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Undifferentiated embryonal sarcoma of the liver: US and CT findings

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Abstract. Six cases of undifferentiated embryonal sarcoma (UES) were reviewed to determine their characteristic features on ultrasonography (US) ($n = 5$) and computed tomography (CT) ($n = 6$). US demonstrated a single large, echogenic mass with some anechoic spaces. Contrast-enhanced CT scan revealed a well-demarcated low-attenuation mass with hyperdense septations of variable shape and thickness. Discrepancy of internal architecture on US and CT was one of the important characteristics of UES. CT numbers were 25–47 HU in low-attenuation areas. Enhancing peripheral rim was found in four cases and some solid portions at the periphery or adjacent to the septa were found in all cases. Two patients who had follow-up US and CT without treatment showed enhancing solid portions, changing to hypodense as the tumor grew. When compared with the pathologic findings, US showed a more accurate representation of internal architecture than did CT. Familiarity with these US and CT findings of UES of the liver will be helpful in the differential diagnosis of primary hepatic tumors in childhood.

Undifferentiated embryonal sarcoma (UES), which was most commonly known in the past as malignant mesenchymoma, is the term applied to a sarcoma of the liver without histologic differentiation [1–3]. UES, although still uncommon, was the fourth most frequent hepatic tumor in the pediatric age group after hepatoblastoma, infantile hemangioma, and hepatocellular carcinoma [2]. Approximately 90 % of cases occur in children up to the age of 15, the remaining 10 % occur between the ages of 15–30. UES occurs as a large, solitary, spherical mass that is usually located in the right lobe of the liver. Despite resection, chemotherapy, and radiation therapy, the prognosis is poor and many patients are dead within 12–24 months of the diagnosis [1–3].

Although Ros et al. [2] described the radiologic and pathologic aspects of this tumor in detail, there has been little radiologic experience reported with these tumors recently [4–7]. Herein, we present six cases of UES with emphasis on the ultrasonography (US) and computed tomography (CT) findings.

Materials and methods

We retrospectively reviewed the US and CT of six patients with hepatic undifferentiated embryonal sarcoma seen between 1985–1992 (Table 1). There were 5 children (3 boys and 2 girls) aged 9–12 years old, and one adult aged 23. The diagnosis was established by surgery in all six cases. In four cases total removal of the mass was possible, allowing radiologic-pathologic correlation. In two patients, 3 and 4 weeks' follow-up US and CT were performed without any treatment.

US was performed in five patients with a Diasonics DRF 400, with a 3.5-MHz and/or a 5.0-MHz transducer. CT was performed in all six patients with a GE CT/T 9800 or 8800 scanner (General Electric Medical Systems, Milwaukee, Wisc., USA) with contiguous 10-mm sections after intravenous injection of 76 % ioxithalamate meglumine and sodium (Telebrix-38, Laboratoire Guerbet, Aulnay-sous-Bois, France).

Results

Patients presented with an abdominal mass with or without abdominal pain, weight loss, and gastrointestinal complaints. Fever, probably due to hemorrhage and necrosis, was prominent in two patients. In none of the six cases was there jaundice at presentation. The duration of symptoms was from a few days to 1 month. Laboratory data included a mild leukocytosis and anemia in three patients and elevated liver enzymes in three. Alpha-fetoprotein level was checked in four patients and was normal.

Abdominal plain radiographs demonstrated a large mass in the right upper quadrant in five patients and in

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Table 1. US and CT findings of undifferentiated embryonal sarcoma of the liver in six patients

Patient no.	Age (years)	Sex	Location of tumor	Size (cm)	Radiologic findings				
					US		CT		
					Echo pattern	Anechoic area	Attenuation[a]	Solid portion	Enhancing rim
1	12	F	right lobe	12 × 18	echogenic	+	low (30)	+	+
2[b]	10	M	right lobe	7 × 11	echogenic	+[c]	low (26)	+	+
3[b]	10	F	both lobes	13 × 18	not performed		mixed	+[d]	−
4	10	M	both lobes	12 × 16	echogenic	+	low (25)	+	+
5	9	F	left lobe	6 × 7	echogenic	+[c]	low (47)	+	+
6	23	M	right lobe	6 × 8	echogenic	+	low (32)	+	−

