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Epithelioid Hemangioendothelioma of the Liver: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Epithelioid hemangioendothelioma of the liver (EHE) is a rare primary solid tumor of vascular origin [1-4]. EHE represents about 0.4% of all sarcomas and only 20% develop in the liver [5]. It occurs in a non-cirrhotic liver and its pathogenesis remains unknown. The involvement is often multinodular and may mimic metastases of a primary extrahepatic lesion and may delay the diagnosis. The clinical and biological signs are non-specific. Anatomopathological diagnosis can be difficult, requiring the precious help of immunohistochemistry. We report the clinical case of a 22-year-old patient admitted with febrile right hypochondrial pain, peritoneal and pleural effusion syndrome with a severe alteration of general condition. The diagnosis of hepatic EHE was proven on immunohistochemical study of the liver biopsy. The evolution was marked by the death of the patient.

Keywords: Liver; Epithelioid hemangioendothelioma; malignancy; anatomopathological diagnosis.

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1. INTRODUCTION

Epithelioid hemangioendothelioma of the liver (EHE) is a rare vascular tumor with intermediate malignant potential, first described at the expense of soft tissue in 1982 by Weiss et al [2,6]. It can involve several organs, including the liver, lung and bone. Hepatic localization is exceptional and usually multifocal, which may radiologically simulate secondary localizations [1,7]. We report the clinical case of a 22-year-old patient with an epithelioid hemangioendothelioma of the liver simulating secondary lesions and proven by immunohistochemical study.

2. CASE PRESENTATION

A 22-year-old patient, chronic user of hookah weaned 6 months ago, was hospitalized for febrile pain of the right hypochondrium. The history of the disease went back to seven months, by the progressive onset of pain in the right hypochondrium associated with a fever quantified at 38-39° degree without icterus, the whole evolving in a context of alteration of the general state made of asthenia, anorexia and uncalculated weight loss. The clinical examination showed a painful hard hepatomegaly with a hepatic arrow at 18cm and ascites without any sign of portal hypertension or hepatocellular insufficiency. Biologically, the blood count showed a normocytic normochromic regenerative anemia at 8 g/dl with hyperleukocytosis at 12470 elements/mm3, a correct platelet count with a high C- reactive protein at 227 mg/l. The hepatic work-up showed a biological cholestasis with gamma-GT at 178IU/L, alkaline phosphatases at 292IU/L with a slight elevation of total bilirubin at 13.6 mg/l with a predominance of conjugated at 8. 1 mg/l and minimal cytolysis on AST at 58IU/l and normal ALT at 23IU/l. albuminemia was low at 26g/l with prothrombin and factor V at 80% and 119% respectively. Abdominal ultrasound showed an enlarged liver, with regular contours, heterogeneous, associated with multiple nodular lesions and a peritoneal effusion layer. The thoracoabdomino-pelvic CT scan with contrast injection showed multiple bilateral diffuse pulmonary nodules of random arrangement, the largest at the right apical level of 10×8mm and left basal level of 9×8mm, with a small right pleural effusion (Figs. 1, 2). At the abdomino-pelvic level, the liver was increased in size by 20 cm, with bumpy contours, heterogeneous, with multiple nodular and patchy lesions, hypodense, confluent, not enhanced after injection of the contrast , the most voluminous one is under capsular defect straddling segments V, VI and VII measuring 180 mm anteroposteriorly, 103 mm in width and 171 mm in height, associated with a medium-sized peritoneal effusion without deep abdominal adenopathies or suspicious-looking bone lesions (Figs. 3, 4). All this suggests secondary pulmonary, hepatic and peritoneal localizations. The etiological investigation was negative in search of a primary tumor, by means of fibroscopy and colonoscopy. Tumor markers (CEA, CA19-9 and alpha-feto-protein) were negative. An echo-guided liver biopsy was performed showing a largely necrotic biopsy core with atypical cells surrounding vascular slits, sometimes scattered, of spindle-shaped and rounded form with a dense and anisokaryotic nucleus and a poorly limited cytoplasm (Fig. 5). On immunohistochemical complement the expression of CD31 and CD34, without expression of HHV8 in favour of a hepatic epithelioid hemangioendothelioma (Figs. 6, 7). The evolution was marked by the worsening of the clinical picture with the appearance of jaundice, bilateral pleurisy and the biological picture with gamma-GT at 214IU/l, PAL at 365IU/l, total bilirubin at 76.4mg/l with a predominance of conjugated bilirubin at 59.7mg/l, acute cytolysis with a level of ASAT at 642IU/l, ALAT at 99IU/l and a PT at 40%. The patient died before recovery of the result from the IHC study.

