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Case Report

Undifferentiated embryonal sarcoma of the liver mimicking venolymphatic malformation

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ABSTRACT

Undifferentiated embryonal sarcoma of the liver (UESL) is a primitive hepatic neoplasm that presents in a variety of forms on ultrasonography, computed tomography, and magnetic resonance imaging. In this case report, we present an UESL with fluid-fluid cysts mimicking a radiographic presentation commonly seen in venolymphatic malformation on magnetic resonance imaging. This is the first described case of UESL, with this radiographic presentation and outlines, the importance of considering this malignant lesion when evaluating liver tumors in children.

Introduction

Primary liver tumors are rare in children and represent less than 2% of all neoplasms in children. Hepatoblastoma and hepatocellular carcinoma account for most pediatric liver malignancies. Undifferentiated embryonal sarcoma of the liver (UESL) comprises 9%-15% of the remaining pediatric liver malignancies, the incidence of UESL remains low with 0.6-1.2 cases per 1 million patients. Ninety percent of patients presenting with UESL are among the pediatric population, most often between 6 and 10 years of age.

Given the rarity of this tumor, reports of its imaging characteristics are scarce in the literature. As a result, UESL is often misdiagnosed as other types of hepatic malignant tumors [1]. One reason for misdiagnosis is the discrepancy between a solid appearance of the tumor on ultrasonography and cystic appearance on computed tomography (CT) [2,3]. Although magnetic resonance imaging (MRI) is commonly used to characterize liver lesions, to date few reports comment on specific MRI findings in UESL. Characterization of these undifferentiated mesenchymal liver tumors on MRI is further made challenging by their variable appearance due to their primitive histology. In a few MRI studies, UESL has been shown to be T1

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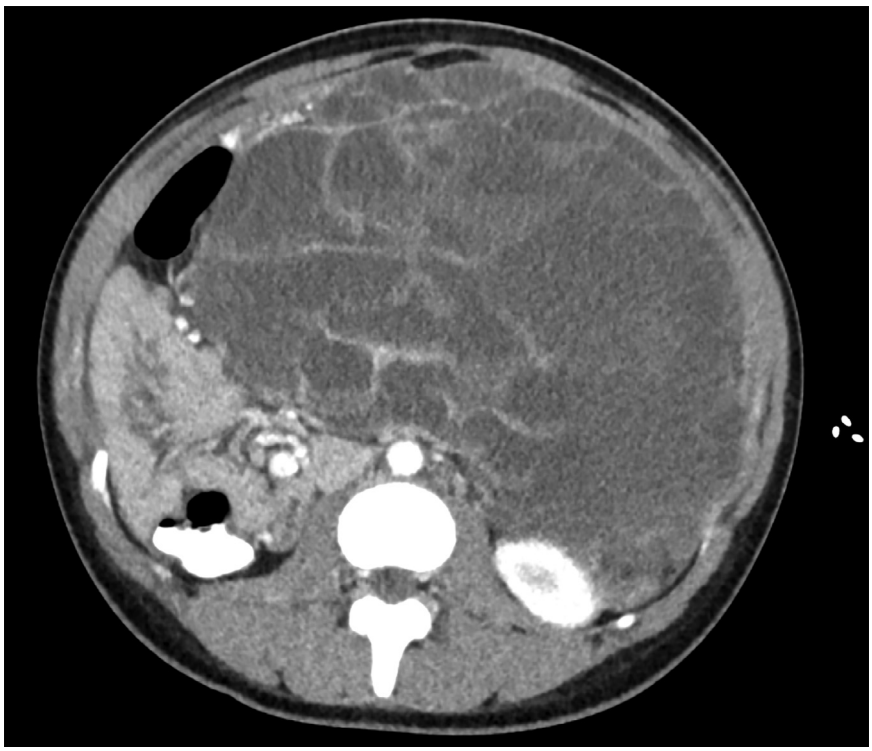

Fig. 1 – Axial enhanced CT of the abdomen during the portal venous phase in this patient with undifferentiated embryonal sarcoma of the liver demonstrates a complex multicystic exophytic mass occupying most of the left abdomen and pelvis inseparable from the left lobe of the liver.

hypointense and heterogeneously T2 hyperintense [2,4]. Our goal in presenting this case report is to review the challenges in distinguishing UESL from other liver neoplasms and share an unusual MRI appearance not previously reported.

