Hepatic epithelioid hemangioendothelioma

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A 36-year-old woman with no relevant medical or surgical history was referred to our attention for abdominal diffuse pain and mild weight loss. No risk factors such as hepatitis, alcohol abuse, oral contraceptive use, or a family history of carcinoma were reported. Liver laboratory studies were unremarkable, except for elevated gamma-glutamyl transferase (138 U/l). Viral serology tests were negative, as were serum carcinoembryonic antigen, CA 19-9, and alpha-fetoprotein.

Abdominal ultrasonography revealed multiple round hypoechoic nodules, which produced no signal at color Doppler. Computed tomography images were obtained before and after contrast injection. The unenhanced phase showed multiple hypodense lesions. On the arterial phase (35”), there was almost no contrast enhancement, whereas in the portal phase (90”), only minimal peripheral enhancement was seen; the inner part of the lesion remaining unenhanced. On T1-weighted magnetic resonance (MR) images, the nodules were hypointense, with an internal area of lower signal intensity in the largest lesions. On T2-weighted MR images, all lesions showed mild signal hyperintensity, with a core of higher signal intensity (Figure 1a). Dynamic contrast-enhanced imaging showed mild, inhomogeneous and progressive centripetal enhancement, although the center of the largest lesion remained minimally enhanced (Figure 1b). Moreover, some nodules tended to merge together, with sub-capsular nodules causing capsular retraction (Figure 1b).

The patient underwent tumorectomy, and excisional biopsy revealed epithelioid hemangioendothelioma. This diagnosis was confirmed at pathological analysis after liver transplantation performed six months later. Macroscopic pathological analysis revealed several coalescent, gray-white hepatic lesions (Figure 2a). Microscopic analysis revealed spindle-shaped tumor cells and epithelioid tumor cells with intracytoplasmic vacuoles, some of which contained red blood cells (Figure 2b). Immunohistochemical staining was positive for the endothelial marker CD31 (Figure 2c).

Epithelioid hemangioendothelioma of the liver is a rare, low-grade malignant neoplasm of endothelial origin. It typically occurs in female patients aged 20–40 years. Its diagnosis can be challenging: it can be discovered accidentally or revealed by unspecific symptoms such as right upper quadrant pain, weight loss, fatigue, and jaundice. Imaging findings can be helpful in guiding the diagnosis; characteristic imaging features of epithelioid hemangioendothelioma include multiple hepatic masses with peripheral location, capsular retraction, nodule coalescence, target-like configuration (so-called “lollipop sign”), and peripheral and delayed contrast enhancement [1].

Characteristic pathological features include intracytoplasmic vascular lumen formation, sinusoidal spreading, vessel obliteration, necrosis in the center of the lesion, and cellular pleomorphism. Positive immunohistochemistry for endothelial markers such as factor VIII, CD31, or CD34 confirms the diagnosis. High cellularity is the most significant factor predicting an unfavorable prognosis [2].

Several treatment strategies are available, including systemic/regional chemotherapy, radiotherapy, hormone therapy, thermoablation, liver resection, and liver transplantation [3]. The latter option is the most common treatment modality, because the vast majority of patients have multifocal lesions at the time of diagnosis [4]. The prognosis depends on whether extrahepatic involvement is present at the time of diagnosis, with survival rates at 1 and 5 years of 80% and 64%, respectively.
Bibliography:


Figures legends:

Figure 1: MR imaging
(a): T2-weighted MR image showing mild signal hyperintensity of the abdominal lesions, with a core of higher signal intensity.
(b) Injected portal phase showing inhomogeneous and progressive centripetal enhancement; the center of the largest lesion remaining minimally enhanced. Note the capsular retraction.

Figure 2: Pathological analysis
(a): Gross appearance of two gray-white hepatic lesions.
(b): The lesions showed spindle-shaped tumor cells and epithelioid tumor cells with vacuoles, some of which contained red blood cells (arrows) (hematoxylin and eosin, ×200).
(c): Tumor cells were positive for the endothelial marker, CD31 (CD31 immunostaining, ×200).