Rare primary liver tumors - MRI pictorial review

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Learning objectives

The aim of this exhibit is to review rare primary liver tumors and to illustrate their magnetic resonance imaging (MRI) findings, showing features that may be consistent or even diagnostic for these entities.
Background

MRI is an excellent method to evaluate focal liver lesions and offers advantages in detection and characterization of common liver lesions. In most of the cases it can obviate the need for biopsy. However, it is not clear that confident diagnosis can be made of tumors that are very uncommon.

With this poster the authors provide a pictorial review of rare primary malignant liver lesions.

Primary malignancies of the liver can be divided according to its origin: hepatocellular, cholangiocellular or mesenchymal.\[1\]

In the group of primary malignant liver lesions with hepatocellular origin the most common is the hepatocellular carcinoma (HCC). Fibrolamellar carcinoma and hepatoblastoma are the least common ones.

From the group of cholangiocellular origin we will discuss biliary cystadenocarcinoma, since isolated intrahepatic cholangiocarcinoma is relatively common.

While HCC and isolated intrahepatic cholangiocarcinoma are excluded, the rarer combined hepatocellular-cholangiocarcinoma will be discussed.

From the group of primary liver lesions with mesenchymal origin we will discuss angiosarcoma, leiomyosarcoma, Ewing’s sarcoma, epithelioid hemangioendothelioma and primary lymphoma.

FIBROLAMELLAR CARCINOMA (Fig. 1)

Fibrolamellar carcinoma (FLC) is a slow-growing tumor that, contrarily to HCC, arises in healthy patients with normal liver and no known predisposing risk factors.\[1\] It usually affects younger adults apparently with no gender predominance.

This tumor is composed of cords of neoplastic hepatocytes separated by lamellar fibrous bands forming a radiating pattern with a central scar that may have calcifications.

On MRI FLC shows hypo or iso-signal on T1-weighted images and iso or hyper-signal on T2-weighted images. The fibrous scar is usually hypointense on both T1 and T2-weighted images. After administration of gadolinium-based contrast agents, there is an intense
heterogeneous enhancement on the arterial phase. FLC may resemble hepatocellular
carcinoma as it may show portal or delayed washout. The scar has minimal or no
enhancement, although it can show enhancement on late phase. If the central scar is
necrotic, it shows hyperintensity on T2-weighted images mimicking the central scar of
focal nodular hyperplasias (FNH) on pre-contrast images.¹¹

Distinctive features from FNH may be the scar characteristics (low-signal on T2-
WI, and radiating appearance extending from the center to the periphery) and the
heterogeneously early enhancement on FLC. ²³

**HEPATOBLASTOMA (Fig. 2)**

Hepatoblastoma is the most common primary liver neoplasm in childhood. It is more
frequent in males and it usually develops in the first 3 years of life.

More often it appears as a large lobulated solid mass, but it can be multifocal in 20% of
cases. Analogous to other sarcomas these lesions may have intratumoral cystic areas,
hemorrhage or necrosis, and may also contain calcifications. On MRI hepatoblastoma is
usually hyperintense on T2-weighted images and hypointense on T1-weighted images.
Areas of hemorrhage appear as hyperintense foci in T1-weighted images.

This tumor is hypervascular, enhancing heterogeneously after intravenous contrast
administration. ¹¹

**MIXED (COMBINED) HEPATOCELLULAR CARCINOMA-CHOLANGIOCARCINOMA
(Fig. 3, 4)**

Mixed Hepatocellular (HCC)-cholangiocarcinoma (CC) is a rare type of primary hepatic
tumor which contain both hepatocellular and cholangiocellular components, also referred
to as "biphenotypic" tumors. ⁴

Usually this type of mixed carcinoma arises in patients with chronic hepatic disease.
The presence of imaging features of both HCC and cholangiocarcinoma is the main
characteristic that allows the radiologist to consider this diagnosis.

This tumor may appear hypointense on T1-weighted images and show intermediate to
high signal intensity on T2-weighted images. On the dynamic study the findings in the
arterial phase varies according to the dominant histological component. The lesion should
show hypervascular behavior if the HCC component is dominant and hypovascular if CC
is the main component. Early ring-enhancement with progressive enhancement centrally,
or heterogeneous early enhancement with partial washout are the typical findings. The use of a hepatocellular agent may be useful to differentiate a mixed-HCC-CC from a CC, since CC more frequently shows complete target appearance on hepatobiliary phase.

