Liver teratoma

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Abstract

True liver teratomas are exceptional tumors, with only 25 cases reported in the radiology literature. Most cases reported were either intraperitoneal or retroperitoneal teratomas that had invaded the liver. The authors present a 27-year-old woman with a liver complex mass found incidentally at computed tomography (CT). The different imaging studies demonstrated that the lesion was composed of macroscopic fat, with a central soft tissue component and a calcification. The patient underwent surgery and the final diagnosis was a benign liver teratoma. (Acta gastroenterol. belg., **2008**, 71, **275-279**).

Introduction

Teratomas are neoplasms that arise from pluripotent cells (1) and contain a diversity of tissues that normally are not found within the organ in which they arise. So they are composed of a multiplicity of diverse tissues that are in distinct topographic relationship to each other and are different to their anatomic site of origin (2). These tumors originate in descending order of frequency from ovaries, testes, anterior mediastinum, retroperitoneum, coccygeal and presacral areas, pineal and intracranial regions, neck and abdominal viscera (2). There have been rare cases of these tumors occurring in the gastrointestinal tract, liver, nasal sinuses, cervix, thyroid and pancreas (3,4). Teratomas of the liver are rare neoplasms with only a few isolated case reports in the radiology literature (5,6).

Most are found incidentally. The diagnosis is usually easy to made because the imaging findings are characteristic.

Mature cystic teratoma can be associated with complications such as rupture, malignant degeneration, or torsion (when located in the ovary) (7). The rupture of the tumors can cause leakage of the liquefied sebaceous contents into the peritoneum (8). Malignant degeneration of mature cystic teratomas consists of differentiated tissues giving rise to carcinoma or sarcoma (7).

Surgery is usually the treatment of choice.

We report a case of an adult female with a benign mature teratoma arising in the right hepatic lobe with relevant imaging features, including ultrasound (US), computed tomography (CT) and magnetic resonance (MR) imaging.

Case report

A 27-year-old woman with a personal history of asthma presented to our hospital with fever and dyspnoea. A thoracic and abdominal CT scan was requested and revealed an incidental liver complex mass.

The topogram (Fig. 1) showed a radiolucent image with a central calcification projecting on the right upper quadrant.

At contrast-enhanced CT (Fig. 2), the mass was round, heterogeneous, and was composed predominantly by fat with a solid soft tissue component, had a central coarse calcification, and didn't enhanced after intravenous contrast administration. The lesion measured 9,7 cm and was located on the posterior segment of the right hepatic lobe.

At ultrasound examination (Fig. 3), the liver mass was heterogeneous and mainly hyperechoic, because of the fat component of the lesion. MR imaging was also performed in order to better characterize the lesion before surgery. The peripheral areas and the central component demonstrated increased signal intensity on the T1-weighted images (Fig. 4A) but decreased signal intensity on the fat saturation T2 weighted images (Fig. 4D), confirming the presence of macroscopic fat, excluding other types of material, such as blood, concentrated protein and melanin that could mimic this appearance. The calcification had low signal intensity on all the pulse sequences and the soft tissue component was relatively hyperintense to muscle on the T1 and T2-weighted images (Fig. 4A, 4B and 4C). The central portion of the mass demonstrated some focal area of increased signal intensity on the contrast-enhanced MR images (Fig. 4E).

The patient underwent surgery. The right hepatic lobe and caudate lobe were resected. On the macroscopic view, the mass was solid and contained mainly fat and also hair and calcifications (Fig. 5).

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Submission date : 22.09.2007
Revised version : 20.04.2008
Acceptance date : 24.04.2008

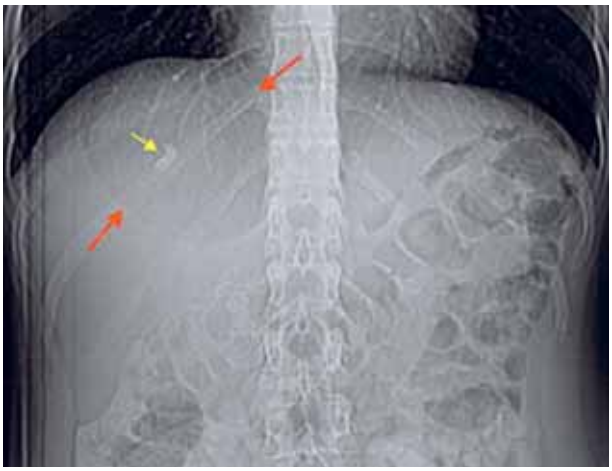

Fig. 1. — Topogram : radiolucent mass (red arrows) with a central calcification (yellow arrow) projecting on the right upper quadrant.

