

乏脂肪型肝脏血管平滑肌脂肪瘤 CT 和 MRI 表现分析及鉴别诊断文献复习

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摘要: [目的] 探讨影像学上表现为乏脂肪型的肝脏血管平滑肌脂肪瘤(hepatic angiomyolipoma, HAML) 的 CT 和 MRI 表现并复习文献, 旨在提高对该病的认识和诊断准确性。[方法] 回顾性分析经穿刺或者手术病理证实的 6 例乏脂肪型 HAML 的 CT 和 MRI 表现及临床资料。[结果] 6 例 HAML 均为女性, 4 例位于肝右叶, 2 例位于肝左叶。6 例 HAML 均单发, 边界清楚。CT 平扫呈稍低密度, 密度均匀, 平扫均未见条状及裂隙状脂肪密度影, 增强扫描动脉期肿瘤呈均匀明显强化, 门脉期强化明显减退; MRI 平扫 T1WI 呈稍低信号, T2WI 呈稍高信号, 正反相位未见明显信号改变, DWI 及 ADC 均呈高信号。增强扫描动脉期肿瘤均匀明显强化, 门脉期强化明显减退。6 例增强模式均为“快进快出”型。6 例病灶均可见粗大强化的血管穿行于病灶或者在病灶周围。6 例病灶内均未见坏死、囊变、钙化及假包膜。[结论] HAML 的影像学表现具有一定的特征性, 了解这些影像特征能够提高该术前诊断的正确率。

关键词: 肝脏血管平滑肌脂肪瘤; CT; MRI; 鉴别诊断

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Analyzing Imaging Features of Hepatic Angiomyolipoma Devoid of Fat for Diagnosis and Differential Diagnosis: A Retrospective Study with Literature Review

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Abstract: [Objective] To evaluate the imaging features of hepatic angiomyolipoma(HAML) devoid of fat so as to improve the understanding and diagnostic accuracy of this disease. [Methods] The clinical data, including computed tomography (CT) and magnetic resonance imaging(MRI) records, of 6 patients with pathologically confirmed HAML devoid of fat were reviewed and analyzed with literature review. [Results] Of the 6 patients, all were female; 4 had lesions located in the right lobe of liver, and 2 had lesions located in the left lobe of liver. All patients had solitary lesions with well-defined boundaries. Non-enhanced CT images showed lesions with slightly low density. There was no limited spot- or arc-like low density in the center or on the edge of lesions with CT values of -30 to -10 HU. Two cases showed slightly low-signal intensity on T1WI and slightly high-signal intensity on T2WI. There were no obvious signal changes on in-phase images, and low-signal intensity was observed on out-phase images. Both DWI and ADC showed high-signal intensity. Contrast-enhanced CT and MRI showed signal enhancement with “fast in and slow out” fashion in the arterial phase, obvious signal decrease in the portal phase, and signals of blood vessels crossing or surrounding lesions in all cases. No necrosis, cystic change, calcification, and pseudocapsule were found in 6 cases. [Conclusion] HAML devoid of fat has certain characteristic imaging manifestations, which can improve the accuracy of preoperative diagnosis of the disease.

Subject words: hepatic angiomyolipoma; computed tomography; magnetic resonance imaging; differential diagnosis

肝脏血管平滑肌脂肪瘤 (hepatic angiomyolipoma, HAML) 是一种罕见的间叶源性肿瘤^[1], 具有丰富的脂肪成分时基本容易诊断。然而, 部分肝脏血管平滑肌脂肪瘤影像学上未见明显脂肪成分, 易与肝脏其他病变相混淆。本文回顾性分析经病理证实的

6 例乏脂肪型 HAML 患者的影像学表现, 总结其特征并复习文献, 旨在提高对该病的认识及鉴别诊断能力。

1 资料与方法

1.1 一般资料

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医院经穿刺或者手术病理证实,并经影像学观察无明显脂肪密度的乏脂肪型 HAML 患者资料共 6 例,均为女性;年龄 42~68 岁,中位年龄 55 岁。3 例体检发现肝脏占位性病变,无任何临床症状。1 例因子宫肌瘤术前检查发现,1 例因肺癌术前检查发现。实验室检查 AFP、CA199、CEA 均阴性,无乙肝及肝硬化病史(Table 1)。其中 4 例行 CT 平扫和动脉期、门脉期增强扫描,1 例行 MRI 平扫和动脉期、门脉期增强扫描,1 例行 CT 及 MRI 平扫加增强扫描。