[a] The number in parentheses is the mean CT number of the mass in HU

[b] Follow-up US and CT performed

[c] Echogenic nodules seen in anechoic spaces

[d] Pathologically a predominantly solid tumor

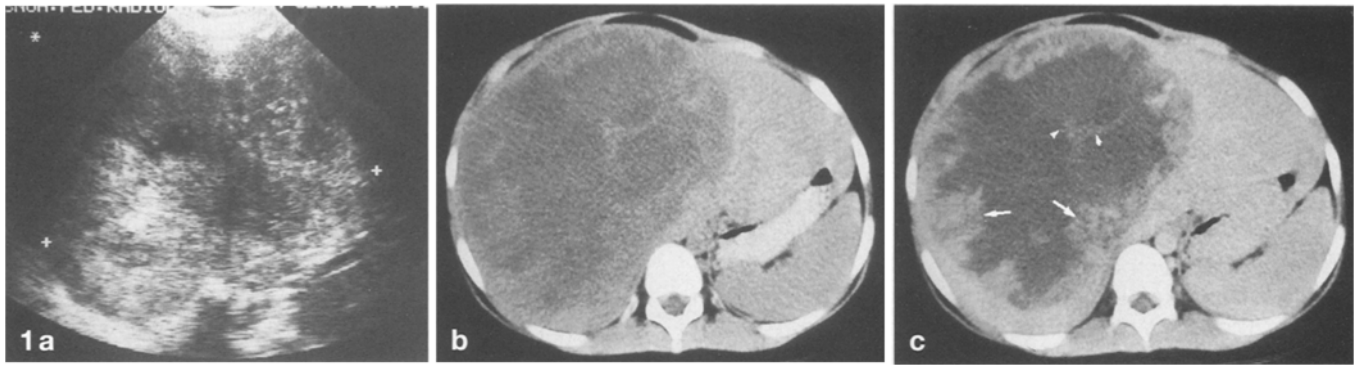

Fig. 1 a–c. *Case 1.*
a US shows a large, predominantly echogenic hepatic mass with multiple anechoic areas.
b, c Precontrast (**b**) and postcontrast (**c**) CT scans show a well-circumscribed predominantly low attenuation mass with a few septa *(arrowheads)* and peripheral solid portions *(arrows)*

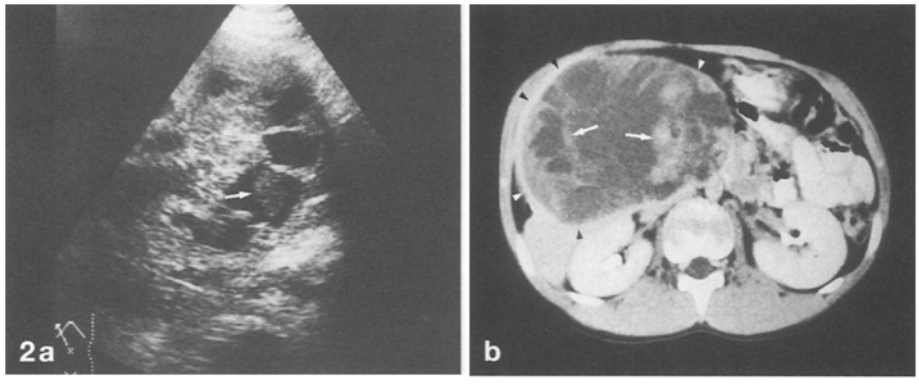

Fig. 2 a, b. *Case 2.*
a US with oblique scan shows a large, predominantly echogenic hepatic mass with multiple small anechoic spaces. Note echogenic nodules within the anechoic spaces *(arrow)*.
b Postcontrast CT scan shows a well-circumscribed low-attenuation mass with ill-defined or linear high-attenuation areas *(arrows)* and enhancing thin peripheral rim *(arrowheads)*

