Radiology Case Reports 13 (2018) 829–833

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Hepatic epithelioid angiomyolipoma with renal metastasis: radiologic-pathologic correlation

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ARTICLE INFO

Article history:
Received 8 April 2018
Revised 10 May 2018
Accepted 13 May 2018

Keywords:
Epithelioid angiomyolipoma
Liver PET

ABSTRACT

Epithelioid angiomyolipoma (EAML) is a rare subtype of angiomyolipomas. Unlike the conventional angiomyolipomas, EAML often contains minimal fat which usually precludes prospective diagnosis on imaging. The imaging findings of EAML may overlap with other benign and malignant hepatic neoplasms. We report a hepatic epithelioid angiomyolipoma in a 47-year-old female which metastasized to the right kidney and recurred after resection in the liver. We analyze the imaging findings of EAML on ultrasound, computed tomography, positron emission tomography and magnetic resonance imaging. Correlation between the imaging and histopathologic findings is made. The estimated annual growth and doubling time of the primary hepatic EAML are calculated. To the best of our knowledge, this is the first published report of positron emission tomography–computed tomography findings and annual growth rate for hepatic EAML.

Introduction

Angiomyolipoma (AML) is a mesenchymal neoplasm composed of adipose tissue, smooth muscle, and abnormal blood vessels in varying proportions. Epithelioid angiomyolipoma (EAML) is a rare subset composed predominantly of perivascular epithelioid cells and often contains minimal fat [1–3]. The tumor occurs most frequently in the kidneys but has also originated in either lobe of the liver. Although often benign, hepatic EAML has demonstrated rare malignant behavior and recurrence following resections [4,5], as well as several recorded instances of metastasis [2,5,6,7]. This paper will discuss a case of a hepatic EAML with metastasis to the right kidney six years following its initial discovery. Imaging features from ultrasound (US), magnetic resonance imaging (MRI), computed tomography (CT) and positron emission tomography (PET) will be analyzed. Similarities and differences on imaging to other hepatic lesions would also be discussed. The annual growth rate of the hepatic EAML will also be estimated.

Case report

A 47-year-old female, nonsmoker with no prior surgical history apart from 2 C-sections, but has a strong family history of lung cancer, colon cancer, and other malignancies was found

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https://doi.org/10.1016/j.radcr.2018.05.007

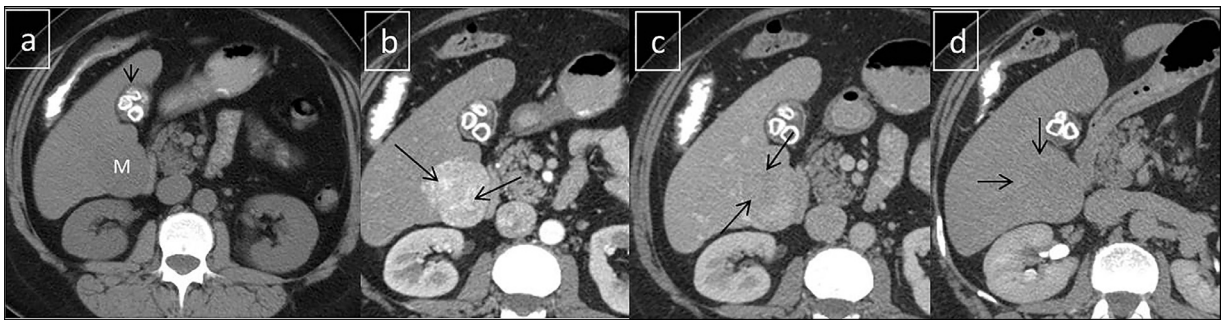

Fig. 1 – First computed tomography examination during the initial presentation (patient age 47 years): the primary lesion in the right hepatic lobe which is slightly hypoattenuating to the remainder of the liver parenchyma (M) on the nonenhanced computed tomography (a). It demonstrates heterogeneous avid enhancement on the arterial phase (b). Note the linear intralesional structures which represent prominent vessels centrally in the lesion and at the periphery (arrows in b). On the portal venous phase, the mass demonstrates mild washout, best seen in the right lateral part (arrows in c). Pseudocapsule is seen on the delayed phase (arrows in d) while the mass demonstrates mild washout (d). Incidental note is made of multiple hyperdense gallbladder stones (short arrow in a).

