Journal of Pharmacy and Pharmacology 6 (2018) 153-156
doi: 10.17265/2328-2150/2018.02.007

Epithelioid Hemangioendothelioma of the Liver—Case Report

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Abstract: EH **(**Epithelioid hemangioendothelioma) is a rare vascular malignant neoplasm. Only 434 cases had been reported until 2006. Symptoms may be nonspecific, but it also can involve hepatic veins resulting in Budd-Chiari syndrome. The main differential diagnosis is cholangiocarcinoma, sometimes only distinguishable through immunohistochemistry study. We report a case of a young woman diagnosed with hepatic EH and listed for liver transplantation.

Key words: EH, liver malignancy, liver transplantation.

1. Introduction

EH (Epithelioid hemangioendothelioma) is a rare vascular malignant neoplasm that affects mostly female patients after the 4th decade [1, 2]. Here we report a case of a female patient with a history of inflammatory bowel disease presenting with nonspecific abdominal symptoms. Magnetic resonance showed a large hepatic lesion and, after unsuccessful surgical resection attempt, biopsy showed a patter compatible with EH.

2. Case Report

Patient ALC, feminine, 46 years old, hairdresser, is natural from the metropolitan region of Porto Alegre, Brazil. She seeks emergency presenting weight loss (approximately 4 kg), abdominal pain (constant, on right hypochondrium, with dorsal irradiation, partial improvement with common painkillers and unrelated to alimentation) and daily fever (37-40 °C at evening) for 50 days. No other symptoms associated. Her evacuation frequency was normal (1-2 evacuations a day, no pathological signs). She has been previously diagnosed with ulcerative colitis (20 years ago) and follows up with a proctologist (previous use of mesalazine, suspended more than 2 years ago). She denied any other comorbidity, continuous use of medication, surgeries and allergies. On physical exam, patient presented a good complexion, conscious and coherent, slightly hypocolored mucosa, anicteric and stable vital signs. Cardiac and respiratory systems examination was normal. Abdominal exam shows a painful right hypochondrium and a palpable enlarged liver (no splenomegalia or ascites). Recently, she did ambulatory routine exams, including thorax radiography, uterine cervix cytopathological test and mammary ultrasound, all normal. In admission, blood tests were done (Table 1), including viral serologies (HIV, hepatitis B, hepatitis C and syphilis) that were negative.

Still in the emergency department, an abdominal ultrasound was performed and it showed heterogeneous echogenicity of the liver, mainly on the right side, due to presence of agglomerated hypoechoic

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Table 1 Laboratory Exams.

Laboratory exams	Referred value	Result
Hematocrit	36%-48%	37%
Hemoglobin	11.6-15.6 g/dL	11.7 g/dL
Leucocytes	3,600-11,000	6,630
Platelets	150.000-400.000	364.000
Albumin	3.5-5.5 g/dL	4.2 g/dL
Total Bilirrubin	0.3-1.2 mg/dL	0.6 mg/dL
Direct Bilirrubina	0.1-0.4 mg/dL	0.3 mg/dL
Alcaline Fosfatase	20-130 U/L	192 U/L
AST	Até 34 U/L	31 U/L
ALT	10-49 U/L	39 U/L
INR (TP activity)	- (> 50%)	1.22 (64%)