Case report

A 11-year-old girl was admitted with a 3-month history of progressively worsening abdominal pain and distention with an acute inability to tolerate oral intake. On physical examination, the abdomen was distended with a palpable nontender irregular mass in the upper quadrants without a fluid wave. Serum laboratory examination results were as follows: alpha-fetoprotein (AFP) 1.0 ng/mL, Carbohydrate Antigen 125 Units/mL, total bilirubin 1.0 mg/dL, alanine transaminase 11 Units/L, aspartate transaminase 23 Units/L, alkaline phosphatase 77 Units/L, lactate dehydrogenase 275 Units/L, and albumin 3.7 g/dL.

A CT scan of her abdomen revealed a large complex multicystic mass that was $13 \times 19 \times 25$ cm in the left abdomen scalloping the left lobe of the liver (Fig. 1). The stomach was displaced into the right hemiabdomen. T2 weighted MRI examination confirmed a $25 \times 19 \times 14$ cm multicystic mass arising from the left lobe of the liver (Fig. 2). There was no rapid washout of contrast in the solid components, but there was progressive enhancement in the central portion of the cystic lesions. Multiple fluid-fluid levels were also observed. This constellation of radiographic findings led to a primary diagnosis of venolymphatic malformation.

Following admission, the patient underwent a left lateral hepatectomy. On gross examination, the tumor was composed primarily of multiloculated cysts separated by dense bands of tissue ranging between 1.2 and 6 cm in thickness. The cysts had focal hemorrhagic areas without necrosis (Fig. 3). Microscopically, the tumor was composed of cyst walls and solid areas containing spindle and stellate to pleomorphic sarcomatoid cells (Fig. 4). Mitoses, including atypical forms, were abundant. Numerous and readily apparent PAS-positive diastase-resistant eosinophilic globules were present (Fig. 5). Immunohistochemical staining of tumor cells revealed positivity for bcl-2, CD10, CD68, desmin, alpha-1 antitrypsin, glypican-3, and vimentin. These finding were consistent with the diagnosis of UESL. Negative staining for ALK-1, CD34, CD117, pan-cytokeratin, HepPar-1, myogenin, and S-100 excluded other entities such as hepatoblastoma, rhabdomyosarcoma, gastrointestinal stromal tumor, anaplastic large cell lymphoma, angiosarcoma, other carcinomas, and melanoma [1,2].

Discussion

A review of the literature demonstrates a paucity of reports in describing imaging characteristics of UESL. Furthermore, given the undifferentiated nature of this mesenchymal sar-

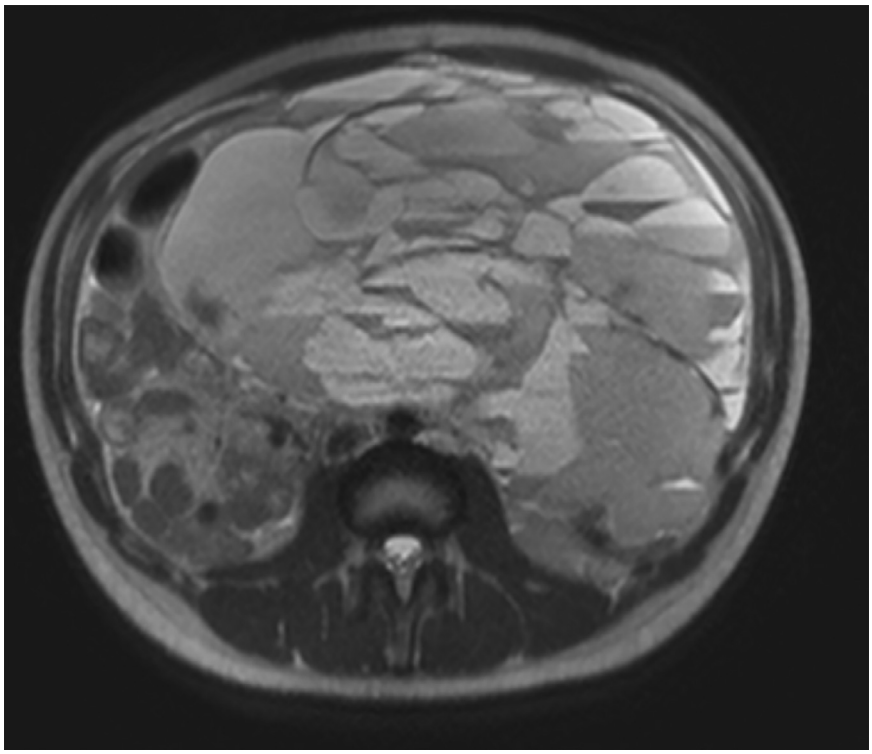

Fig. 2 – T2-weighted imaging MRI of the abdomen in this patient with undifferentiated embryonal sarcoma of the liver demonstrating multiple fluid levels.