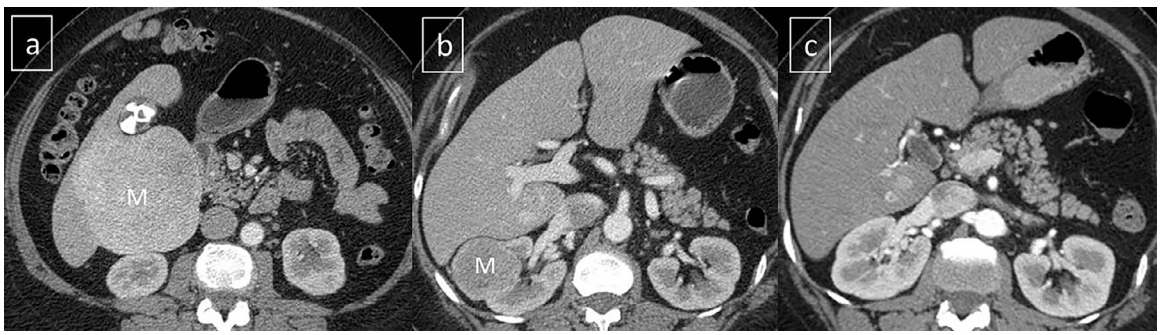

Fig. 2 – The follow-up portal venous phase computed tomography performed 6 years and 4 months after the initial presentation (patient age 53 years) (a and b); showing significant interval growth of the right hepatic lobe mass (M in a) and a new development of an interpolar partially exophytic right renal mass (M in b). The previous arterial phase computed tomography during the initial presentation at the same level of (b), is provided for comparison (c).

to have a solitary right lobe hepatic mass in outside abdominal ultrasound. The mass measured approximately 4.2 cm in longest dimension. The patient underwent a CT abdomen and pelvis with and without IV contrast according to the triphasic liver CT protocol for further evaluation of the hepatic mass. A well-defined hyperdense lesion was seen in segment VI of the liver with estimated measurements of $3.8 \times 4.6 \times 4.7$ cm (Fig. 1). It appeared solid with extension to the liver capsule and partially exophytic. There was avid arterial enhancement which filled in, appearing almost homogenous, on delayed imaging. Two additional hepatic lesions were present, one adjacent to the dominant growth and one anteriorly, measuring 6 mm and 5 mm respectively. These were thought to represent hepatic hemangiomata.

A subsequent CT was performed 6 year and 4 months after the initial presentation for evaluation of vague upper abdominal pain. The scan showed that the hepatic lesion had grown in size to an estimated $10.9 \times 9.7 \times 11.2$ cm (Fig. 2). The mass demonstrated arterial enhancement and faint washout in some areas. There was no significant necrosis, central scar, intralesional fat, or calcification. The adjacent lesion had grown to 1.9 cm and the anterior lesion had grown to 2.4 cm. Incidentally, the scan revealed that a new solid, homogenous, exophytic mass had developed in the interpolar region of the right kidney (Fig. 2), measuring 4.6×5.1 cm. The mass was slightly hyperdense on the unenhanced images (34 Hounsfield units). On the contrast enhanced images, it measured up to 77 HU in the arterial phase with washout to 57 HU in the portal venous phase. This raised the possibility of an indolent malignancy and subsequently US-guided biopsies were conducted on the hepatic and renal masses after 3 months and 7 months of the later CT examination, respectively (Fig. 3). Pathologic analysis of the hepatic biopsies showed a tumor consisting of epithelioid-like cells arranged in clusters with thin interweaving vascular channels and a few spindle cells. Adipose tissue was present in one of the cores, as well as areas of inflamed fibrous macrophages, evidence of remote hemorrhage, and admixed hematopoietic cells. The sample was positive for HMB-45 and Melan A. Similar features were noted in the core samples from the renal lesion. However,