3. DISCUSSION

Epithelioid hemangioendothelioma is a rare solid tumor, vascular in nature, of uncertain evolution, with intermediate malignancy between benign hemangioma and angiosarcoma, first described in 1982 by Weiss et al [8,2,6]. It usually develops in soft tissue or bone. This disease usually affects subjects in their fourth decade with a slight female predominance [3,9]. Primary hepatic localization of EHE is exceptional and approximately 50% of cases are initially metastatic, with pulmonary or bone involvement at the time of diagnosis [1,2]. No etiological factors have been identified, and there is no evidence of contraceptive role [4,6,9]. The discovery circumstances of liver HEE are very heterogeneous and non-specific. In the majority of cases, it may present with right hypochondrial pain, altered general condition, and fever as in our clinical case. The discovery can also be fortuitous during an abdominal ultrasound. And exceptionally, it can be revealed by a Budd Chiari syndrome following invasion of the suprahepatic veins, hemoperitoneum by intraperitoneal rupture, hepatocellular insufficiency or portal hypertension [10,11].

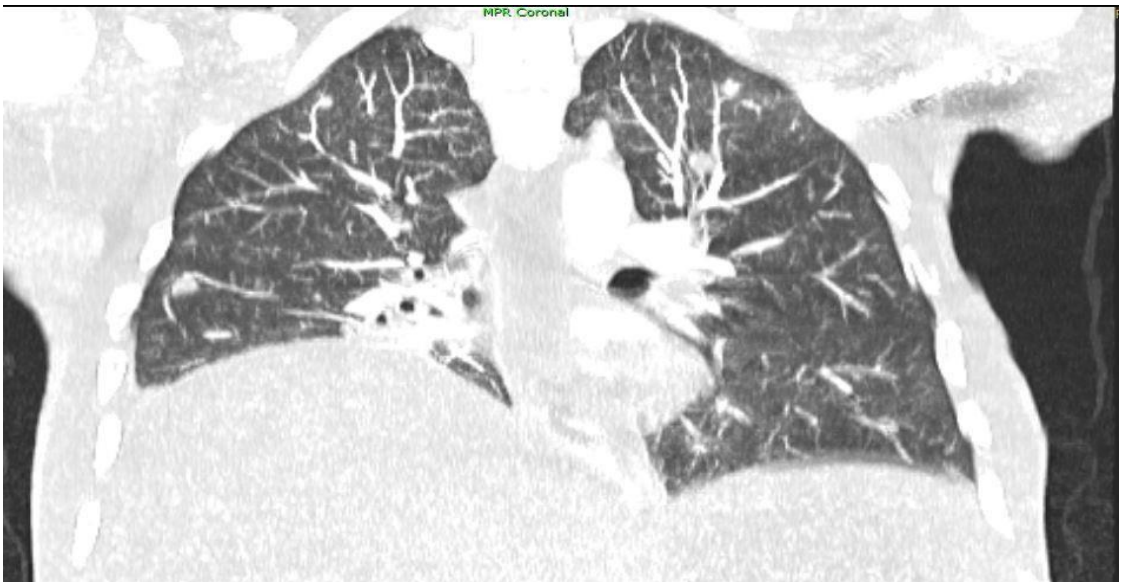

Fig. 1. Chest CT longitudinal section with injection of multiple bilateral diffuse pulmonary nodules,right apical and left basal, with a small right pleural effusion

