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Int J Case Rep Images 2020;11:101113Z01SA2020.
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Hepatic angiomyolipoma: A case report and review of the literature

Sanae Amalik, Othman Ayouche, El Amrani, Fatima Zahra, Laila Jroundi

ABSTRACT

Introduction: Angiomyolipomas are benign tumors. They can be asymptomatic or cause various types of atypical abdominal pain, which are uncommon consequences of this pathologic growth. The advent of new radiologic modalities has allowed for early diagnostics and treatment of these entities. **Case Report:** We report the case of a 60-year-old female, with no history of chronic disease, who consulted of right upper abdominal quadrant pain. We conducted a Doppler ultrasound and a contrast enhanced computed tomography (CT), which demonstrated a double component fatty hepatic lesion with a dysplastic vascular network. The choice of a therapeutic abstention with sonographic surveillance every three months was made, with a further possibility of embolization and surgical resection. **Conclusion:** Early diagnosis followed by surgical resection is the mainstay of management of large hepatic masses. Computed tomography angiography is the modality of choice for the detection and evaluation of hepatic angiomyolipomas. Magnetic resonance imaging (MRI) and liver biopsies are recommended for difficult cases with no fatty components. In our case, we made the choice of abstention with sonographic surveillance.

Keywords: Angiomyolipoma, Benign, Computed tomography, Hepatic, Solid mass

Sanae Amalik[1], Othman Ayouche[1], El Amrani[1], Fatima Zahra[1], Laila Jroundi[1]

Affiliation: [1]Emergency Radiology Department, Ibn Sina Hospital, Mohamed V University, Rabat, Morocco.

Corresponding Author: Sanae Amalik, Emergency Radiology Department, Ibn Sina Hospital, Mohamed V University, Rabat, Morocco; Email: sanaeamalik@gmail.com

Received: 07 January 2020
Accepted: 12 March 2020
Published: 06 April 2020

How to cite this article

Amalik S, Ayouche O, Amrani E, Zahra F, Jroundi L. Hepatic angiomyolipoma: A case report and review of the literature. Int J Case Rep Images 2020;11:101113Z01SA2020.

Article ID: 101113Z01SA2020

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doi: 10.5348/101113Z01SA2020CR

INTRODUCTION

Angiomyolipomas are rare mesenchymal benign tumors with a female predominance [1]. Right hepatic localization is classic. The size is extremely variable ranging from 1 cm to several tens of cm, occurring most commonly in renal parenchyma with a few descriptions of hepatic cases. The association of a tuberous sclerosis of Bourneville, commonly admitted for renal localization, is rare for hepatic angiomyolipoma. Its diagnosis is difficult, and there is no codified treatment [2]. Complications are exceptional which consists of spontaneous ruptures with unknown physiopathological mechanisms [3].

CASE REPORT

We report the case of a 60-year-old woman, admitted for the exploration of a right upper abdominal quadrant pain for the past five months treated with pain medication, in a context of conserved general state.

The clinical examination found an isolated right upper quadrant tenderness. The biological assessment that consisted of the hepatic transaminases and blood serum numeration was normal. After which, a sonography exploration was conducted concluding for the presence of a solid heterogeneous hepatic mass of the right hepatic lobe measuring 5 cm.

Abdominal CT revealed the presence of a heterogeneous solid hepatic mass of the sixth segment, oval, well-defined, irregularly contorted, containing a double component: tissue enhanced early after injection, as well as a predominant fat component, enhancing in the early arterial phase persistent in the equilibrium phase with early venous filling of drainage vein (Figure 1), measuring 59 × 47 × 71 (AP × T × H). It is characterized by a significant dysplastic vascular network arising from two branches of the right hepatic artery (Figure 2). The diagnosis of angiomyolipoma was strongly suspected.

In view of the importance of clinical symptomatology, a biopsy with histological examination was performed and confirmed the diagnosis of hepatic angiomyolipoma.

Gross examination of mass shared a brownish encapsulated mass measuring 5 cm. Immunohistochemistry has shown positively labeled cells with Melan A and MB 45 antibodies. The choice of an abstention with sonographic surveillance every three months was made, with a further possibility of embolization and surgical resection.

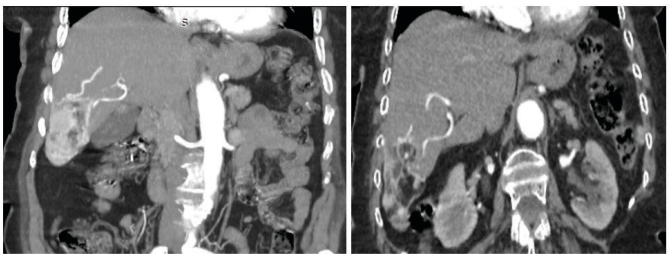

Figure 1: Coronal reformation of an abdominal CT in arterial phase: dysplastic vascular network arising from the hepatic artery.