Fig. 3. — Hepatic US with color Doppler showed a heterogenous mass, mainly hiperecoic because of the fat component, abutting the right hepatic vein.

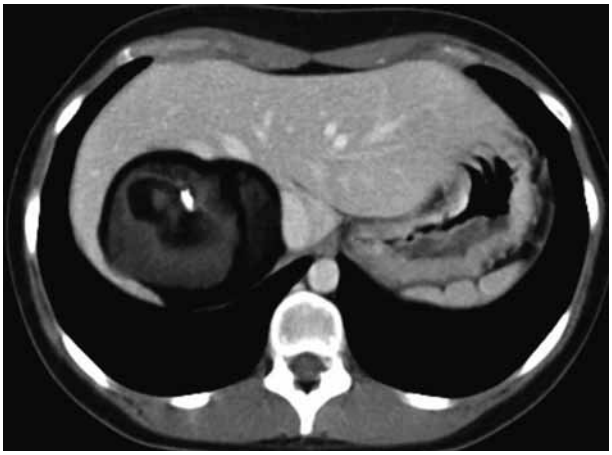

Fig. 2. — Abdominal CT scan (portal phase) showed a well defined lesion in the posterior right lobe of the liver. The lesion was a combination of fat, calcification and soft tissue components, with no enhancement after intravenous contrast administration.

The final pathologic diagnosis was a benign liver teratoma.

Two years after the surgery the patient remains well.

Discussion

True liver teratomas are extremely rare ; of the 25 hepatic teratomas described in the literature, only five have occurred in adults (9).

Most so-called "hepatic teratomas" represent either intraperitoneal or retroperitoneal teratomas that have invaded the liver (10).

The majority of the cases were in female children below the age of three (1), likely reflecting their proposed congenital origin (11).

A teratoma must contain tissue derivatives of at least two or more germ cell layers -namely, ectoderm, mesoderm and endoderm- and have evidence of organ formation (12). This combination of tissues is unrelated to the organs where the tumor originates (5). Embryologically, there are thought to arise from primordial germ cells that arrest along their migration path from the allantois hindgut to the gonads during the first weeks of life (13), hence there common midline and paramedial location (11). During this migratory process, a few of the primordial germ cells may become isolated in an aberrant location. Under normal conditions, germ cells not invested by the sex cords will degenerate and die. Teratomas are thought to arise from unincorporated pluripotential germ cells, which fail to involute and continue to undergo mitosis (14,15).

Four histologic variants of teratoma are described : 1-mature teratoma, 2-immature teratoma, 3-teratoma with malignant transformation, and monodermal teratoma (16). Dermoid cysts are a special form of mature teratoma in which there is predominantly an ectodermal derivation. There are characteristically uniloculated cysts lined by skin, complete with special structures such as sebaceous glands, hair follicles, and teeth, and are filled with off-white, cheesy, sebaceous material (17).

Most are discovered incidentally, as in this case, as there is no typical clinical presentation (11). Compression of the surrounding structures can cause symptoms of abdominal distension, nausea and vomiting (9).

Hepatic teratomas are usually well encapsulated lesions and are easily resectable from the surrounding hepatic parenchyma (5), but in our patient the right hepatic lobe and caudate lobe had to be also resected.

Plain radiographs are useful in demonstrating the different densities of the lesion, namely, the fat opacity component which appear lucent and the calcifications (as we can see it on the figure 1 of our case).

At US these tumors may have a variety of appearances. The most common manifestation is a cystic lesion with a densely echogenic tubercule (Rokitansky nodule)

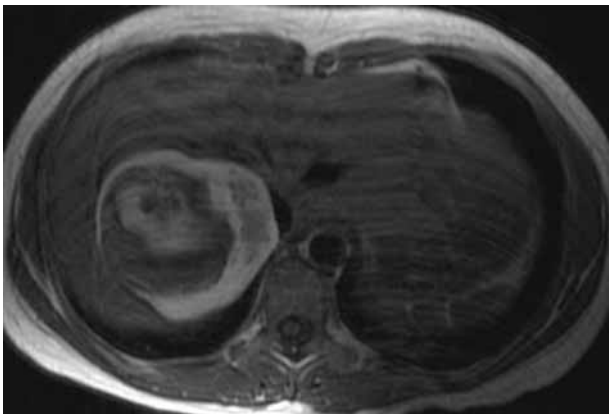

Fig. 4A. — Axial T1-weighted MR image : the peripheral and the central areas demonstrated increased signal intensity, and the soft tissue component was isointense to hyperintense relatively to muscle. The calcification had low signal intensity.