1.2 检查方法

采用西门子 64 排 CT 机进行 CT 检查。扫描参数为 120kV, 300mA, 层厚 5mm, 层间距 5mm。应用高压注射器经肘静脉注射对比剂碘普胺(含碘 300mg/ml), 注射速率为 2.5ml/s。CT 增强扫描动脉期、门脉期分别选择在注射造影剂后 30s 和 60~70s。

采用西门子 3.0T MRI 机进行 MRI 检查。采用相控阵头线圈。扫描序列包括:①横断面 FSE T1WI: TR 400~500ms, TE 10~20ms; ②横断面脂肪抑制 FSE T2WI: TR 4000~4600ms, TE 90~100ms。矩阵 380×224, 激励次数 2 次, FOV 19cm×23cm, 层厚 4mm, 层间距 1mm。增强扫描对比剂采用 Gd-DTPA, 经肘静脉以 2.0ml/s 流率注射 0.2mmol/kg, 随后注入生理盐水 20ml。增强后行横断面、矢状面和冠状面 T1WI, 参数同平扫。

1.3 影像学评价

由 2 名中级以上职称的放射科医师共同阅片, 达成一致意见。观察内容包括病灶部位、大小、形态、边缘、密度或信号、钙化、强化方式、淋巴结转移等。增强 CT 扫描强化程度分级标准: 轻度强化: 病灶实性部分强化较平扫增加 10~20HU; 中度强化: 病灶实性部分强化较平扫增加 21~40HU; 明显强化: 病灶实性部分强化较平扫增加 41HU 以上。MRI 强化程度标准: 轻度强化: 病灶实性部分强化程度与颈部

肌肉强化相近; 中度强化: 强化程度高于肌肉低于血管; 明显强化: 病灶强化程度与血管强化相近或高于血管。

1.4 病理学检查

标本均常规固定、包埋, 连续切片后分别行 HE 染色及免疫组织化学染色。免疫组织化学检测抗体包括 S-100、HMB45、Melan-A、SMA、Ki67、Hepahepatocyte、gly-3、glypican-3、ALK 等。由 2 名病理科副主任医师回顾性分析所有病理切片, 最后得出结论。

2 结 果

2.1 影像学表现

6 例乏脂肪型 HAML 中肿瘤位于肝右叶 4 例(肝 V 段 2 例和 VI 段 2 例), 肝左叶 2 例(肝 II 段 1 例和 III 段 1 例)。6 例均表现为类圆形、边界清、浅分叶状单发肿物。肿瘤长径为 1.0~4.5cm。4 例 CT 平扫均呈稍低密度, 密度均匀, 平扫未见大片状、条状及裂隙状脂肪密度影, 增强扫描动脉期明显强化, CT 值约 105~160HU, 强化净值约 50~110HU, 门脉期明显减退, 门脉期病灶与正常肝实质呈低密度、等密度或略高密度(Figure 1)。1 例 MRI 扫描 T1 信号为稍低信号, T2 为稍高信号, 正反相位未见明显信号改变, DWI 及 ADC 均呈高信号, 增强扫描动脉期呈明显强化, 门脉期明显减退。1 例行 CT 及 MRI 增强扫描(Figure 2)。6 例病灶中心或周围均可见粗大的血管, 均未见坏死、囊变、钙化及假包膜。

