Radiologic-pathologic comparison of undifferentiated embryonal sarcoma of the liver in a 61-year-old woman

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Mrs. V, a 61-year-old woman with no particular history, was seen for impairment of general health and abdominal pain. The physical examination found a tender mass in the right hypochondrium. The laboratory tests revealed the presence of inflammation (CRP: 350 mg/L, 15,000 neutrophils), cytolysis (ALT: 130 IU/L, AST: 150 IU/L: 4 N), anicteric cholestasis (alkaline phosphatase: 450 IU/L); tumor markers AFP (8 ng/mL) and CA 125 (27 U/mL) were normal.

The ultrasound (Fig. 1a, b) showed a voluminous hypo-hyperechoic heterogenous solid hepatic mass that had developed to the detriment of segment IV (19 × 25 × 21 mm), and responsible for dilatation of the left intrahepatic bile ducts. On the CT scan, it was tissular, heterogeneous, and compartmentalized, presented hemorrhagic changes, and had arteries running through it (Fig. 1 c, d). After injection, moderate enhancement was seen in the arterial phase, increasing in the delayed phase. The hepatic parenchyma was normal; the staging was negative.

On the MRI (Fig. 2), the lesion was clearly hyperintense on the T2-weighted images with multiple walls and cavities of varying sizes. After injection, homogeneous peripheral enhancement was seen with uptake by the septa, but not central enhancement.

The surgical resection, which was performed quickly due to the impaired general health, confirmed the cystic, encapsulated intrahepatic mass (Fig. 3a, b) centered in segment IV. The resection was complete. The surgical specimen weighed 3550 g and measured 27 × 22 × 10 cm. It was composed of multiple cavities of varying size with a necrotic
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Figure 1. a: ultrasound: cross-section, arrowhead: multiple hypoechoic lesions within an iso-echoic solid tumor lesion; b: ultrasound: longitudinal section, arrowhead: multiple hypoechoic lesions within an iso-echoic solid tumor lesion; c: CT scan: axial section without injection of contrast product, arrow: spontaneously hyperdense hemorrhagic changes within the tissue portion of the tumor; d: CT scan: sagittal section after injection of contrast product, curved arrow: arteries within the tumor.

Discussion

Characterization of an aggressive lesion that is clearly hyperintense on T2-weighted images is tricky in adults, since such lesions are rare.

Benign cystic lesions can be easily ruled out based on their symptoms, serologies, and clinical context: hydatid cyst, alveolar echinococcosis, biliary cystadenoma, biliary cysts, peribiliary cysts, and abscesses.

With malignant cystic tumors, there are often multiple cystic metastases and the primary one is most often found: endocrine, pancreatic, colonic, or ovarian.

Biliary cystadenocarcinoma is the malignant transformation of a biliary cystadenoma. It is the most common aggressive primary cystic lesion. This lesion is predominant in women over 60 years of age. It is characterized by the presence of multiple enhancing septa, associated with a fleshy nodule, which were absent in our case.

Much more rarely, aggressive cystic lesions of the liver may be sarcomas, and particularly angiosarcoma, which predominantly affects children. This is an aggressive multicystic vascular tumor that is moderately hyperintense in T2-weighted images and heterogeneous in T1-weighted images due to hemorrhagic changes. It is usually multifocal with splenic involvement. The outcome is rapidly unfavorable.

Myxoid sarcoma predominantly affects soft tissues and occurs most often in young subjects. Hepatic involvement is rare. Rare cases are described in the literature, including one in the round ligament [1]. Such tumors appear cystic due to their clear hyperintensity on T2-weighted images, which conflicts with the ultrasound data (tissue component).
That hyperintense signal reflects the myxoid stroma. Their enhancement is characteristic, with peripheral nodular contrast uptake that progresses over time.

The histology in our case study revealed an undifferentiated embryonal sarcoma (UES). The immunohistochemistry found specific positive markers: vimentin and alpha-1 antitrypsin [2]. And these lesions, originally described in 1978, predominantly affect children and adolescents [3]. In 2008, only 67 patients over the age of 20 presented with a UES; the mean age at the time of diagnosis was 35 years (range 18–60 years) [3]. On the ultrasound, these lesions were solid iso-hyperechoic masses [4]. The CT scan revealed a heterogeneous stroma: cystic with hemorrhagic changes, non-calcified, and partially enhancing after injection of contrast product [4]. The MRI signs showed a hypointense lesion on T1-weighted images, a clear hyperintense lesion on T2-weighted images corresponding to the myxoid matrix, and areas of spontaneous hyperintensity on T1-weighted images indicating hemorrhagic changes. The combination of a cystic component and heterogeneous enhancement with the different acquisition techniques should initially suggest a diagnosis of sarcoma, particularly if there are signs of severity, such as dilatation of the intrahepatic bile ducts [5]. In that case, these lesions should not be biopsied, but a surgical opinion should be immediately requested.

Despite their very poor prognosis (median survival 29 months), the currently recommended therapeutic management consists of surgical resection followed by adjuvant chemotherapy, which is what was done in our case study [6]. There is a high risk of recurrence after surgery; the mean post-therapeutic follow-up is 14 months [7].
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Figure 3. a: Macroscopic examination: orange-red encapsulated tumor (arrow head) measuring 27 × 22 × 10 cm, weight: 3550 g; b: macroscopic examination: split multicystic specimen (stars); c: microscopic examination (× 10 magnification): connective-appearing tumor proliferation of spindle-like polymorphic cells. Curved arrow: formed blood elements; d: microscopic examination (× 40 magnification, PAS staining): Elevated nucleocytoplasmic ratio. Arrows: intracytoplasmic eosinophilic granulations.

Conclusion

Undifferentiated embryonal sarcomas are very aggressive and rare tumors in adults. Due to the combination of a pseudocystic component on T2-weighted images contrasting with the tissue component fund on sonograms and heterogeneous contrast uptake, it is advisable to seek a surgical option immediately in order to avoid transparietal biopsies.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