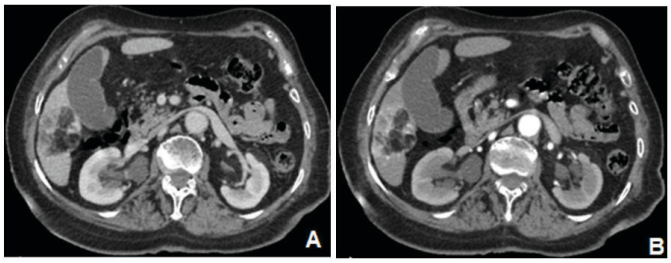

Figure 2: Axial contrast-enhanced abdominal CT. (A) Heterogeneous fatty mass with a homogenization in portal phase. (B) Tissular component with early enhancement in arterial phase.

DISCUSSION

Angiomyolipoma is an exceptional benign mesenchymal tumor of the liver with a potential for malignancy transformation. It can be classified into four histological types according to tissue differentiation into: mixed, fat, muscle, and vascular, with no specific imaging representation. Smooth muscle is the most specific cell component for histological diagnosis, for which HMB 45 (marker for smooth muscle cells) is positive. Hepatic localization is rare since fewer cases have been described in the literature.

Due to the variable proportion of the three tissue components, hepatic angiomyolipoma can take several aspects in imaging.

The presence of fat, if detected in imaging, is suggestive of the diagnosis; if not, the diagnosis is more difficult and the tumor may be mistaken for hepatocellular carcinoma or other benign liver injury [4]. There is a clear predominance of women with an average age close to 50 years [1]. The association with tuberous sclerosis of Bourneville is rare, unlike renal angiomyolipoma [5].

In addition, the reported size varies between 1 and 25 cm with a mean of 6.1 mm with a location in the right hepatic lobe in most cases [2]. Only one recurrence was reported six years after the removal of the initial tumor [2].

Hepatic angiomyolipoma is an asymptomatic tumor, which explains why its discovery is mostly fortuitous. However, in the case of a large tumor, it can be a source of abdominal pain. The diagnosis is made essentially by imaging. The most commonly reported ultrasound appearance is that of a single heterogeneous echogenic mass of well-limited variable size with posterior reinforcement (fibrous component) [1].

In CT scans, we often describe a hypodense heterogeneous mass with a fat quota lower than –20 HU with significant and early contrast enhancement of the vascular contingent [1].

In MRI, usually the presence of a fat component can help make the diagnosis with the discovery of a T1 hyper signal that saturates on FATSAT sequences. Angiomyolipomas typically have at least one T1 hyper signal portion. The fatty portion of the angiomyolipoma is vascularized and intensifies with early onset, whereas the fatty portion of hepatocellular carcinoma is relatively avascular and therefore not elevated after injection of contrast [1, 6].

However, in its absence there is a low signal on T1 sequences. An angiomyolipoma low in fat is a challenge for the radiologist, since there are few arguments for the diagnosis. It is therefore advisable, just as its renal location, to search for small intratumoral fat islands (CT without injection and specific MRI sequences).

In the case of large intratumoral vessels, the presence of an early-filled hepatic drainage vein is also highly suggestive of the diagnosis [6, 7].

Angiomyolipoma is a well-limited, hyper vascular tumor at the arterial phase with persistent contrast enhancement at venous and equilibrium phases [6, 7]. The peak-time enhancement is later than that of hepatocellular carcinoma, but this characteristic is difficult to assess in daily practice.

The absence of peripheral capsule is also a sign reported in several series for the diagnosis of hepatic angiomyolipoma, especially in the presence of a hyper vascular tumors, even low in fat, in a non-cirrhotic liver. Early and persistent contrast enhancement may suggest

an inflammatory adenoma, but there is no capsule or circumferential pseudocapsule in angiomyolipomas, whereas this sign may be visible in inflammatory adenomas under the name of the reversed halo sign [6, 7].

In doubtful cases, the performance of a liver biopsy makes the diagnosis easier. The standard treatment for hepatic angiomyolipoma is surgical resection.

Therapeutic abstention may be discussed if the tumor is less than 5 cm and the diagnosis has been confirmed histologically after liver biopsy, as the malignant transformation risk is exceptional [2–4]. We proposed for our cases an abstention with a sonographic surveillance every three months for the first year.

CONCLUSION

Hepatic angiomyolipomas are rare mesenchymal benign tumors. The advent of new noninvasive diagnostic imaging techniques with increased sensitivity for asymptomatic disease has led to more frequent diagnostics. Although surgery resection is recommended for large masses, abstention is the norm for asymptomatic tumors less than 5 cm. In our case, we made the choice of abstention accompanied with a sonographic surveillance.

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Author Contributions

Sanae Amalik – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Othman Ayouche – Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

El Amrani – Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Fatima Zahra – Design of the work, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Laila Jroundi – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.