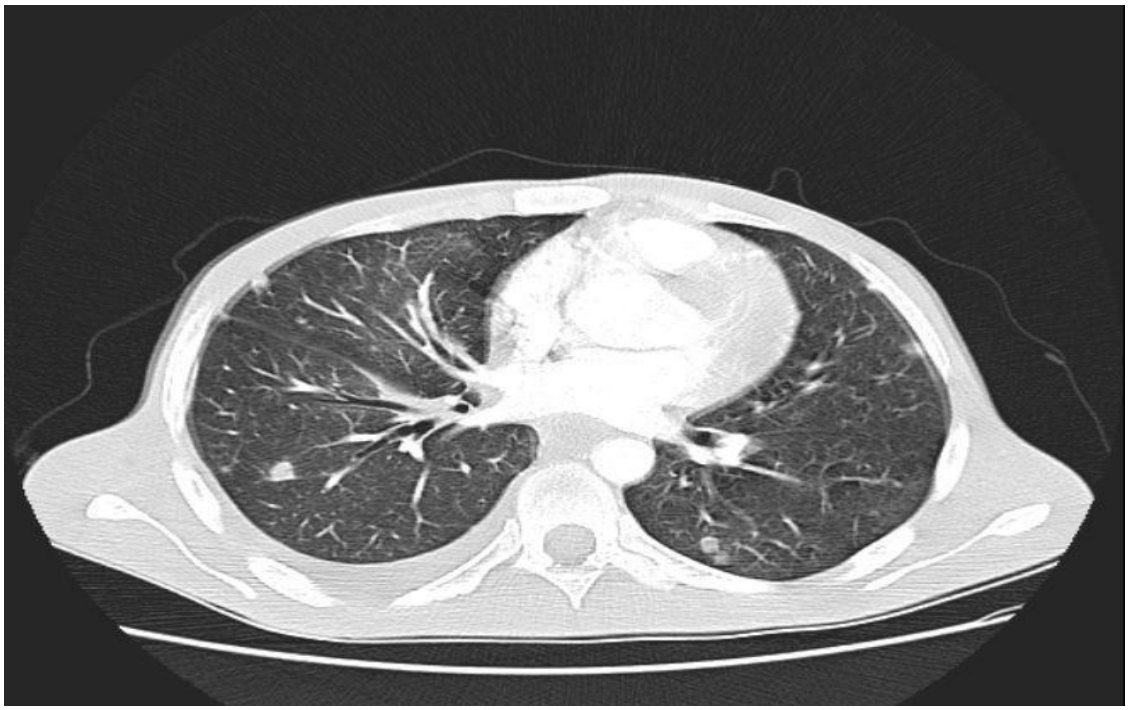

Fig. 2. Chest CT axial section with injection of multiple bilateral diffuse pulmonary nodules, right apical and left basal, with a small right pleural effusion

Biologically, there is no specific marker and liver balance disturbances are inconsistent at the time of diagnosis. Moderate elevation of alkaline phosphatases is observed in 75% of cases, gamma-glutamyl transferases in 16% of cases and transaminases in 10% of cases [2,12,6,13]. Tumor markers (alpha-fetoprotein, CEA and CA19-9) were negative. The radiological appearance is not unambiguous; in the early phase, it is often multinodular with a halo appearance. These lesions will coalesce to form diffuse, infiltrative lesions that may mimic metastases of a primary extrahepatic lesion and may delay

diagnosis [7]. On CT scan, nodular lesions are hypodense on non-injected time with halo contrast on injected time and become isodense on portal time [12]. On magnetic resonance imaging, the lesions are hypersignal on T2 sequence with a necrotic center, often hyposignal after gadolinium injection [1,3,10]. Our patient had a CT appearance in favor of secondary hepatic, pulmonary and peritoneal lesions.