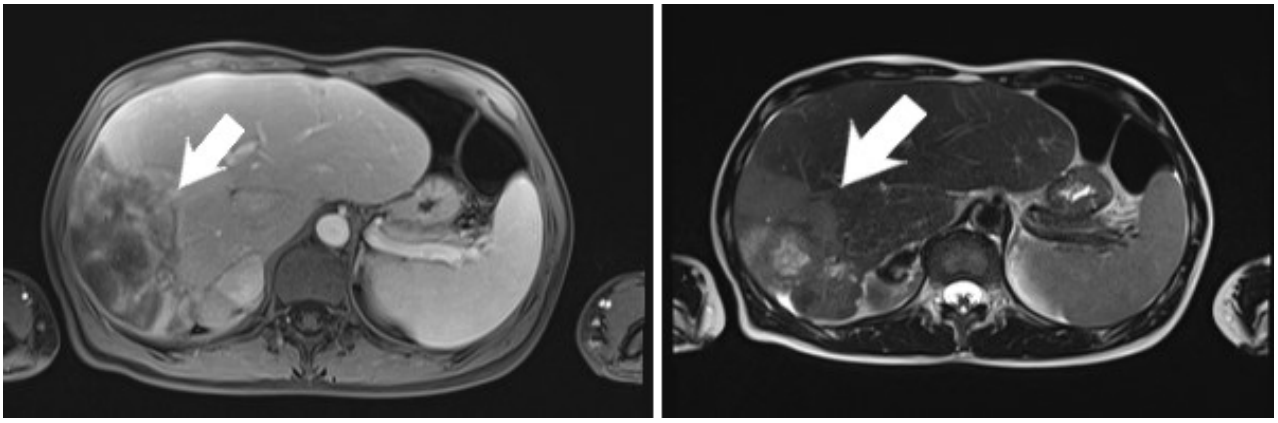

Fig. 1 Abdominal magnetic resonance images. Left image: T1; Right image: T2; White arrow: Infiltrative lesion invading nearly the entire right lobe, heterogeneous signal intensity and central necrosis.

nodular images (the largest measuring 5.2 cm) suggesting metastasis or another etiology. Patient underwent a colonoscopy that detected just a hyperemic area on the hepatic angle compatible with ulcerative colitis activity (anatomopathological test excluded neoplasia). Next, magnetic resonance imaging (Fig. 1) revealed a rounded edge liver, compensatory augmentation of left and caudate lobe and a single infiltrative lesion, invading nearly the entire right lobe and part of segment IV, with heterogeneous signal intensity and contrast enhancement in the late phase associated with central necrosis and subtle intrasegmental biliary duct dilation and right lobe atrophy, compatible with primary neoplasia (cholangiocarcinoma was considered). The lesion extended towards the hepatic hilus causing obstruction of the right portal vein and right hepatic vein. The median hepatic vein was partially invaded by the tumor on its distal branches. Also, the lesion had an intimate relation with the gallbladder. It was noted a small amount of perihepatic and perivesicular fluid.

Tumoral markers were tested (alpha-fetoprotein, carcinoembrionary antigen and CA 19.9) and were all in the range of normality. After staging (no evidence of secondary implants) and hepatic volumetric measurement, the patient underwent laparotomy with a right hepatic trisegmentectomy plan for curative purposes. However, during the surgery two lesions were found on the contralateral hepatic lobe (intraoperative frozen section analysis confirmed adenocarcinoma). The resection plan was aborted and the surgeon opted to biopsy the original lesion for histological definition (Fig. 2). Histopathological findings, in association with immunohistochemical study (Vimentina, Factor VIII, CD31 and CD34 positive), were compatible with EH of the liver. The