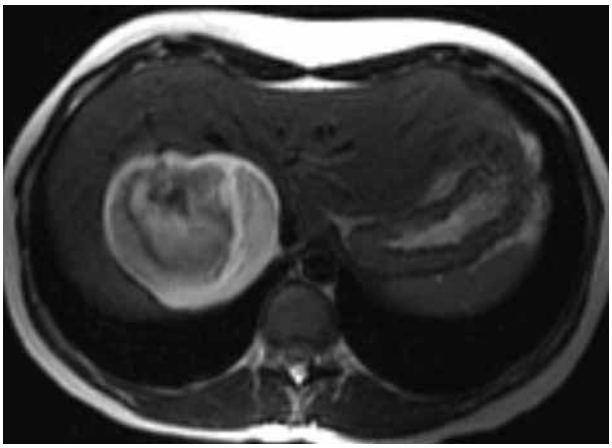

Fig. 4B. — Axial T2-weighted MR image : the peripheral and central areas demonstrated increased signal intensity. The soft tissue component was also hyperintense. The calcification had low signal intensity.

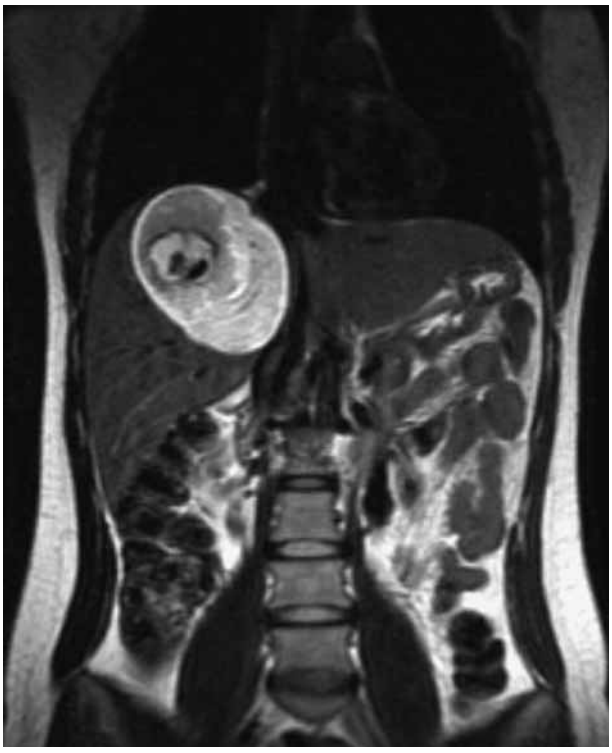

Fig. 4C. — Coronal T2-weighted MR image : the peripheral and central areas demonstrated increased signal intensity. The soft tissue component was also hyperintense. The calcification had low signal intensity.

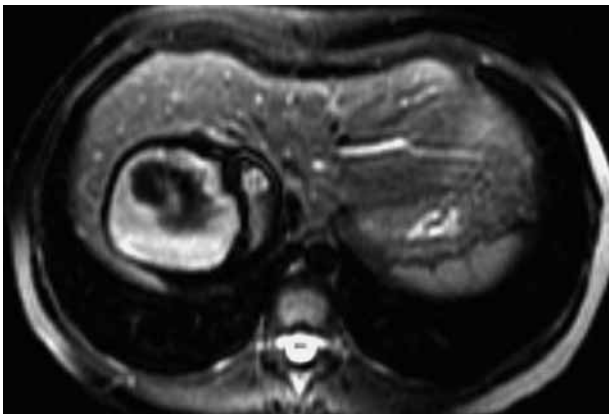

Fig. 4D. — Axial fat saturation T2-weighted MR image : the fatty component had decreased in signal intensity like the subcutaneous macroscopic fat.