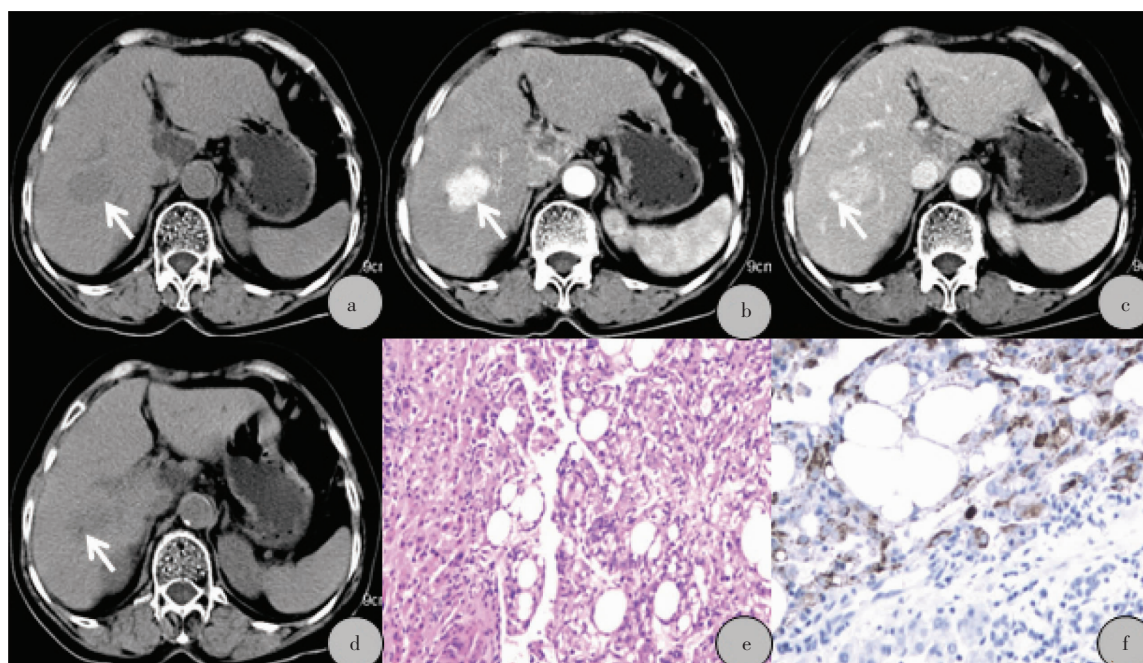
2.2 病理结果

大体观: 肿瘤多呈单发、类圆形、无包膜的肿块。镜下: 3 例为上皮样血管平滑肌脂肪瘤, 其脂肪细胞不明显, 主要由上皮样细胞组成; 3 例为血管平滑肌脂肪瘤, 其中 1 例可见明显簇样分化成熟的脂肪细胞, 2 例见散在脂肪细胞。瘤周肝组织均无肝硬化改

Table 1 Clinical characteristics of 6 cases of HAML

No.	Age (years)	Gender	HBsAg	Cirrhosis	Tumor location segment	Tumor size (cm)	Preoperative diagnosis	Pathological characteristics
1	42	F	Negative	No	III	4.5×3.5	HCC	(epithelioid)HMB45+, SMA+, Melan-A+
2	59	F	Negative	No	VI	1.0×0.9	Metastasis	(epithelioid)HMB45+, SMA+, CD34+, Vim+
3	43	F	Negative	No	II	2.8×2.5	HCC	(epithelioid)HMB45+, SMA+, CD31+, Vim+, Melan-A+
4	60	F	Negative	No	V	1.9×1.3	Hemangioma	HMB45+, SMA+, Melan-A+, CD31+, CD34+
5	51	F	Negative	No	VI	2.2×1.7	HCC	HMB45+, SMA+, Melan-A+, CD34+, CD31+
6	68	F	Negative	No	V	3.2×2.1	Hemangioma	HMB45+, SMA+, Melan-A+

Note: F; female; HBsAg; hepatitis B surface antigen; HCC; hepatocellular carcinoma.



Note: a: The lesion in the right anterior liver lobe was 3.2cm×2.1cm in size (arrow), which showed slightly low density and lobulated in shape in non-enhanced CT images.
b: Contrast-enhanced scanning showed that lesion was significantly enhanced in arterial phase, CT value was 160HU, there was no early enhanced pseudocapsule around the lesion.
c: The lesion decreased in the portal phase obviously, CT value was 109HU, which was slightly higher than liver parenchyma and the enhanced blood vessel was seen around the lesion.
d: The dots of low density was seen in the lesion (arrow), CT value was 23HU, fat component was considered by radiologist combined with postoperative pathology.
e: Tumor disorder showed predominance of smooth muscle cells and fat cells, with much vascular tissues (HE staining ×200)
f: Strongly positive staining for HMB-45 (immunohistochemical staining ×200).