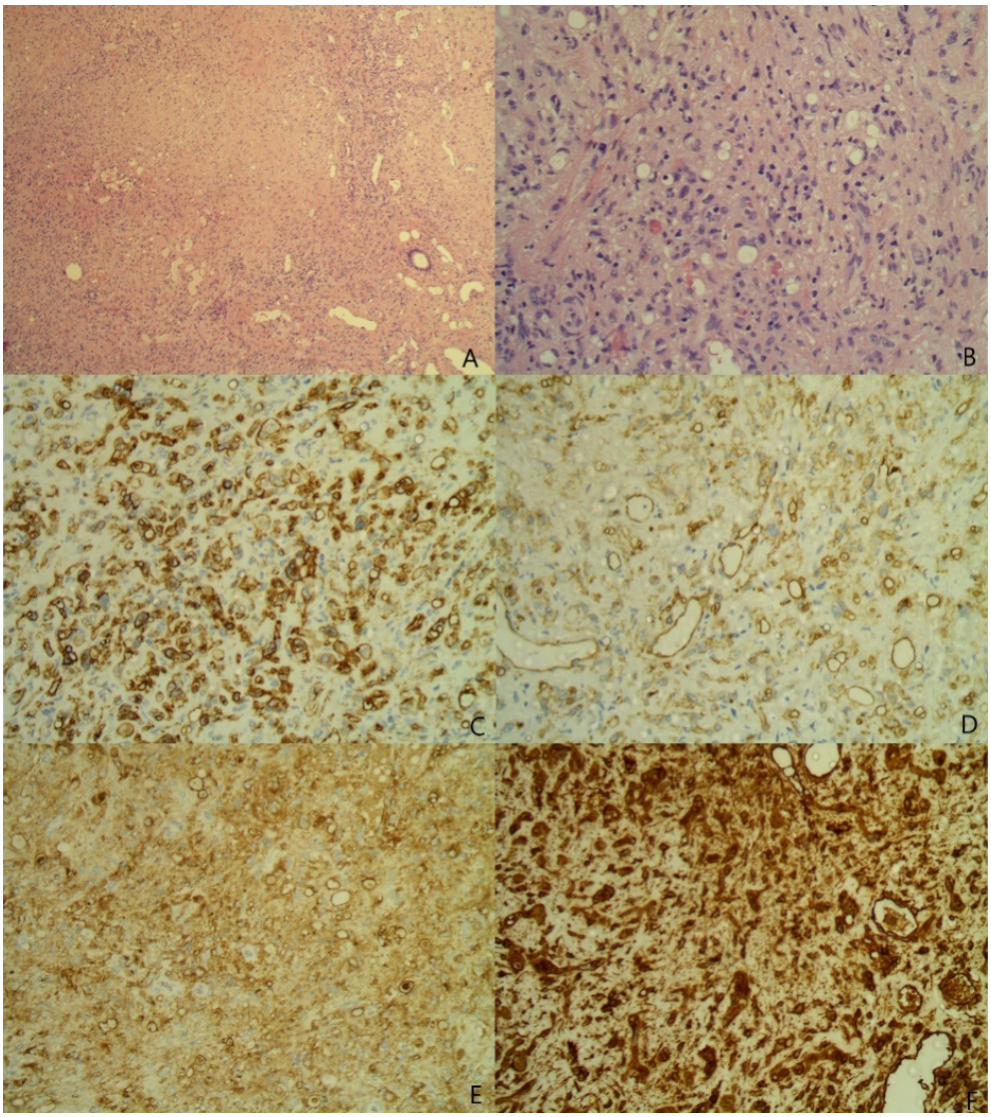

Fig. 2 Pathological findings compatible with EH. A: Hematoxylin and eosin stain (HE), 10× augmentation; B: HE, 40× augmentation; C: Immunohistochemical panel—CD31 positive for malignant cells; D: Immunohistochemical panel—CD34 positive for malignant cells; E: Immunohistochemical panel—Factor VIII positive for malignant cells; F: Immunohistochemical panel—Vimentina positive for malignant cells.

patient was referred to oncology and liver transplant service, but she died of liver insufficiency two months after the diagnose.

3. Discussion

We reported a case of EH, a rare vascular malignant neoplasm of unknown etiology that affects the liver and other organs. From 1984 to 2006, 434 cases were reported and only 252 of them were primary of the liver [3]. Main symptoms include constitutional symptoms, palpable abdominal mass and abdominal pain, but it also can result in Budd-Chiari syndrome when hepatic veins are involved [4]. Imaging exams show one or multiple intensively vascularized or calcified masses that can affect the entire liver [5]. Despite being considered an indolent tumor, it has high regional and distant metastasis rates (up to 75%) [6]. Anatomopathological study reveals a lesion in the central zone, hypocellular in relation to the peripheral zone, with preserved lobular architecture. The main differential diagnosis is cholangiocarcinoma (sometimes only distinguishable through

immunohistochemistry study of CD31, CD34 and factor VIII) [7, 8]. Treatment usually requires surgical approach (resectable cases) or liver transplant, due to the limited response to radiotherapy and chemotherapy [9]. Liver transplant, despite its rare indication (0.8% of all EH cases), shows similar results to non-neoplastic indications, with a disease-free survival in 5 years of 46%-82% [10, 11].

4. Conclusions

The reported case of EH had a poor outcome, despite all measures taken and the possibility of liver transplantation. As it is a rare disease, there are few treatment options other than lesion resection. It is important to emphasize the significance of early diagnose, which relies on accurate imaging interpretation and histological/immunohistochemical evaluation for differential diagnosis.

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