The main differential diagnosis is metastatic disease, even in the absence of a known primary malignancy given the fact that mixed HCC-CC is such a rare entity. The presence of central necrosis and the multiplicity of lesions favors the diagnosis of metastasis.

**BILIARY CYSTADENOCARCINOMA (Fig. 5)**

Biliary cystadenocarcinoma is a rare cystic neoplasm which arises from biliary epithelium and is typically mucinous. It usually occurs in middle-aged woman and there are no associated pathogenic factors.

This lesion presents as a large lobulated cystic mass, with one or multiple cystic compartments.

On MR imaging, biliary cystadenocarcinoma typically shows high signal intensity on T2-weighted images and variable T1-weighted signal intensity due to proteinaceous or blood products content. Septal or mural calcifications (depicted as signal voids on all sequences) and fluid-fluid levels are occasionally seen. Post-contrast sequences may demonstrate enhancement of the capsule, septa, and any mural nodules.

Despite the distinction between biliary cystadenoma from biliary cystadenocarcinoma may not be determined by imaging alone, the combination of thick irregular septations and nodules is suggestive of cystadenocarcinoma, while thin septations suggest cystadenoma.

The features that distinguish both cystadenoma and cystadenocarcinoma from other cystic liver lesions are the multiloculated appearance with variable signal intensity on T1 and T2-weighted images and the septal/solid nodules enhancement.

**HEPATIC EPITHELIOID HEMANGIOENDOTHELIOMA (Fig. 6, 7)**

Hepatic epithelioid hemangioendothelioma (HEHE) is a rare malignant neoplasm. It is more common in women and is usually detected incidentally, with no proven risk factors associated. This tumor is in the spectrum of vascular tumors and has clinical and biological behavior intermediate between hemangioma and angiosarcoma.
This entity may have two different patterns: early multifocal nodular type, and a more advanced diffuse type. The multiple nodules tend to coalesce and form diffuse large masses. [1,7]

On MRI these lesions show low signal intensity on T1-weighted images and moderate to markedly high signal intensity on T2-weighted images. At the dynamic evaluation lesions appear to have "target" or "halo" sign, with a multi-layered appearance, characterized by peripheral rim enhancement on the arterial phase with inner border of low signal and central core delayed enhancement. These features reflect the active proliferation of the cellular peripheral margins of these lesions and their dense fibrous central stroma. [7]

The lesions are frequently peripheral and capsular retraction of the liver is commonly found[1], a feature that is non-specific since it may be present in a spectrum of both malignant and benign entities.[9]

The HEHE make differential diagnosis with hemangiomas as both may present with high T2 signal and peripheral enhancement, although HEHEs have a thin rim while hemangiomas usually have nodular discontinuous rim enhancement. HEHE usually preserve a round morphology, are more numerous and show a persistent inner border of low signal compared to the centripetal homogenous progression of hemangiomas. [7]

Another differential to keep in mind is metastasis, which may have central enhancement and peripheral washout (hypervascular metastases). However, in HEHE the low signal inner border does not represent washout, but rather represents an area of true hypoenhancement, since there is no hypervascular enhancement on early phases. [7]

**PRIMARY LIVER LYMPHOMA (Fig. 8)**

Lymphoma of the liver can be either primary or secondary, the latter being the most common. Primary liver lymphoma is very rare and it can present as a single lesion, as multiple lesions or as a diffusely infiltrated liver. **Burkitt lymphoma (Fig. 9)** is a type of high-grade non-Hodgkin lymphoma and is usually associated with HIV/AIDS.

A solitary lesion is the most common manifestation and usually has variable mild to moderate high signal lesion on T2-weighted images, with low signal on T1-weighted images. [10]

Because of its poor vascularity, after intravenous contrast injection the majority of these tumors remain hypointense, with a thin peripheral rim probably due to a process of vasculitis within the adjacent liver parenchyma [11]
A distinctive finding is that the tumor encases the vessels instead of deviating them.

