



A Rapid Recurrence of a Hepatic Epithelioid Hemangioendothelioma after Liver Transplantation

Sahir Omrani, Wael Ferjaoui*, Mohamed Hajri, Ghofrane Talbi, Karouia S, Gharbi L, Bayar R and Khalfallah MT

Department of General Surgery, Mongi Slim Hospital, Tunisia

Abstract

The epithelioid hemangioendothelioma is a rare malignant vascular tumor that may occur within the liver. Liver transplantation is indicated in the hepatic multifocal and bilobar forms. In this article, we report the case of a 63-year old man who was diagnosed with Hepatic Epithelioid Hemangioendothelioma (HEHE) and treated by liver transplantation. However, the patient presented a rapid recurrence within 3 months.

Keywords: Liver; Recurrence; Hepatic epithelioid hemangioendothelioma; Transplantation

Introduction

HEHE is a rare vascular neoplasm of intermediate malignant potential [1]. It occurs in lung, spleen, brain, stomach and mostly in liver. Its incidence is very low. That is why there is no consensual therapeutic approach. Indeed, therapeutic options may include chemo or radiotherapy but also surgery. Liver transplantation can be indicated for unresectable cases with acceptable disease free survival [2]. However, some cases of recurrent hepatic HEHE after liver transplantation have been reported in the literature. In this issue, we present a case of rapid recurrent HEHE in a 63-year-old man who underwent orthotopic liver transplantation.

Case Presentation

A 63-year-old man with history of insulin-dependent diabetes was admitted with a 2-months history of right upper quadrant pain and 10 kg weight loss. On admission, physical examination was normal except tenderness in the right upper quadrant abdominal and hepatomegaly. Blood investigations revealed normal liver function. Abdominal ultrasound exam showed various round lesions all along the liver. An abdominal computed tomography scan was done and showed multiples lesions with low density and involving all the liver. These lesions are suggestive of liver metastases (Figure 1).

Fibroscope of the stomach and colonoscopy were performed in search of the primary tumor. They were normal. Percutaneous liver biopsy was, therefore, done and the histology and immunohistochemistry concluded to HEHE. The diffusion of lesions classified this tumor as unresectable. The patient was transplanted with liver graft from cadaveric donor. A cross section of the explanted liver revealed poorly limited white area inside the liver parenchyma (Figure 2). Histologic examination showed wide infiltration by epithelioid cells that express endothelial markers as CD31 and CD34. These results confirmed the diagnostic of HEHE.

The post-operative period was uneventful. He was maintained on Tacrolimus with no evidence of rejection. He presented 3 months later with abdominal pain and jaundice. Abdominal computed tomography showed multiple hepatic lesions with low attenuation (Figure 3).

Therefore, the biopsy was repeated and confirmed the recurrence of HEHE. He was started on palliative chemotherapy.

Discussion

HEHE is a rare tumor of vascular origin individualized in 1982 by Weiss and Enziger [3]. Its incidence is less than 1 in 1 million [4]. The epithelioid hemangioendothelioma occurs frequently in the liver, but can also touch other organs such as lungs, bones, brain, heart or retroperitoneum [3]. It can be asymptomatic. However, the patient may present, as in our case, nonspecific pain in the right upper quadrant of the abdomen. Other symptoms as jaundice and hepatosplenomegaly may be

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*Correspondence:

Wael Ferjaoui, Department of General Surgery, Mongi Slim Hospital, Tunisia, Tel: +216-52430099;

E-mail: farjaouiwael4@gmail.com

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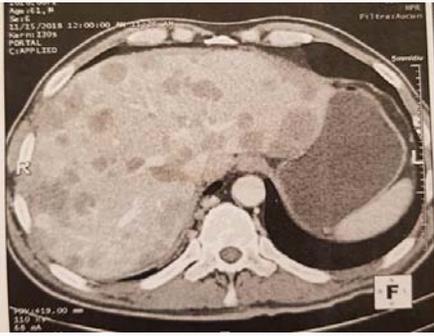


Figure 1: Abdomen computed tomography showing multifocal and bilobar lesions of HEHE.



Figure 3: Abdomen computed tomography after liver transplantation showing recurrence of HEHE.



Figure 2: Hepatectomy specimen showing a white mass with irregular margins within hepatic parenchyma.

found. In biology, there is no specific marker. Radiologically, HEHE appears as hypodense multifocal nodules on computed tomography. Differential diagnosis is that of liver metastases.

Microscopically, the lesions have many similarities with hepatic hemangioma or angiosarcoma with a proliferation of neoplastic cells of appearance epithelioid, most often along sinusoids. Endothelial specific markers such as CD34, CD31, vimentin and factor VIII-related antigen are often present [3]. Evolution without treatment is unpredictable and life expectancy can be long. Both therapeutic possibilities are hepatectomy and liver transplantation. Unique lesion can be treated by radical resection, with regular monitoring in order to ensure there is no recurrence in the remaining liver parenchyma. However, lesions may be multifocal and bilobar, as in our patient case. Liver transplant is, therefore, indicated [5]. Liver may be from cadaveric donor or, rarely, from living donor (only 6 cases from living donor liver transplantation for HEHE are reported in the literature [6]. Recurrence of HEHE after transplantation is possible and some cases were reported in the literature. Several factors are associated with this recurrence, such as the large tumor burden [6]. Indeed, Among 110 patients undergoing liver transplantation for HEHE between 1987 and 2005, The United Network for Organ Sharing reported 12 patients who died of recurrent HEHE within 5 years.

The average time of recurrence is 49 months. The longest recurrence reported is up to 12 years after Liver transplantation [6] and the shortest recurrence is 3 months [6]. Our case, therefore, is among the shortest recurrence of HEHE. There is no consensual treatment for recurrent HEHE. Palliative chemotherapy with agents targeted against vascular endothelial growth factor such as thalidomide is reported in the literature.

Conclusion

Recurrence of HEHE after liver transplantation is exceptional and unpredictable. All cases of recurrence should be reported and published in order to establish risk factors as well as the suitable management of these recurrent cases.

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