Hepatic epithelioid haemangioendothelioma: diagnostic imaging

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Section: Abdominal imaging
Area of Interest: Liver
Procedure: Contrast agent-intravenous
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: Ultrasound-Colour Doppler
Imaging Technique: Percutaneous
Imaging Technique: MR
Imaging Technique: MR-Angiography
Imaging Technique: MR-Diffusion/Perfusion
Imaging Technique: PET

Special Focus: Haemodynamics / Flow dynamics
Tissue characterisation Case Type: Clinical Cases

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Patient: 36 years, female

Clinical History:

The patient was referred to our hospital for an incidental detection of hepatic nodules during ultrasonographic follow up for pyeloureteral junction stenosis. Previous ultrasonographic investigations performed in a private imaging center were normal. Blood chemistry was normal; serum levels of tumour markers (a1-fetoprotein, carcinoembryonic antigen, CA19-9 and CA125) were within normal limits.

Imaging Findings:

Ultrasonography of the liver evidenced peripheral hypoechoic nodules without vascular signals at color Doppler examination (Fig. 1).

Nodules were hypoattenuating on contrast-enhanced CT (Fig. 2); no abnormal metabolic activity were appreciable on whole body F18-FDG-PET (Fig. 3).

Hepatic lesions were hypointense on T1W-images (Fig. 4a) and hyperintense on T2W-images on a 3T-MR equipment (Fig. 4b, c), with different signal intensity on DWI (Fig. 5); some masses showed a target-like pattern on T2W-images (Fig 4b, c). Superficial nodules were associated to a focal capsule retraction; none of them exhibited a pseudocapsule (Fig. 4d). Dynamic MRI with liver-specific contrast agent (Gd-BOPTA) demonstrated progressive but slow enhancement of nodules, which did not retain the contrast medium in the hepatobiliary phase (Fig. 6a-d).

Histology from US-guided specimen showed sparse single atypical epithelioid uni- or multivacuolated cells and thin capillaries embedded in a dense fibrous matrix positive for CD34 markers at immunostaining (Fig. 7).
Discussion:

Hepatic epithelioid hemangioendothelioma (HEH) is a rare, low-grade primary malignancy of endothelial origin with a variable clinical course [1]. Presenting symptoms are nonspecific and diagnosis is often incidental [1]. Vascular invasion can cause a Budd-Chiari syndrome or portal hypertension [2]; metastases (usually in the lungs) or an abdominal diffusion can occur [1].

HEHs comprise a nodular type and a diffuse type, the extremes of a continuous spectrum from a localized to an extensive liver involvement [1]. Small cords or nests of dendritic-shaped cells and epithelioid cells with cytoplasmic vacuoles embedded in a fibrous stroma are found at microscopy; regressive changes may result in cell atrophy and a densely hyalinized stroma in the tumour core [1, 3]. HEH shows a higher peripheral cellularity and tendency to infiltrate acini; invasion of peripheral vasculature produces a narrow avascular peritumoural ring [1, 3]. Small underrepresentative biopsies from HEH or from sclerosing or hypocellular areas may be extremely difficult to be differentiated from similar lesion, such as intrahepatic cholangiocarcinoma, angiosarcoma and sclerosed haemangioma. Immunohistochemistry is valuable for the final diagnosis by evidencing a positivity in the cell tumours for at least one of endothelial markers (FVIII-RAg, CD34, CD31) [1].

HEH nodules are usually hypoechoic at ultrasonography; hyperchoic or isoechoic nodules may exhibit a hypoechoic rim [2, 3]. Sonographic pattern does not correlate to the tumour size [3]. No blood flow signals are evident on Doppler imaging [2, 4]. Nodules are hypoattenuating on non-enhanced CT images and show some enhancement with a target-like appearance on post-contrast scans; a delayed enhancement depends on the predominance of regressive changes [2, 3, 5]. Calcium deposits are found in 20% of cases [1, 3]. Superficial nodules can cause a capsular retraction [3]. Nodules can resemble a lollipop when a hepatic or portal vein terminates at or just within the periphery of the nodule [6]. Hypermetabolic activity on F18-FDG PET depends on tumour cellularity; usefulness of additional delayed imaging (dual-time-point technique) is questioned in differentiating HEH from benign lesions [7, 8]. HEHs are hypointense on T1W-MRI, occasionally with a haemorrhagic component, and hyperintense with a more hyperintense core on T2W-MRI (target sign) [2-5, 8]. The target pattern is also conspicuous on CE-MRI; a progressive centripetal enhancement is appreciable in the delayed phases [2-5, 8]. The avascular rim can produce a three-layer target-sign [3, 5]. Despite of reported cases [5], HEH can show a low ADC value likely because of a dense stroma.

Differential Diagnosis List: Hepatic epithelioid haemangioendothelioma, Hypovascular metastases, Lymphoma, Intrahepatic multifocal cholangiocarcinoma

Final Diagnosis: Hepatic epithelioid haemangioendothelioma

References:


Description: Peripheral hypoechoic nodule with no vascular signals. Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
**Description:** Hypoattenuating hepatic nodules. Amputation of a portal branch entering the more central nodule is appreciable. **Origin:** Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Figure 3

Description: T1 out-of-phase GE imaging (TR/TE 100/1.23): hypointense hepatic nodules. Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Description: HASTE T2W-FS-MRI (TR/TE 1600/95): high signal intensity of hepatic masses; the lesion located in the central portion of S8 shows a more pronounced hyperintense core. Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Description: Coronal HASTE T2W-MRI (TR/TE 1400/89): target-like appearance of two hepatic nodules. Pyelectasis of the right kidney is visible. Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Description: T1 out-of-phase GE imaging (TR/TE 100/1.23): a peripheral nodule with capsular retraction. Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Figure 4

a

Description: DWI b50: two hyperintense subcapsular nodule in the right lobe of the liver.

Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy

b

Description: DWI b800: restriction to water diffusion is more pronounced in the more ventral nodule.

Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Description: ADC map: the nodules have different ADC values (700 mm2/s and 1700 mm2/s). Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Figure 5

Description: Arterial phase: nodules appear hypointense relative to the liver; some of the lesions exhibit some peripheral enhancement (target sign). Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Description: Delayed venous phase: centripetal filling of nodules. Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Description: 10 minutes after the contrast medium administration: nodules are homogenously hyperintense relative to intervening parenchyma. Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
**Description:** Hepatobiliary phase: lesions do not take up the liver-specific contrast medium.

**Origin:** Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy
Figure 6

Description: Sparse single atypical epithelioid uni- or multivacuolated cells (upper half, yellow circles - H/E stain x200) and thin capillaries embedded in a dense fibrous matrix positive for CD34 markers at immunostaining (lower half). Origin: Emanuele D’Amore, U.O. Anatomia e Istitutopatologia, Ospedale San Bortolo, Vicenza, Italy
Description: No abnormal hyperactivity; however, the liver shows an inhomogeneous activity. Origin: Sergio Savastano, UO Radiologia, Ospedale San Bortolo, Vicenza, Italy