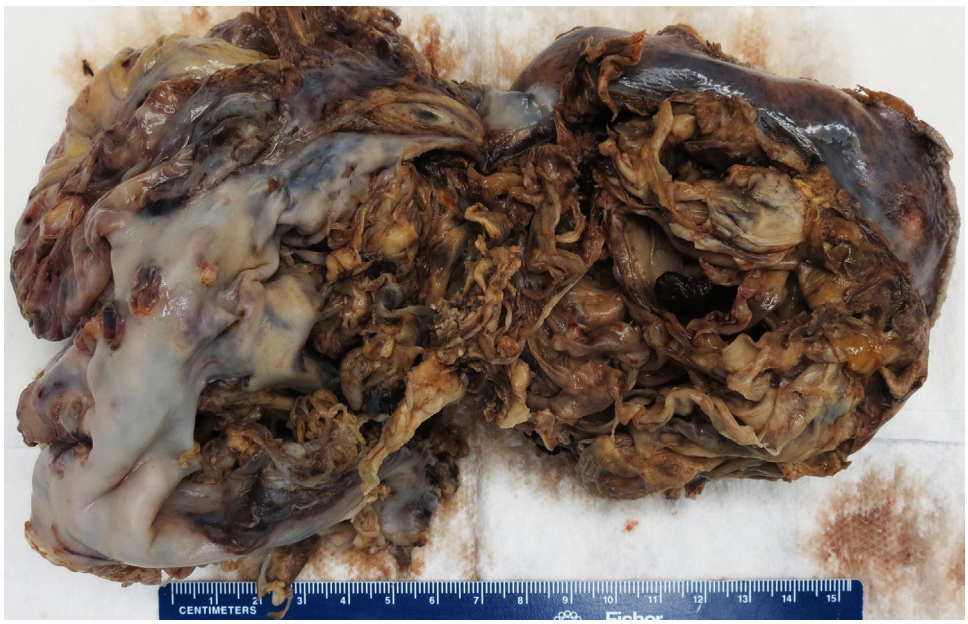

Fig. 3 – Cut surface of the partially collapsed and predominantly multicystic tumor mass. Some cysts contain clotted blood.

coma, characterizing imaging findings of typical UESL becomes challenging due to varied constituents of the tumor and resultant varied imaging appearance.

On ultrasonography, UESL typically appears as a solid mass that ranges from hypo- to hyperechoic compared to normal liver tissue with small anechoic areas. These anechoic areas correspond to focal hemorrhage, necrosis, and cystic degeneration [5]. In contrast, UESL typically appears as a hypodense cystic mass with internal septations in CT scans. A pseudocapsule may also be observed as an enhancing peripheral rim

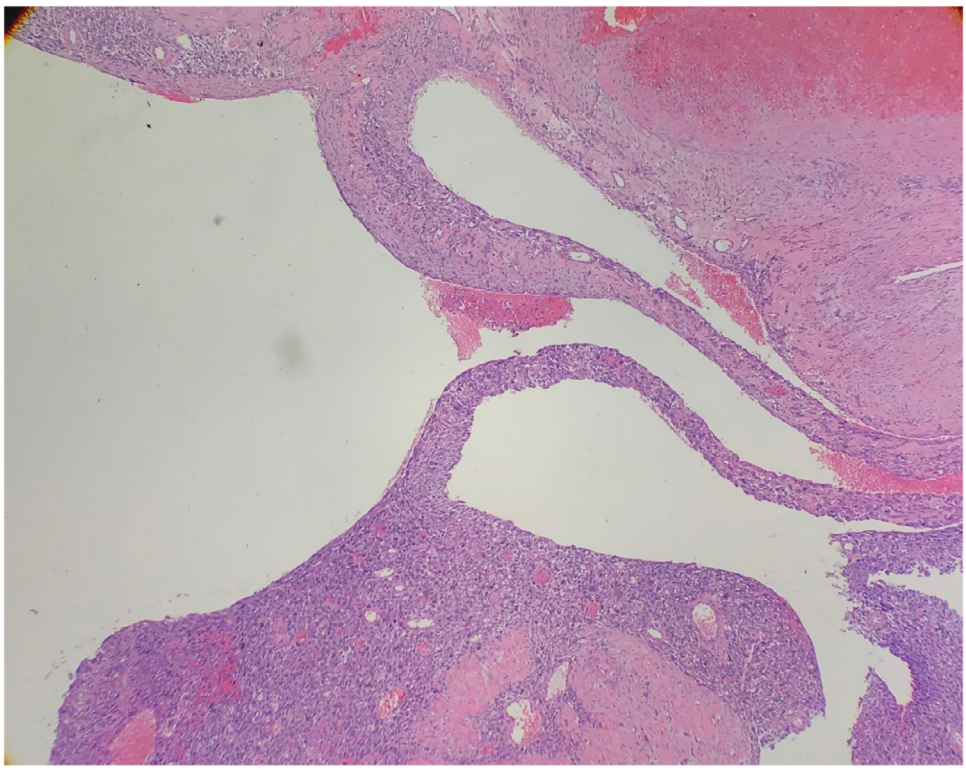

Fig. 4 – Cyst walls composed of malignant neoplastic cells. The cysts are devoid of any epithelial lining (H and E staining, low magnification).