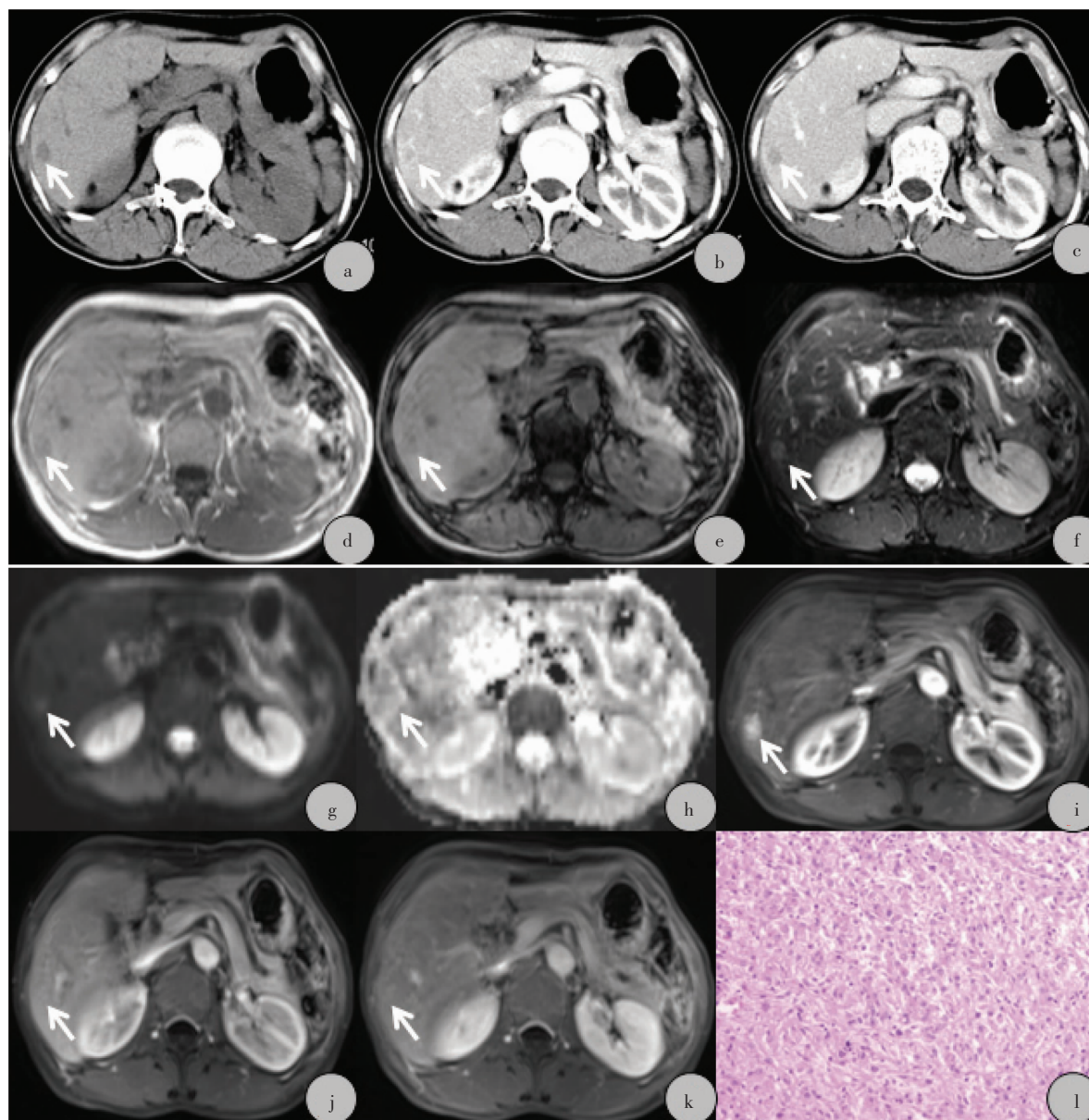
Figure 1 Pathological and CT features of a pathologically diagnosed HAML

变。6例均行免疫组化检测，结果显示 HMB45、SMA、Melan-A 均为阳性 (Table 1)。

3 讨论

肝脏血管平滑肌脂肪瘤分经典血管平滑肌脂肪瘤和上皮样血管平滑肌脂肪瘤，本组病例中有3例为上皮样血管平滑肌脂肪瘤，肝脏上皮样血管平滑肌脂肪瘤具有恶变潜能^[2]，Luo等^[3]认为当病灶出现坏死、核异常分裂等提示其恶变。肝脏血管平滑肌脂肪瘤常发病于中青年女性，本组患者平均年龄54岁，且6例患者均为女性，与相关文献报道一致^[4]。该肿瘤大多表现为单发，也可多发，大约5%~10%患者伴结节性硬化和肾血管平滑肌脂肪瘤^[5]。本组研究对象均为单发，所有患者均未伴发结节性硬化及肾血管平滑肌脂肪瘤。肝脏血管平滑肌脂肪瘤影像学表

