesion that can be of any echogenicity at B-mode ultrasound and with a hypervascular appearance at color Doppler imaging [6].

In our case, CEUS was performed following B-mode ultrasound for lesion characterisation. There are only two case reports of PEComa studied with CEUS by Della Vigna and Akitake [7,8]. SonoVue was used as a contrast reagents by Della Vigna and Sonazoid was used by Akitake [7,8]. In both cases tumors showed same enhancement pattern as in our case - homogeneous hyperenhancement on the arterial phase, isoechogenicity on the portal vein phase, and iso- to hyperechogenicity on the late phase. In non-cirrhotic liver, this pattern of enhancement is considered diagnostic for FNH or adenoma according to EFSUMB guidelines and it can cause misdiagnosis. This pitfall in diagnosis was observed in our case an also in the case published by Della Vigna [3,7].

The MRI of the liver with hepatospecific contrast agent gadolinium-ethoxybenzyl-DTPA (GD-EOB-DTPA) was performed because of the growth of the lesion. GD-EOB-DTPA is magnetic resonance imaging contrast agent distributes into the hepatocytes and bile ducts during the hepatobiliary phase, therefore indicating hepatocytes containing lesions. Our tumor showed hyperenhancement in arterial phase, but was completely hypointense in the hepatospecific phase the, indicating lack of hepatobiliary function and ruling-out the diagnosis of FNH.

The diagnosis of PEComa was made based on histological examination of the tumor and subsequent additional immunohistochemical examination. The majority of reported cases presented benign tumors but some PEComas can show malignant potential with local recurrences and distant metastasis [9]. There is no particular treatment protocol for hepatic PEComas. Most tumors were surgically treated with good results. Only in one case of malignant hepatic PEComa sirolimus was used as neoadjuvant chemotherapy agent [10].