REVISTA ESPAÑOLA DE
ENFERMEDADES DIGESTIVAS
The Spanish Journal of Gastroenterology

Title:
An incidental hepatic epithelioid hemangioendothelioma in a patient with chronic hepatitis: lost in the maze

Authors:
Lan Zhang, Yanru Zhou, Jiajia Zhang

DOI: 10.17235/reed.2021.8322/2021
Link: PubMed (Epub ahead of print)

Please cite this article as:
Zhang Lan, Zhou Yanru, Zhang Jiajia. An incidental hepatic epithelioid hemangioendothelioma in a patient with chronic hepatitis: lost in the maze . Rev Esp Enferm Dig 2021. doi: 10.17235/reed.2021.8322/2021.

REVISTA ESPAÑOLA DE
ENFERMEDADES DIGESTIVAS
The Spanish Journal of Gastroenterology

An incidental hepatic epithelioid hemangioendothelioma in a patient with chronic hepatitis: lost in the maze

Lan Zhang[1], Yanru Zhou[1], Jiajia Zhang[2]

1 MRI Department, the First Affiliated Hospital of Henan University of Chinese Medicine, Zhengzhou, Henan, China

2 Department of Radiology, Gold Coast University Hospital, Southport, Queensland, Australia

Correspondence to:

Lan Zhang, M.D., MRI Department, the First Affiliated Hospital of Henan University of Chinese Medicine, No 19 Renmin Road, Zhengzhou, Henan, China

Telephone: +86-0371-66036752

Email: 13837187787@163.com

Conflict of Interest

All authors have no conflicts of interest to declare.

Ethical Approval

This study was approved by the local institutional review board and informed consent was obtained from the patient for the publication.

Financial Disclosure: The authors declared that this study has received no financial support.

Dear Editor,

Hepatic epithelioid angioendotheliomas (HEHE) are rare vascular tumors with low to intermediate malignancy (1). Imaging plays an important role in the diagnosis of HEHE. However, imaging characterization of HEHE still remains challenging, since HEHE is commonly misdiagnosed as hepatocellular carcinoma, cholangiocarcinoma or metastases (2).

REVISTA ESPAÑOLA DE
ENFERMEDADES DIGESTIVAS
The Spanish Journal of Gastroenterology

A 60-year-old woman with a 20-years history of hepatitis B presented with 2 weeks of dull pain in the right upper quadrant and intermittent nausea. Her serum alpha-fetoprotein(8gn/ml), carbohydrate antigen125(10.3U/ml), carbohydrate antigen 199 (9.9U/ml)were within the normal range, and her liver function was graded as Child-Pugh A. Magnetic resonance imaging (MRI) showed a solitary lesion with a multi-layered target appearance resembling a "maze" in Segment VIII of the liver. On diffusion-weighted image and T2-weighted images the mass showed six layers with alternate high and low signal (Fig. 1a, b). However a three-layered target appearance was recognizable on T1-weighted images (Fig.1c). On Gd-EOB-DTPA enhanced-MRI, the lesion showed rim enhancement on the arterial and portal phase (Fig.1d, e), and became a low signal target with a hypointense core on the hepatobiliary phase (Fig.1f). The patient underwent laparoscopic lobectomy and histopathology confirmed hepatic epithelioid hemangioendothelioma (HEHE).

One of characteristic imaging features “halo sign” or target appearance consists of three layers of varying signal on MRI, which is known to be related to necrosis, prior hemorrhage, thrombosis and stroma (3). Interestingly, our case shows a solitary lesion with as many as six layers. Overall appearances are more like a "maze", which is a challenge for diagnosis. This maze-like manifestation actually makes hepatocellular carcinoma or metastasis as the main differential being very unlikely. Other differentials such as cholangiocarcinoma, atypical hemangioma and angiosarcoma are also unlikely to show more than three layers. The lesion shows a seed-like central hypointense core on the hepatobiliary phase as "core pattern". Recognition of core pattern can further assist in the differential diagnosis from hepatocellular carcinoma or cholangiocarcinoma (4).

In conclusion, our case highlights HEHE needs to be taken into consideration when a multi-layer target lesion is found incidentally in the patient with chronic hepatitis – don’t get lost in the maze.