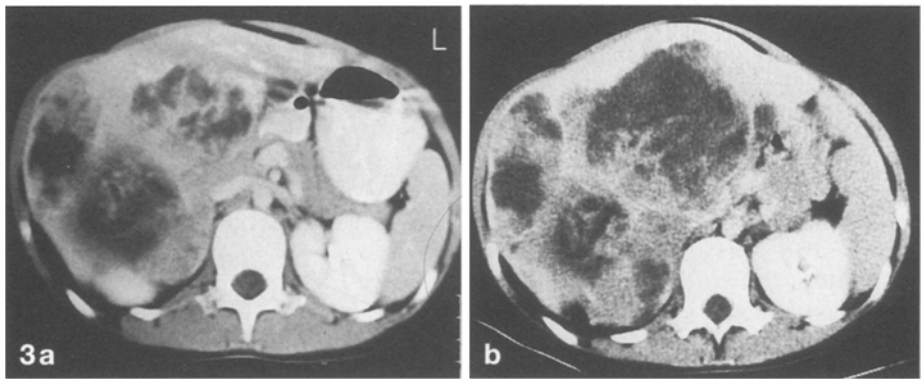

Fig. 3 a, b. *Case 3.*
a Postcontrast CT scan shows a large right hepatic mass extending to the left lobe. The liver bulges due to the size of the mass. The tumor has multifocal low-attenuation areas with intervening enhancing areas.
b Follow-up CT obtained 15 days later, without any treatment, shows increased tumor size and enlarged cystic portion of the tumoral mass

the left upper quadrant in one. Calcifications were not detected. Chest radiographs revealed elevation of the right hemidiaphragm in three patients.

US showed in each case a large, well-defined mixed echogenic hepatic mass, with multiple small anechoic spaces (Figs. 1, 2), measuring from 7 cm to 18 cm in greatest dimension (mean 13 cm). The mass was confined to the right lobe in three patients, to the left lobe in one, and grew from the right into the left in two (Fig. 3). Multiple lesions were not found. In all cases, the mass was predominantly echogenic. Anechoic or hypoechoic areas were variable in size and shape. Well-de-

fined round cystic areas were found in three cases (Fig. 2). In two cases, echogenic nodules were found within the anechoic spaces (Fig. 2 a). In one case, calcification foci were detected.

CT scan demonstrated a well-circumscribed low-attenuation mass in each case, with a few enhancing septations and peripheral solid portions (Figs. 1–3). On precontrast scan, the mass was hypodense compared with normal parenchyma and was unenhanced or only slightly enhanced (range 5–12 HU) after contrast injection. In three cases, ill-defined high-attenuation areas were found within the low-attenuation mass (Fig. 1). On postcontrast CT scan, low attenuation of the mass became more prominent. Mean attenuation values of the lesions were 25–47 HU. In two cases, punctate calcifications were found at the periphery of the mass. Septations were seen in five cases and varied in number and size (Fig. 1). Solid portions were found in all cases. They were ill-defined and found at the periphery of the mass and/or adjacent to the septum. CT numbers were 60–72 HU. In four cases, the tumor was surrounded by an enhancing thin rim (Fig. 2 b, 3 a) 2–5 mm across. On precontrast scan this was isodense with, or slightly hypodense to the adjacent liver parenchyma. The rim was especially well delineated in the mass with very low attenuation.

The tumors grew rapidly. In the two patients who each had 3 and 4 weeks' follow-up US and CT, an increase in the size of the tumor and the larger, cystic portion was seen. In one case, pulmonary metastasis and tumor growth into the inferior vena cava was found.