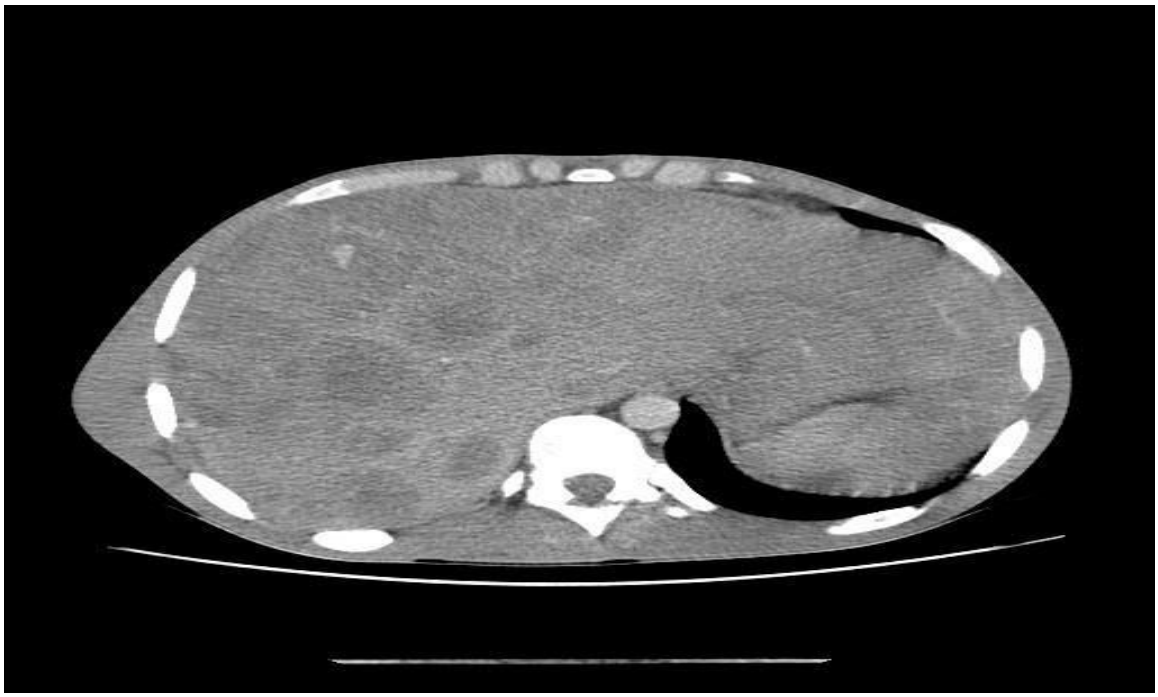

Fig. 3. Axial section abdominal CT the liver was increased in size with bumpy contours, heterogeneous, seat of multiple lesions nodular and patches, hypodense, confluent non-enhanced after injection of PDC