Reference

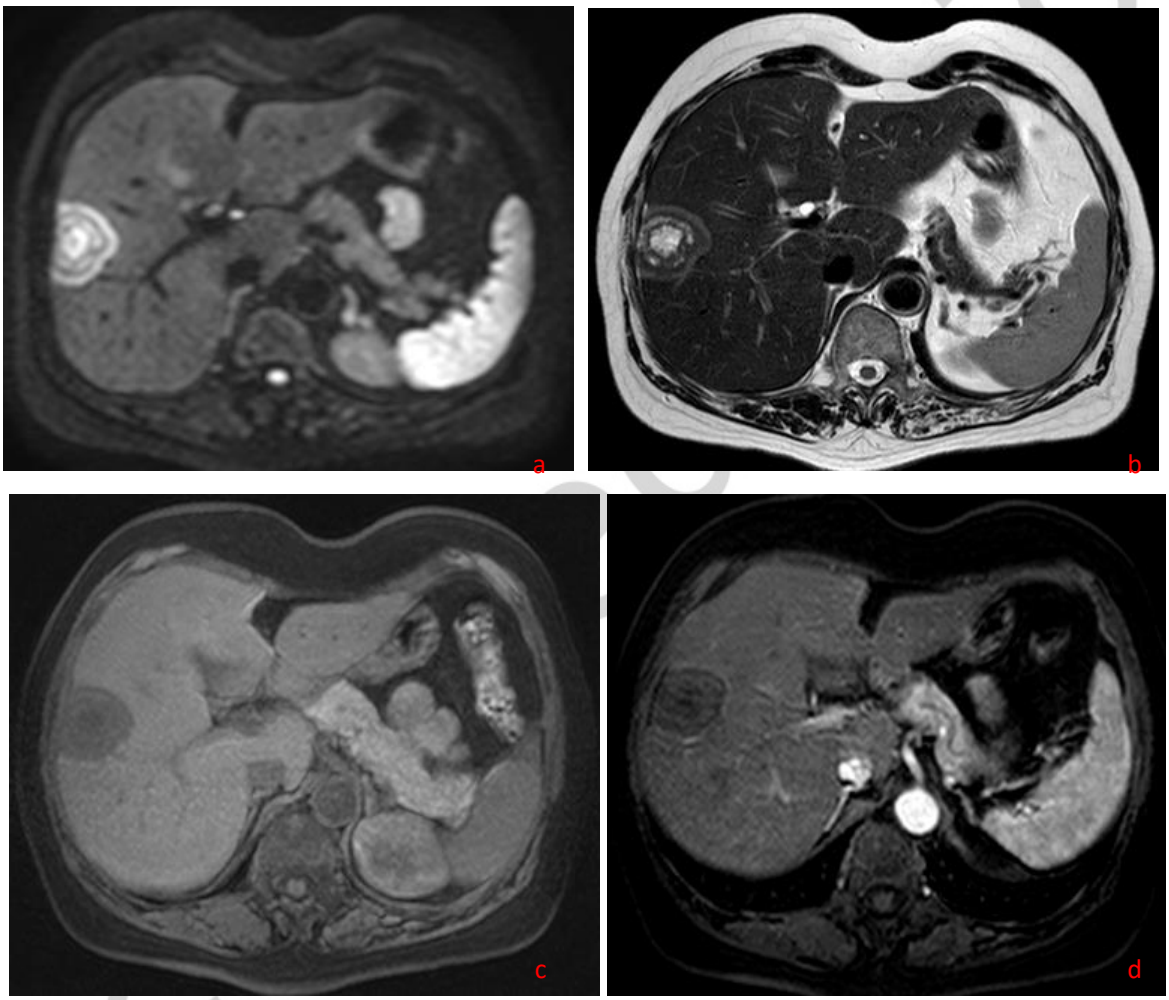
1 Alventosa Mateu C, Urquijo Ponce JJ, Herráez Cervera B, García Llabata L. Epithelioid hemangioendothelioma: a rare cause of a liver tumor. Rev Esp Enferm Dig 2021;113:69-70.

2 Cifrián Canales I, García Bernardo CM, Contreras Saiz E. Hepatic epithelioid hemangioendothelioma: a rare vascular tumor with an unpredictable diagnosis. Rev Esp

Enferm Dig 2021;113:300-301

3 Epelboym Y, Engelkemier DR, Thomas-Chausse F, et al. Imaging findings in epithelioid hemangioendothelioma. Clin Imaging 2019;58:59-65.

4 Lee JH, Jeong WK, Kim YK, et al. Magnetic resonance findings of hepatic epithelioid hemangioendothelioma: emphasis on hepatobiliary phase using Gd-EOB-DTPA. Abdom Radiol (NY) 2017; 42:2261-71.



REVISTA ESPAÑOLA DE
ENFERMEDADES DIGESTIVAS
The Spanish Journal of Gastroenterology

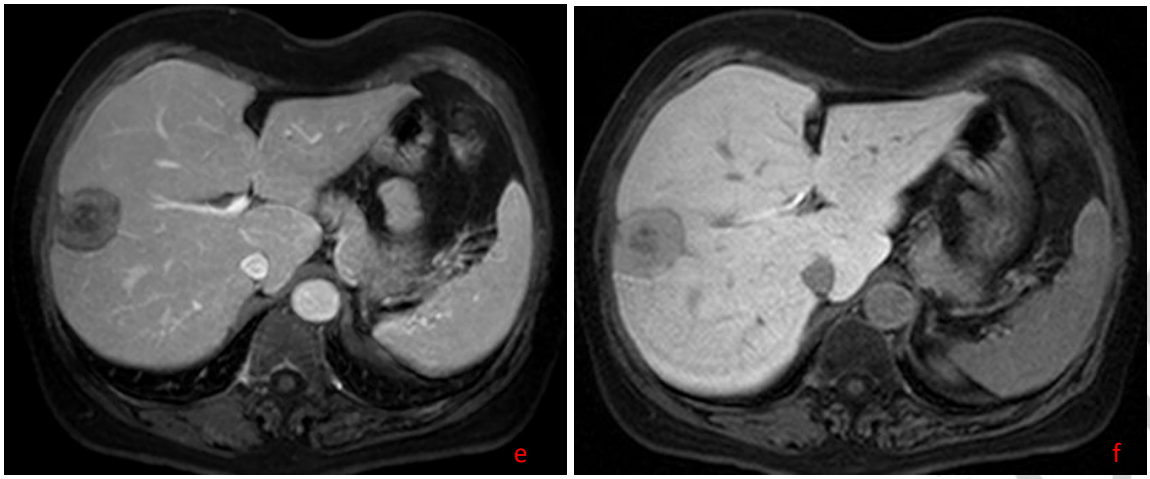


Fig.1 The tumor shows a six-layered heterogeneous signal intensity on DWI (a) and T2WI (b), and a three-layered target appearance on T1WI (c) with rim enhancement on the arterial and portal phase (d, e), and as a low signal target with a hypointense core on the hepatobiliary phase (f).