Radiologic-pathologic correlation was done in four cases. Macroscopically, the tumors were large, well defined spherical masses with multiple cystic spaces. There was sharp demarcation between the tumor margin and adjacent normal liver. In four cases, thin fibrous pseudocapsule was found. The cut surface of resected masses had a variegated, predominantly yellowish to tan, glistening appearance and had cystic areas of variable size that contained necrotic debris, hemorrhagic fluid, clotted blood, and gelatinous material. Multiple solid portions, found at the periphery of the mass and/or adjacent to the septation, had viable tumor tissues.

Discussion

The histologic pattern of UES is that of a rapidly growing, undifferentiated tumor with frequent mitosis [1]. This rapid growth is probably responsible for the areas of necrosis and subsequent cystic degeneration. In two of our series, increase of the tumor size was detected even at 3 or 4 weeks' follow-up. UESs are occasionally unresectable at presentation. Tumor growth into the inferior vena cava and right atrium has also been noted [8]. Metastases usually go to the lungs and skeleton. In our series, pulmonary metastasis and tumor growth into the inferior vena cava was found in one case. Treatment is by surgical excision when feasible. Prognosis of UES is poor, with a median survival time of less than 1 year. It is, however, important to achieve an accurate and early diagnosis of UES since aggressive, combined surgical and chemotherapeutic treatment may increase survival time. Preoperative chemotherapy has been reported to allow complete resection of a UES previously considered inoperable [9, 10].

Pathologically, the lesions can range from solid to primarily cystic and the imaging features correspond. On US, UES appears as a large intrahepatic mass which may be predominantly echogenic with many small anechoic spaces, or may be cystic. In all of our series, the appearance was of a predominantly echogenic mass with anechoic spaces of various shapes. In two cases, echogenic nodules were found within the anechoic spaces. A cystic mass with echogenic, polypoid areas arising from its wall has also been seen [2]. On CT scans, UES appears as a large intrahepatic mass that has lower attenuation values than the surrounding liver. The abundant myxoid matrix of the tumor may be the cause of the hypodense appearance on CT scans. On US, UES usually appears as an echogenic solid mass, but on CT, it shows as a well-circumscribed, very low attenuation mass of cystic appearance, suggestive of a benign lesion.

When used to examine pathologic specimens, US gives a more accurate representation of internal architecture than does CT. CT can depict a dense, peripheral, enhancing thin rim of tissue that corresponds to the fibrous pseudocapsule seen on gross pathologic examination. Multiple solid portions found at the periphery of the mass and/or adjacent to the septation have been found to have viable tumor tissues.

The differential diagnoses include mesenchymal hamartoma, hepatoblastoma, hydatid cyst, infantile hemangioendothelioma, hepatocellular carcinoma and metastasis [2, 4, 11]. Differentiation may be possible based on the patient's age, alpha-fetoprotein levels and radiologic findings. The typical presentation of UES in older children or young adults does not resemble that of hepatoblastoma or infantile hemangioendothelioma. Metastasis and hepatocellular carcinoma are unlikely to be predominantly cystic or to contain small cystic spaces within a solid mass. Alpha-fetoprotein levels are not elevated in UES [9, 10], while they may be elevated in hepatoblastomas and hepatocellular carcinomas. Radiologic findings do not easily permit differentiation from mesenchymal hamartoma since both tumors have variable degrees of cystic change [11–13], but in our experience, UES is more solid. The rarity of UES in patients less than 5 years of age helps to distinguish it from mesenchymal hamartoma of the liver, which usually presents between the ages of 4 months–2 years.

Imaging findings of UES were quite characteristic. US demonstrated a single large, echogenic mass with some anechoic spaces and CT scan revealed a well-demarcated low-attenuation mass with hyperdense septations of variable shape and peripheral solid portions. Enhancing peripheral rim, representing fibrous pseudocapsule of the tumor, was occasionally found. Discrepancy of internal architecture on US and CT was one of the important characteristics of UES. We conclude that US and CT findings of UES are characteristic and familiarity with these findings will be helpful for differential diagnosis of primary hepatic tumors in childhood.

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