**SARCOMAS**

Sarcomas in general usually show prominently low T1 and high T2 signal. Post-contrast enhancement features include early heterogeneous peripheral enhancement and diffuse enhancement followed by progressive heterogeneous enhancement.

**EWING’S SARCOMA (Fig. 10)**

Ewing's sarcoma is a rare tumor that occurs predominantly in the long bones in pediatric population but can also occur in soft tissues and viscera. Is has been reported to occur in the liver as primary site although it is rare and there are few cases in the literature, the reason why there is a poor knowledge about its imaging features. In the few described cases in the literature there have been reports of large solid tumors with cystic lesions, an enlarged liver without a clear image of a nodule, and a multilocular cystic lesion. [12]

**ANGIOSARCOMA (Fig. 11)**

Primary hepatic angiosarcoma is a highly malignant and rapidly progressing tumor. It usually affects patients with 60-70 years and has a male predominance. [13]

The appearance on MRI is variable due to its heterogeneous structure with hemorrhagic and hypervascular nature. On T1-weighted images, dominant lesions are hypointense and contain hyperintense areas suggestive of hemorrhage. On T2-weighted images mass lesions are heterogeneous with high signal intensity areas of hemorrhage or necrosis and also low intensity areas of hemosiderin deposition, fibrosis or recent hemorrhage. [13,14] An image of fluid-fluid level in these lesions is typical of intra-lesional hemorrhage. [14] On the dynamic study there is markedly heterogeneous pattern of enhancement on arterial and portal phase while on delayed phase there is progressive filling. [13,14]

**LEIOMYOSARCOMA (Fig. 12)**

Primary liver leiomyosarcoma is a rare entity that arises from smooth muscle of the intrahepatic vascular structures or bile ducts. No underlying etiologic factors are known
even though it has been reported association with AIDS, Epstein-Barr virus, among others.\textsuperscript{[15]}

On MRI this tumor shows homogenous or heterogenous hypointensity on T1-weighted images and hyperintensity on T2-weighted images. \textsuperscript{[15]}
Fig. 1: Pathology proven fibrolamellar hepatocellular carcinoma in a otherwise healthy 16 year-old male: axial T2-weighted fat-suppressed images (a), and T1-weighted fat-suppressed before (b) and after administration of gadolinium in the late hepatic arterial phase (c) and the venous phases (d). Coronal T1-weighted images after gadolinium administration in the delayed phase (e). A large lesion in the right hepatic lobe is hyperintense on T2-weighted images and hypointense in the unenhanced T1-weighted images. A central scar radiating from the center is depicted, showing low signal on T2-weighted images. At dynamic study there is heterogeneous moderate enhancement of the lesion on the late arterial phase images with negligible enhancement of large radiating scar. In this case the lesion retains contrast on late phase images appearing moderately hyperintense to background liver while portions of central scar enhance. Note the healthy features of the surrounding liver parenchyma.
**Fig. 2:** Hepatoblastoma: large liver lesion in a 3 year-old child showing moderate heterogeneous high signal on T2-weighted images with dispersed intralesional cystic areas (a) and low signal on T1-weighted images without and with fat-suppression (b,c). On the dynamic study it shows heterogenous enhancement after intravenous contrast administration on the arterial phase (d) with subtle washout on the portal-venous phase (e). Some areas of hyperintensity are depicted on T1-weighted images probably due to hemorrhagic foci.

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Fig. 3: Mixed HCC-Cholangiocarcinoma in 58-year-old man: The lesion is heterogeneously hyperintense on T2-weighted fat-suppressed images (a) and hypointense on pre-contrast fat-suppressed T1-weighted images (b). On the dynamic study it shows early peripheral ring-enhancement on the hepatic arterial dominant phase (c) with portions showing peripheral washout and portions showing progressive enhancement in the central region on the interstitial phase (d). Note the progressive enhancement in the central region (d).

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**Fig. 4:** Mixed HCC-Cholangiocarcinoma in a 55-year-old woman hepatitis B and C and cirrhosis: The lesion is hyperintense on fat-suppressed T2-weighted images (a), with nodular ring-enhancement on hepatic arterial dominant phase (b). There is progression of the enhancement on the interstitial phase (c) with contrast retention.

