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Mesenchymal Hamartoma: Exceptional Liver Tumor in Adults

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ABSTRACT

Hepatic Mesenchymal Hamartoma (HMH) is a congenital benign tumor, with an uncertain pathogenesis. Most of them are diagnosed during childhood and they are exceptional in adults. We present a case of a young adult with a hepatic tumor in the segments IV-V, initially described as an adenoma and then, five years later definitely diagnosed with HMH. The tumor was completely resected with free margins because of its volume increase and risk of malignization. We study the clinical presentation until the final pathology. The analysis of its possible etiologies, is very important as well as the diagnosis value and surgical planning with images, especially the computed tomography (CT Scan) and magnetic resonance imaging (MRI) in a tumor in which a mesohepatectomy of the segments IV-VIII was performed and its result allows us to suggest its indication in central benign tumors.
Key words: liver tumors, hamartoma mesenchyimal, benign tumors, liver surgey

INTRODUCTION

Liver tumors in young adults have a low incidence and most of them are blastomas. Among them there is the Mesenchymal Hepatic Hamartoma (HMH). It is a benign tumor, congenital, with an unknown pathogenesis. Most of them are diagnosed during childhood and they represent approximately 8% of all pediatric tumors. They are rare in adults and there are only 15 cases published in the English literature (1,2). This report analyzes a clinical case which has three aspects of interest: the previous biopsic diagnosis of liver adenoma at 14yo, the definitive diagnosis of HMH after surgery 5 years later, and the complete resection of the tumor with atypical mesohepatectomy.

CASE REPORT

Young-man of 19yo. with clinical record of right inferior lobectomy and medial right one secondary to a Pleuropulmonary Blastoma followed of adjuvant therapy when he was two years old. When 14yo. during an oncologic control a hepatic nodule of 33 mm in segment IV. The biopsy diagnosis was hepatic adenoma.

He continued with clinical and image follow up. A CT scan and MRI made at 19yo showed a liver nodule of 59 x 42 mm (*fig. 1*). Surgery was indicated

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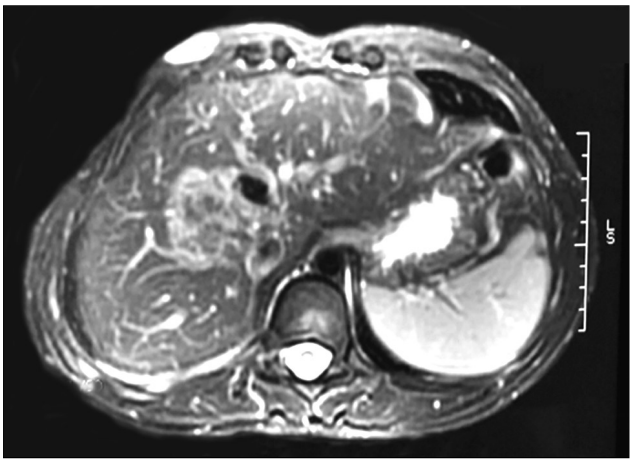

Figure 1 - MRI that shows the relation of the tumor with the inferior vena Cava and the suprahepatic veins

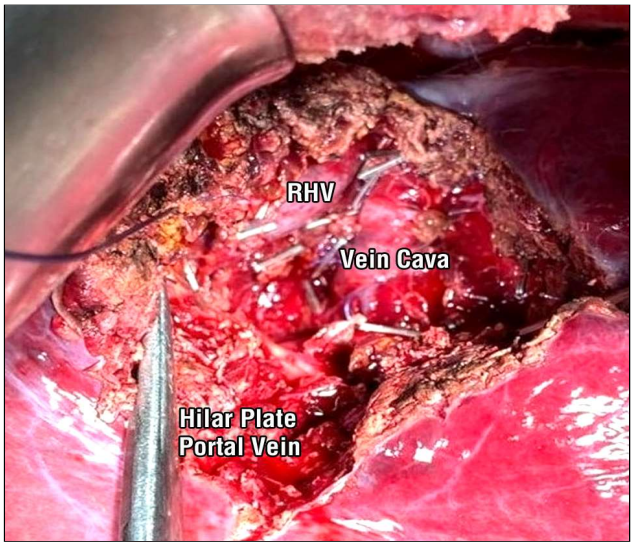

Figure 2 - Intraoperative view of the liver after the surgical resection. In the liver bed we can see the right suprahepatic vein (RHV), the inferior vena Cava and the Hilar plate

because of the risk of malignant transformation of the adenoma which reached the size of 6 cm in diameter.