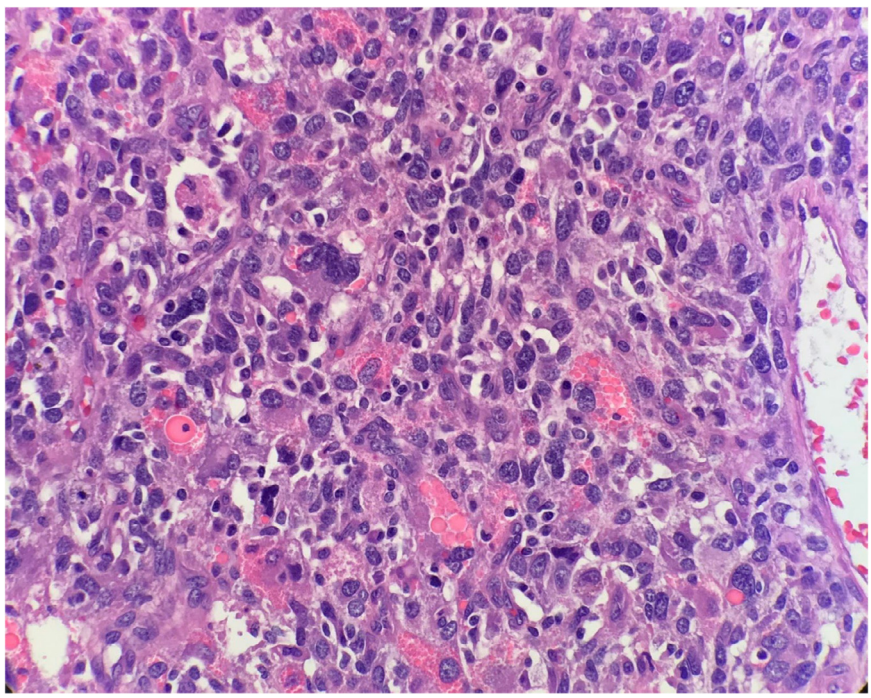

Fig. 5 – Solid area with sheets of hypercellular tumor containing atypical sarcomatoid cells as well as pleomorphic multinucleated tumor cells. Mitoses are abundant and eosinophilic globules are easily found (H and E staining, high magnification).

with high attenuation [6]. Areas of hemorrhage or necrosis which appear with the same attenuation as muscle may appear, although calcifications are relatively uncommon [6]. The cystic appearance of UESL on CT can lead to radiological misinterpretation as a benign liver lesion which is further supported by a normal AFP level.

Reports of the MRI appearance of UESL are scarce. In 1 report, UESL appeared to have the same intensity as cerebrospinal fluid on T1- and T2-weighted imaging [5]. The tumor commonly presents with a central area of hypointensity on T1-weighted images and hyperintensity on T2-weighted images which correspond to an area of focal hemorrhage and necrosis on gross pathology [7]. Internal debris and septations are better visualized on T2-weighted images. Compared to CT imaging, MR imaging is superior for visualizing areas of hemorrhage and necrosis as well as determining resectability, local invasion and involvement of adjacent venolymphatic structures and the biliary tree [5].

An appearance of UESL consists entirely of fluid-fluid levels, characteristic of venolymphatic malformations, as in our case has not been described previously in literature. Venolymphatic malformations classically have fluid-fluid levels on MRI, secondary to hemorrhage and lymphatic debris [8]. The description of peripheral enhancement of the cystic lesion and intrinsic enhancement of the adjacent abnormal T2 hyperintense soft-tissue lesion on T1 fat-saturated postcontrast image describes the findings of this patient MRI findings.

Laboratory data is often obtained to aid in formulating a differential diagnosis for liver masses, but AFP and CEA levels are typically normal in patients with UESL. Pathologic diagnosis is the gold standard of UESL and is made with the aid of immunohistochemistry. The combination of positive immunostaining results (eg, CD68, vimentin, etc.) and negative results (eg, myogenin, pancytokeratin, etc.), as seen in our case, is key to the diagnosis of this undifferentiated neoplasm.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2019.04.005.

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