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Fig. 5: Biliary cystadenocarcinoma: multiloculated lesion with high signal intensity on axial T2-weighted images (a), and low signal on T1-weighted images before (b) contrast administration. The thick septa and the solid mural nodules show slight enhancement on post-contrast T1-weighted images at the venous (c) and interstitial phases (d). Note the thick and irregular septa and the mural/septal nodularity suggestive of cystadenocarcinoma.

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Fig. 6: Diffuse type of epithelioid hemangioendothelioma: Multiple nodules that tend to coalescence in the right hepatic lobe, showing mild to moderately high signal intensity on coronal T2-weighted images (a) and moderate to low signal intensity on T1-weighted out-of-phase images (b). During the dynamic study they show peripheral enhancement on the arterial phase on T1-weighted images (c) with some lesions showing persistent peripheral enhancement and few lesions showing central core enhancement on the interstitial phase (d).

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Fig. 7: Multifocal nodular type of epitheloid hemangioendothelioma: the lesions show moderate high signal intensity on fat-suppressed T2-weighted images (a), low signal on T1-weighted images without (b) and with fat-suppression (c). After contrast administration the lesions show peripheral enhancement on arterial phase with central hypointensity (d) and delayed central enhancement of most lesions (e).

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Fig. 8: Primary lymphoma. The lesion shows low signal intensity on fat-suppressed T2-weighted images (a) and on unenhanced T1-weighted in (b) and out-of-phase (c) images.
After intravenous contrast injection the lesion is hypovascular and shows low signal intensity on the arterial phase (d), with mild progressive enhancement on the venous phase (e - axial, f - coronal). Note minimal mass effect and the presence of portal vein branches encased by the lesion.

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**Fig. 9:** Burkitt lymphoma. The tumor is mild hyperintense on T2-weighted images (a), and hypointense on fat-suppressed T1-weighted images (b). On the dynamic study it shows heterogeneous progressive peripheral arciform enhancement on the arterial (c), portal venous (d) and interstitial (e) phases on fat-suppressed T1-weighted images. On
the delayed phase (e) there is progressive filling of the central area. Note minimal mass effect and the presence of portal vein branches encased by the lesion.

Fig. 10: Ewing’s sarcoma. This large tumor is heterogenous with mild hyperintesity on T2-weighted images (a, b). Note the moderate to high central signal on the fat-supressed T2-weighted images (a, b) and mild high signal on T1-weighted images (c), probably reflecting blood content. This lesion is hypovascular with slight progressive enhancement towards the interstitial phase on fat-supressed T1-weighted images (d, e, f).
**Fig. 11**: Hepatic angiosarcoma: the tumor is heterogeneous on fat-suppressed T2-weighted images with high signal intensity areas of hemorrhage/necrosis and low intensity areas of hemosiderin deposition, fibrosis or recent hemorrhage. (a). On fat-suppressed T1-weighted images the tumor is predominantly hypointense and contain hyperintense areas suggestive of hemorrhage (b). On the dynamic study there is heterogeneous enhancement on arterial (c) and portal phase (d) of fat-suppressed T1-weighted images, while on delayed phase there is progressive filling through the 3minute (e) and 10min (f) phase.

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Fig. 12: Primary hepatic leiomyosarcoma: the tumor is heterogeneous with moderate hyperintensity on T2-weighted images (A) and hypointensity on unenhanced fat-suppressed T1-weighted images (B). On the dynamic study there is heterogeneous enhancement on the arterial and venous phase (c, d) with a central area of hypointensity that persists on the 10min phase (e).

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Findings and procedure details

The authors review MR cases of some uncommon primary liver lesions. A pictorial review was made with examples including lymphoma, sarcomas, epithelioid hemangioendothelioma, fibrolamellar hepatocellular carcinoma, hepatoblastoma, mixed hepatocellular-cholangiocarcinomas and biliary cystadenocarcinoma.
Conclusion

Less common primary liver lesions may pose a challenge for the diagnosis; however some distinctive MR features may suggest the correct diagnosis.

It is important for the radiologist to be familiar with these uncommon entities and their MRI findings.

The knowledge of their main MRI characteristics allows the radiologist to recognize some of the most important findings and to narrow differential diagnoses.
References