现具有一定特征性，一般影像学表现出脂肪密度或信号时较容易诊断，但因其组成成分存在一定差异，特别是该肿瘤缺乏脂肪成分时，易致误诊情况发生^[6]。本组病例均为影像学上缺乏脂肪型病例。肝脏血管平滑肌脂肪瘤组织形态学存在一定差异，但其免疫组织化学染色最为显著的特点为表达 HMB45 及 SMA 阳性^[7]。本组6例患者 HMB45 及 SMA 均阳性，免疫组织化学染色有利于患者诊断。6例患者病灶周围均可见粗大血管影，较多文献将其定义为引流血管^[8]，增强扫描后呈现为动脉瘤样强化，可能为血管平滑肌脂肪瘤中的血管瘤型者，该类型肿瘤所含脂肪成分较少，因此于影像学上难以见显著脂肪组织影像。经 CT 扫描后，其主要表现为类圆形低密度灶，经 MRI 扫描检查，其主要表现为长 T1 和稍长 T2 信号，且边界清，弥散不受限。经动态增强扫描后，动脉期出现明显强化，门脉期减退，呈现“快进



- Note: a: The lesion in the right posterior liver lobe was 1.0cm×0.9cm in size (arrow), which showed slightly low density and in non-enhanced CT images.
 b: Contrast-enhanced scanning showed that the lesion was slightly enhanced in arterial phase and the enhanced blood vessel was seen around the lesion.
 c: The lesion decreased in the portal phase obviously.
 d: Low density component was seen on in-phase.
 e: The tissue demonstrated no decreased signal intensity on out-of-phase image.
 f: The mass showed slightly high signal intensity on MR T2-weighted images.
 g, h: High signal intensity both on DWI and ADC images.
 i, j, k: Contrast-enhanced scanning showed that the lesion was significantly enhanced in arterial phase (i), the lesion decreased in the portal phase obviously (j) and delayed phase (k).
 l: Tumor disorder showed predominance of epithelioid-spindle cells, with no fat tissue (HE staining ×200).

Figure 2 A HAML patient initially diagnosed as metastatic tumor from lung cancer, underwent CT examination, pathologically proved as HAML

快出”的强化模式,本组病例有3例误诊为原发性肝细胞肝癌;由于动脉期强化非常明显,呈“灯泡征”被误诊为肝海绵状血管瘤,故 HAML 需要与上述疾病进行鉴别。很多文献中 HAML 表现为富血供占位,且周围或内部出现引流血管的机会较高(67%~82%),本组病例均出现,明显较其他文献高,可能与本组病例仅纳入乏脂肪型 HAML 有关,而其他文献报道的并未区分出含明显脂肪或乏脂肪,因此,该征象可能为乏脂肪型 HAML 的重要征象之一^[9-12]。目前功能影像学的研究应用于 HAML 与肝脏其他疾病的鉴别诊断上也非常热门^[13]。

HAML 需与以下疾病鉴别诊断:①肝细胞肝癌:患者往往有肝炎、肝硬化病史,AFP 阳性,两者的强化方式均呈“快进快出”,动态增强早期多数肝细胞肝癌呈显著结节状或团块状强化^[14,15],HAML 动脉期为均匀的明显强化,比常见肝癌的强化程度更加高,肝癌门脉期迅速下降为低密度或低信号,而 HAML 门脉期病灶虽然强化退出,但是大部分较周围肝实质呈略高或等密度或者信号,肝癌的假包膜呈现延迟强化,而 HAML 假包膜早期即可强化等,肝癌在 MRI 的 DWI 上信号高于 HAML,肝癌的 ADC 值降低,表现为弥散受限,而 HAML ADC 值也呈高信号,表现为弥散未受限。也有部分肝癌中含脂肪成分,主要为肝细胞脂肪变性所致,而 HAML 为成熟的脂肪细胞^[16]。②肝海绵状血管瘤:肝血管瘤呈 T2WI 高信号,增强后逐渐向中心推进性强化,而 HAML 动脉期即为均匀明显强化。③肝局灶性结节增生:病灶动态增强早期明显强化,门脉期病灶的密度或信号仍高于肝实质,约 50%可见肿瘤中心星芒状疤痕,可与 HAML 鉴别。④转移瘤:原发恶性肿瘤病史,肝内病灶呈大小均匀多发结节,中央易出现坏死,CT 增强环形持续性强化伴中央无强化区,呈牛眼征,如有原发肿瘤病史时较难鉴别。

综上所述,可以从以下几个方面来考虑肝脏病灶是否为血管平滑肌脂肪瘤:无肝炎、肝硬化病史的肝占位,有脂肪信号,当影像学脂肪显示不明确时,CT 及 MRI 检查动态期明显均匀强化,类似于“灯泡征”,磁共振弥散不受限,门脉期及延迟期为低、等或略高密度或者信号,周围可见粗大的血管影,应考虑本病的可能。

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