Tumoral markers such as CEA and CA 19.9 were normal.

Surgical approach was performed by bilateral subcostal type Mercedes Benz´s incision.

After mobilization of the liver, in intraoperative ultrasound we could observe once lesion in segments IV and VIII between the right and medial suprahepatic veins (*fig. 2*). A central hepatectomy was performed (segments IV and VIII) with macroscopic free margins. The operation was completed with a cholecystectomy.

Postoperative evolution without complicantions. Hospital discharge: 4 days.

The pathologic study of the piece informed: macroscopy: hepatic tumor of 65 x 51 x 42 mm, weight of 51 grams and multinodular surface with off-white and brownish areas, alternating with yellowish sectors (*fig. 3*).

Histology: in the liver´s parenchyma we can see epithelial and stromal components and hepatocytes without nuclear pleomorfism or mitosis, biliary ducts are surrounded by abundant fibrotic stroma of the sclerotic type with isolated mixoyd sectors with fusiform cells and inflammatory cells as well. Histopathologic conclusion: Mesenchymal Liver Hamartoma (*fig. 4*). Follow up: 2 years. Asymptomatic and with normal quality of life.

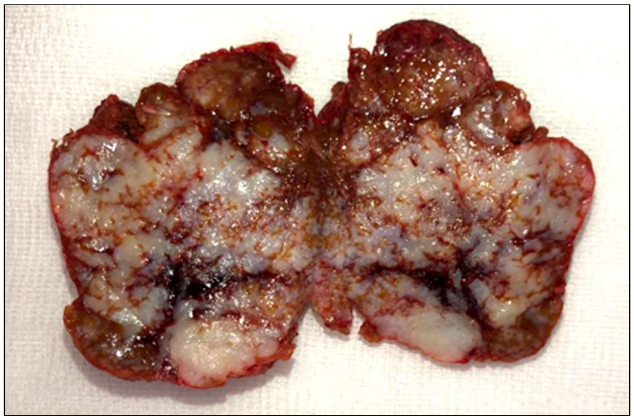

Figure 3 - Macroscopic view of the specimen open in the middle. We can observe the cystic solid component of the tumor with a multinodular aspect with off-white and brownish areas

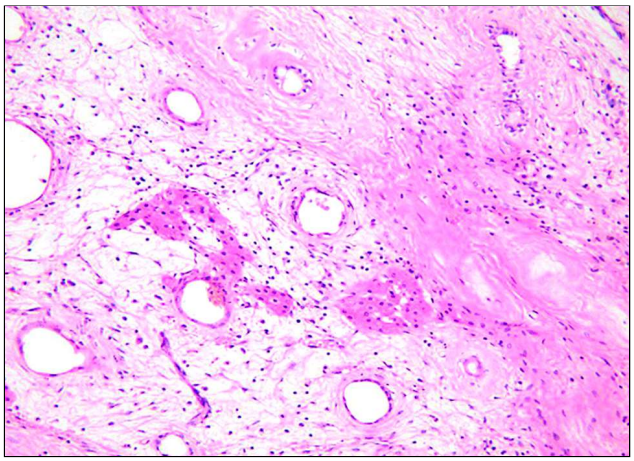

Figure 4 - Hepatic microscopy view (H&E staining). It can see the histologic pattern of HMH with a lesion integrated by epithelial and stromal components and hepatocytes´s grouped without nuclear pleomorfism or mitosis. The biliary ducts are surrounded by a fibrotic stroma of the sclerotic type, with isolated mixoyd sectors with fusiform cells and inflammatory cells

DISCUSSION

The HMH is a benign primary tumor almost exclusive of childhood although cases in young adults

have been reported (3).

It was described in 1959 by Edmoston. It represents an 8% of pediatric tumors and the second most common of all benign tumors (4,5,6).

Other histopathologic patterns associated to this entity include lymphangioma, hamartomas of the biliary tract, mesenchymomas, pseudo cystic mesenchymal tumors and cystic hamartomas.