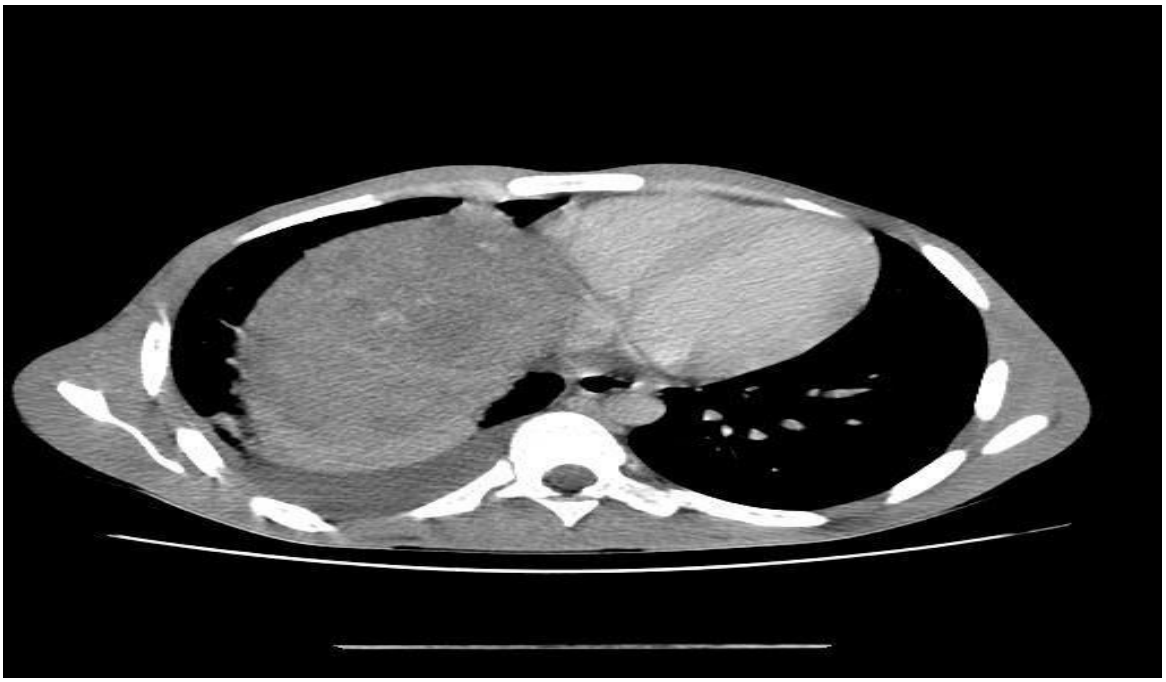

Fig. 4. Axial section abdomino-pelvic CT rising peritoneal effusion

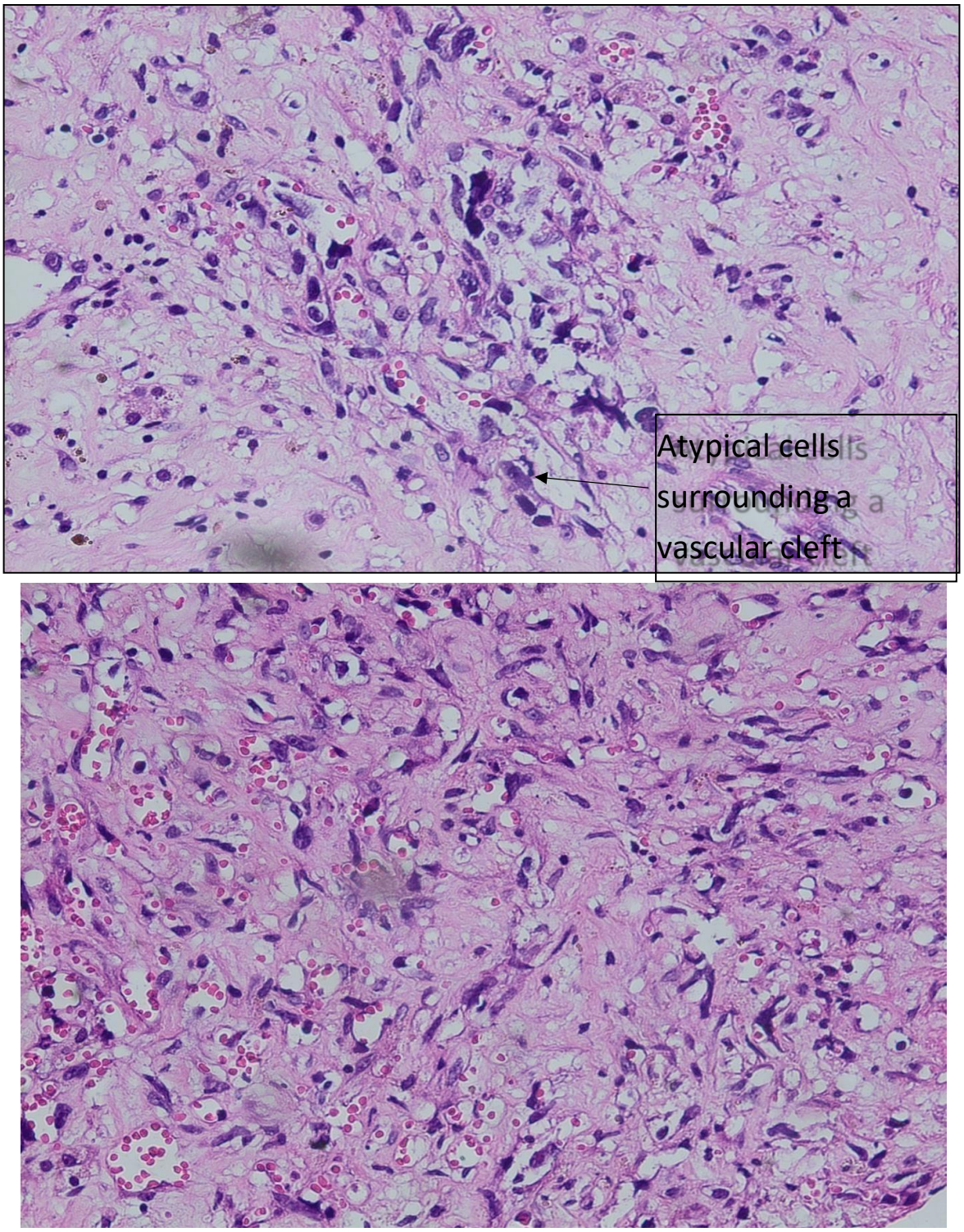

Fig. 5. Liver biopsy (Hematoxylin and eosin stain, original magnification x 20): It is a fibrous fragment with vascular slits bordered by atypical cells with a dense, irregular nucleus and an eosinophilic cytoplasm that is sometimes vacuolated

Extrahepatic localizations are common and present in 30% of cases at the time of diagnosis, and our patient had peritoneal and pulmonary metastases at the time ofdiagnosis [11].

The histological diagnosis is difficult and often erroneous because of the histological similarities with hemangioma and angiosarcoma. For this reason, immunohistochemical study is essential

to confirm the diagnosis by showing the positivity of endothelial cell markers such as factor VIII, CD31 and CD34 [7,14].

The evolution of EHE is unpredictable, ranging from the slow-growing form with no impact on the general state to the darker form rapidly progressing to hepatocellular failure. Management is adapted to each individual case and is based on radiological surveillance, surgical removal of lesions, chemotherapy or liver transplantation [1,6,11]. The overall five-year survival of patients with EHE is estimated to be 28-30% depending on the series [3,4].

