MRI rare finding: Absence of the left liver lobe

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ABSTRACT

We report a rare case of left liver lobe absence in an 80-year-old male patient discovered during an MRI scan. The main imaging features of this condition are briefly reviewed, together with its pathogenesis and the most common associations and differential diagnoses.

1. Introduction

Absence of the left anatomical lobe of the liver is a rare condition. Few cases have been described in the literature [1–6]. Absence of a lobe of the liver can be due to several events, such as neoplasia, chronic damage, trauma, prior surgical resection, and agenesis [5]. Agenesis is defined as the absence of liver tissue without any evidence of disease or previous surgery; it may be associated with other anatomical alterations, such as hypertrophy of other liver segments, colic interposition between the liver and diaphragm, anomalous position of the gallbladder. Agenesis is usually asymptomatic, with normal liver function parameters; it is noted incidentally at autopsy, or in radiological studies.

2. Case report

Our patient was an 80yo man, without any history of surgery or liver disease who underwent an ultrasound examination at our institution for new onset of aspecific abdominal pain, which was not diagnostic because of severe abdominal meteorism. However, a vascular anomaly (a porto-caval shunt) was suspected and therefore a magnetic resonance study was requested.

The MR exam showed the absence of the left lobe and of the left hepatic vein and artery and recanalization of the paraumbilical vein (Fig. 1). The right lobe of the liver appeared without any parenchymal alteration. Portal vein was slightly dilated (16 mm). There were signs of portal hypertension: splenomegaly, dilatation of the splenic vein, recanalization of paraumbilical vein. Gallbladder was in its normal place. It could be noted an interposition of colic tract between the liver and anterior abdominal wall (Fig. 2), responsible of the technical difficulties encountered when performing the ultrasound examination. Portocaval shunt was not confirmed. Another finding was the incomplete rotation of the left kidney with an anteriorly placed hilum and a laterally located ureter (Fig. 3).

3. Discussion

Lobar absence of the liver is usually incidentally found; it is mostly an asymptomatic condition which has no consequences in patient prognosis. However, it is essential to understand if it is acquired (atrophy) or congenital (agenesis). In fact, in case of absence of one lobe or a part of hepatic parenchyma, some other conditions should be excluded, such as cholangiocarcinoma, cirrhosis or hydatid cysts [3].

Even if there could be many different acquired conditions linked to liver atrophy, they all have one common pathophysiology: portal vein and/or biliary obstruction. The relationship between portal vein obstruction and atrophy is an expression of the architecture of hepatic parenchyma, in which portal vein circulation is essential, more than arterial supply, for the normal function of hepatocytes. Apparently, portal vein disturbances is more problematic than bile ducts drainage obstruction; in fact, the attempt to provide surgically a biliary drainage in an atrophic lobe is usually vain [7].

Some cases of lobar atrophy caused by hepatolithiasis and portal flow disturbances were reported, not necessarily linked to a malignant pathology [8]. However, the majority of cases of lobar atrophy described were caused by hilar cholangiocarcinoma [7,9,10]. This type of tumor has an aggressive behavior, responsible of venous obstruction as an early manifestation because of its crucial location and the low resistance of the veins walls to compression [7]. To differentiate atrophy from congenital absence of the liver, it is useful to look at...
some characteristic imaging findings that can help in difficult cases. For example, it is not uncommon that in secondary chronic pathologies may be present a compensatory hypertrophy of a not affected liver segment [11]; it can be noted a “countour step” between atrophic and normal liver parenchima with associated dilated biliary ducts. Moreover, the atrophied liver goes through pathophysiologic changes as oedema, fibrosis and arterio-portal shunting. These changes can be detected as alterations in signal in MR and density on CT, such as: low attenuation on pre-contrast CT, higher attenuation on arterial phase in contrast enhanced CT (arterial blood flow supplies the paucity of portal flow), decreased signal intensity on T1-weighted images and an increase of signal on T2-weighted images in MR [11].

In our case, there were no evidences of biliary ducts dilatations, or portal vein paucity. The imaging study showed no morphologic alterations of the liver, neither sufferings of the tissue, and no deficit in parenchymal function were found during the delayed phase of contrast enhanced MR. All these signs, in addition to the non-rotation of the left kidney and the negative anamnestic history of the patient, pointed to agenesis of the left lobe.

The agenesis may be due to an interruption of development in a specific embryologic stage. As suggested by Prithishkumar et al. [1], variations of liver morphology may be due to a partial arrest of development of the hepatic epithelial trabeculae in the left lobe of liver at developmental stage 13 or 14 [12,13]. From the stage 13 begins the development of tissue architecture of the liver. Other studies have shown that knockout of WT1 gene (Wilm’s tumor 1 homologue) in these developmental stages, a marker of mesothelial cells, yields smaller livers [14]. These livers were hypoplastic due to a quantitative reduction of both hepatoblasts and hepatocytes [15].

Developmental absence is a diagnosis of exclusion. Even if it is not dangerous in itself, it is necessary to evaluate anatomic variations that can be associated, especially in the case of patients who are candidates to abdominal surgery. It is important to be aware of this condition, which could otherwise cause imaging misinterpretation. For our patient, the imaging features of bowel interposition were similar to those of Chilaiditi’s syndrome, which is often found also in patients with normal hepatic morphology, and which made difficult the first US examination. The vascular structures were also misinterpreted, and at first this misunderstanding raised the suspicion of a porto-caval shunt.

Generally, it is important to search a possible cause for lobar absence firstly, in order to exclude a life threatening pathology (e.g. cholangiocarcinoma). Once a possible metabolic, carcinomatous, traumatic, vascular or infectious cause has been excluded, it is important to evaluate the associated anatomic anomalies [16]. The awareness of this condition is crucial for the radiologist, who may be the first to orientate to the right diagnosis without any misinterpretation [3].

In our opinion, to completely understand the whole anatomical picture, including associated vascular anomalies and other anatomical variations, the imaging modality of choice should be contrast enhanced CT or MR, because of their great spatial and contrast resolution. However, MR yields more information about parenchymal structure and composition; furthermore, it gives the possibility to study intra and extra hepatic biliary ducts only with strong T2-weighting, without the use of any contrast injection (cholangio-MR) and with no radiation exposure. Moreover, hepato-specific contrast agents may be useful to
characterize liver tissue, and to obtain a detailed picture in doubtful cases, in order to make the correct diagnosis.

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Conflicts of interest

No conflicts of interest.

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