It is a congenital pathology and its histogenesis is uncertain. It is considered a growth anomaly, composed by multiple cysts with internal septa (7). It is made of unorganized biliary ducts, immature mesenchymal tissue full of liquid and hepatocytes. This lesion's probable origin are alterations of the connective tissue of the Porta tract or as a result of a regional ischemic process , rather than a true neoplasm (4,7,8). It has also been postulated that its origin is the balanced translocation between the two long arms of chromosomes 11 and 19. Likewise, it has been demonstrated that a rupture point in chromosome 19, 19q13.4, is a carrier of a karyotypic anomaly. Therefore, with this data we can support the hypothesis that states that maybe a subgroup of the HMH has a neoplastic origin (6). This structural association with chromosome 19 allows to associate the HMH with the embryonic sarcoma.

Most of the HMH remain asymptomatic for a long time. Seventy seven per cent of them affect the right liver lobe and the rest the left lobe or both (9). In big and evolved tumors the clinical manifestation are: a tumor in the right superior quadrant, shortness of breath, fever, and an elevated right hemi diaphragm (9,10).

In our case the diagnosis was late because the initial diagnosis was adenoma at 14yo. Only with a tumor of 6 cm after surgical resection we could stablish the definitive diagnosis of HMH.

Usually, the diagnosis of HMH is made by image studies and confirmed with biopsy. The hepatic function is normal and it does not influence the diagnosis. The ultrasound shows a hepatic image well delimited, round, heterogeneous, due to its solid and cystic components. It is made of cysts of variable sizes, with thin walls septa mixed with solid parts which appears poorly or not vascularized at all when studied with Doppler-US. The most accurate preoperative diagnosis is achieved with cytology with fine needle aspiration in order to stablish differences with hepatoblastoma, pediatric hepatocellular carcinoma, hemangioendothelioma, undifferentiated sarcoma, smooth muscle tumors and inflammatory pseudo tumor (11,12).

The abdomen CT scan and the MRI are usually used to clarify vascular structures and therefore to help planning the surgical resection. In this case the MRI (*fig. 1*) showed a hypo intense imagen in T.1 and a hyper intense one in T.2; heterogeneous without bleeding and fatty areas; with mild arterial enhancement, especially peripheral in a late phase. It does not have an image pattern which suggests adenoma or any other frequent benign nodular lesions such as hemangioma and focal nodular hyperplasia. In the rest of the liver, other small nodules are associated with adenomas.

HMH is a benign tumor and most authors recommend total resection of the tumor. They consider it enough to treat hamartoma. In case of partial incomplete resection recurrent benign lesions may appear (2).

Central hepatic tumors that affect the segments IV-V-VIII and I must be removed by a mesohepatectomy, which indication for benign tumors is only about 11.7 % (13). We consider this technique as an alternative to extended hepatectomy because it offers up to 5% loss of normal parenchyma, less than the other, although the latter may be limited by the risk of small liver syndrome or post-resection liver failure (14,15).

In the present case a partial mesohepatectomy was performed (*fig. 2*) affecting segments IV and VIII including the medial suprahepatic vein, without the most common post-operative complications such as bile leak, bleeding, and hepatic insufficiency (13) Regarding all this information, a recent meta-analysis has demonstrated that mesohepatectomy is an attractive procedure due to the advantages of keeping hepatic parenchyma, with less bleeding and less surgical time comparing with extended hepatectomies (13).

CONCLUSION

We have seen a rare liver congenital tumor in a young adult, of the segments V and VIII; with initially diagnosis of adenoma and five years later with definitive diagnosis of HMH. Its histogenesis is complex and uncertain. It supposedly originates from connective tissue alterations of the Porta tracts as a result of a regional ischemia; while other hypothesis proposes a chromosomic alteration, which leads to think of a neoplastic etiology.

Diagnosis was made with image studies which show a well delimited tumoral pattern, with heterogenic structure due to its solid and cystic components with poorly vascularized septa. A biopsy was not necessary to rule out other histopathologic patterns. The resection was indicated because of the increased size of the tumor. Even though HMH is a benign tumor most authors recommend a complete resection of it. A

partial mesohepatectomy was performed and its effectiveness allows us to suggest this surgical indication in central benign tumors.

Conflicts of interest

No conflicts of interest.

Ethical approval

The procedures were adjusted to the norms of the ethics committee of the Maciel Hospital, of the Human Rights Committee of Helsinki. The signed consent of the patient was obtained.

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