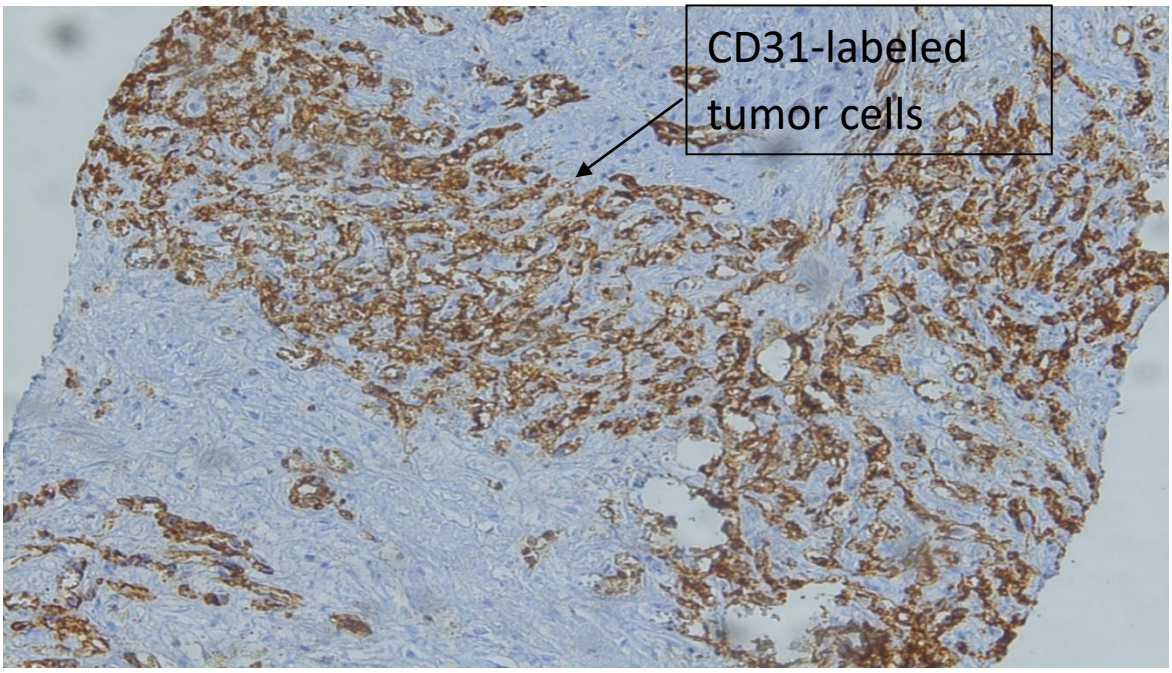

Fig. 6. The tumor cells express CD31

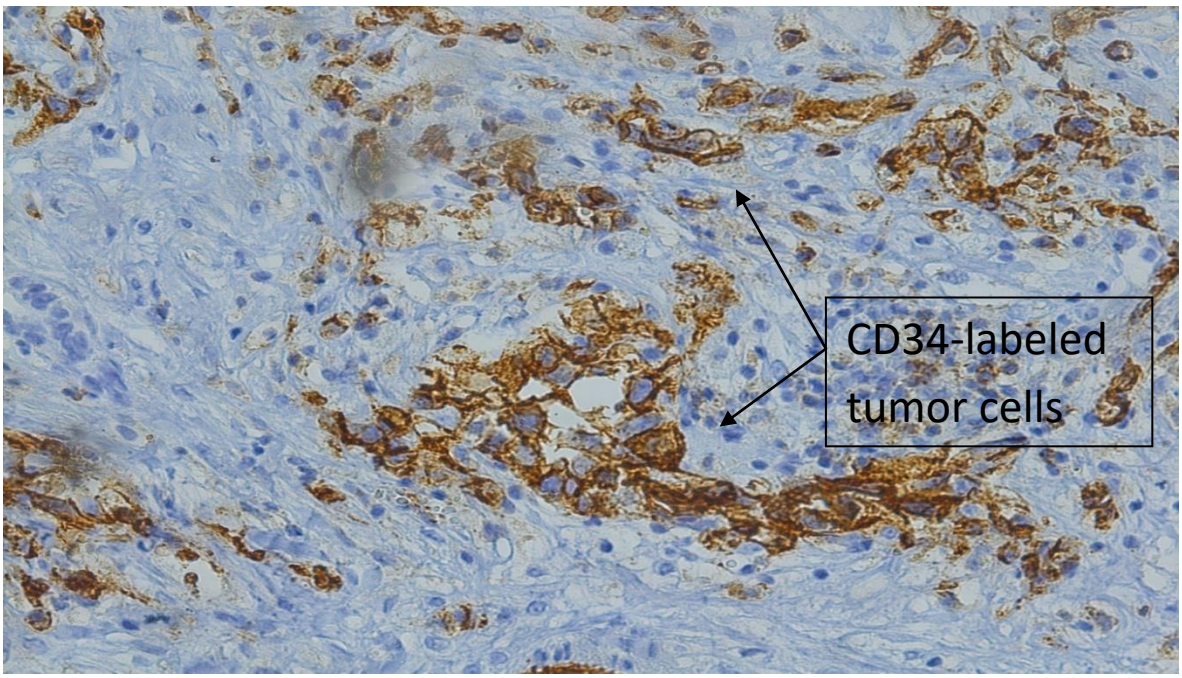

Fig. 7. The tumor cells express CD34

4. CONCLUSION

Epithelioid hemangioendothelioma of the liver is an orphan disease. Primary hepatic localization is rare. Its diagnosis remains difficult, requiring the use of an immunohistochemical study. The prognosis is variable and may be poor in some cases. The therapeutic management is adapted according to the presentation of the hepatic involvement, the symptomatology of the patient and the rapidity of the evolution.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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