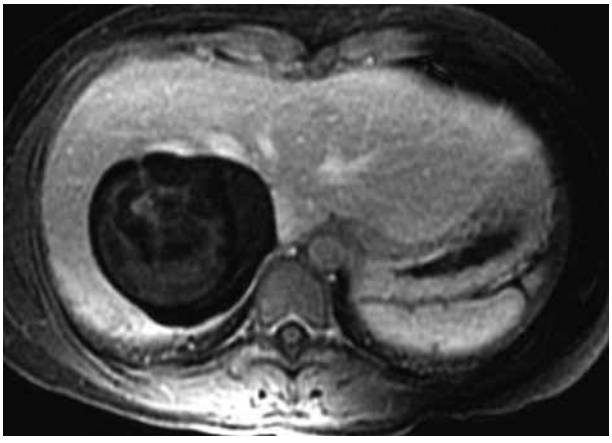

Fig. 4E. — Axial contrast-enhanced MR image showed minimal enhancement.

projecting into the cyst lumen (18). The second manifestation is a diffusely or partially echogenic mass (as we can see it on our figure 3) with the echogenic area demonstrating sound attenuation owing to sebaceous material and hair within the cyst cavity (19,20). The third manifestation consists of multiple thin, echogenic bands caused by hair in the cyst cavity (7). Fat-fluid levels, which occur due to the presence of sebum, are pathognomonic, with the macroscopic fat ranging from hypoechoic to hyperechoic, depending on the differences in acoustic impedance with the adjacent soft tissues (21).

Fig. 5. — Photograph of the gross pathologic specimen showed a lobulated mass, in the right hepatic lobe, that contained copious amounts of hair and macroscopic fat.

The appearance on CT and MR imaging depends on the amount of fat present (22). At CT, fat is of low attenuation (see figure 2) compared with normal liver parenchyma, with a range of -10 to -100 HU (22), and calcifications are markedly hyperattenuating foci (11). In this case there is also a normoattenuating soft tissue component.

Fat is high in signal intensity on T1-weighted MR images (23) (Fig. 4A). The signal intensity of the sebaceous component on T2-weighted images is variable, usually approximating that of fat (24). This combination of different signal intensities on T1- and T2-weighted images is not specific for fat and must be distinguished from blood, concentrated protein and melanin (25,26).

Frequency-selective fat suppression techniques reduce the signal intensity from lipid containing voxels and thus can be used for the detection of lipid (27). Saturation techniques are appropriate when evaluating fatty masses such as dermoid cysts and angiomyolipomas (27).

Calcifications are usually inferred by a lack of signal (11).

In this case, no cystic component is present.

The finding of a mass containing fat, fluid, and calcification is virtually diagnostic of a teratoma (28).

The differential diagnosis of fat-containing liver tumors includes a liposarcoma, a hepatocellular carcinoma (HCC) and an angiomyolipoma.

Primary liver liposarcoma is extremely rare (29). At US liver liposarcoma appears as a poorly defined, lobulated, infiltrating echogenic tumor with shadowing and heterogeneity secondary to areas of hemorrhage and necrosis (30). At abdominal CT and MR imaging the tumor is almost fatty, lobulated with a few, random distributed vascular structures and a small area of nodular enhancement (31), with no calcifications. Most hepatic liposarcomas are metastatic (10).

HCC accounts for 90% of primary malignant hepatic neoplasms (22). The majority of cases of HCC occur in patients with underlying cirrhosis. Areas of fatty metamorphosis and calcifications can also be identified, but fat deposition in HCC is usually patchy (22,10). Because of its arterial blood supply, HCC characteristically appears as a transiently hyperattenuating mass during the hepatic arterial phase of enhancement (22). Fibrolamellar HCC occurs in younger patients without underlying liver disease. Calcifications are present in 33%-55% of fibrolamellar carcinomas. Calcifications are punctate, nodular, or stellate and are usually located within the scar and less commonly in the tumor periphery (32). Fibrolamellar HCC usually doesn't contain fat.

Angiomyolipoma is a benign, unencapsulated mesenchymal tumor that is composed of varying proportions of three elements : smooth muscle cells, thick-walled blood vessels, and mature adipose tissue (10). In one study, angiomyolipoma (11/12 lesions) appeared as a hypodense lesion on unenhanced CT scans and markedly enhanced on the arterial phase with central vascular opacification in eight lesions (33). On the portal venous phase, eight lesions remained enhancing with central vessels seen in six lesions. The presence of central vessels within the lesions may be a characteristic feature of angiomyolipoma (29).

Teratomas present a variety of morphologic features, with different combination of fat, fluid and calcification. They occur most commonly in the ovaries, testes, anterior mediastinum, retroperitoneum, sacrococcygeal region, and cranium (21). Occasionally they can be found unexpectedly in atypical locations.

In the majority of the patients the diagnosis can be made before surgery, with CT and/or MR imaging which demonstrate perfectly the different tissues.

In conclusion, true liver teratomas are rare tumors. The presence of fat is the most striking feature in these lesions, and is an important clue to guiding a differential diagnosis.

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