Undifferentiated Embryonal Sarcoma of the Liver: Report of 2 Cases

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ABSTRACT

Undifferentiated embryonal sarcoma (UES), also known as undifferentiated liver sarcoma (ULS) is a rare, primary malignant neoplasm of the liver, affecting mostly pediatrics and young adults.

Clinical symptoms and plain radiographs are usually nonspecific. Sonographic, CT and/or MRI findings are more sensitive and can help to differentiate this tumor from other conditions.

The differential diagnosis includes hydatid cyst, cystic hepatoblastoma, mesenchymal harmatoma and biliary neoplasm.

In this illustration, we would like to report 2 patients with UES who were 9 year-old and 13 year-old girls. Radiologic and pathologic imaging will be discussed.

Keywords: Undifferentiated embryonal sarcoma, liver mass, liver tumor

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INTRODUCTION

Undifferentiated embryonal sarcoma (UES), also known as undifferentiated liver sarcoma (ULS) is a rare, primary malignant neoplasm of the liver, affecting mostly pediatric patients and young adults. It is the third most common malignant hepatic tumor in children,following hepatoblastoma and hepatocellular carcinoma. This tumor was first described by Stocker and Ishak in 1978.[1]

We report two cases of patients with UES who were 9 and 13 year old girls. Radiologic and pathologic findings will be discussed.

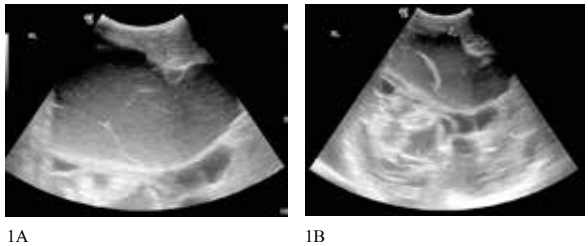
CASE 1

A 9 year-old girl presented with gradual enlargement of the right side of the abdomen over approximately one month. She also experienced early satiety and weight loss. On physical examination, the patient had no jaundice. There was a palpable mass in the right upper quadrant of the abdomen, approximately 10 cm below the right costal margin. The liver span was estimated at 15 cm.

The complete blood count was normal. Liver function tests were within normal limits other than for a mildly elevated alkaline phosphatase (167 units). Serum alpha-fetoprotein (AFP) was 1.13 IU/ml.

Ultrasound of the abdomen demonstrated a mixed solid and cystic mass lesion within the right upper abdomen, probably arising from the liver. (Fig 1A, B) The right kidney appeared normal.

CT scan showed a large hypodense mass within the right lobe of the liver. There were multiple internal septations and there was peripheral and septal enhancement.



1A 1B

Fig 1A and B. Ultrasound of the upper abdomen demonstrates a large mixed solid and cystic lesion in the right upper quadrant of the abdomen.

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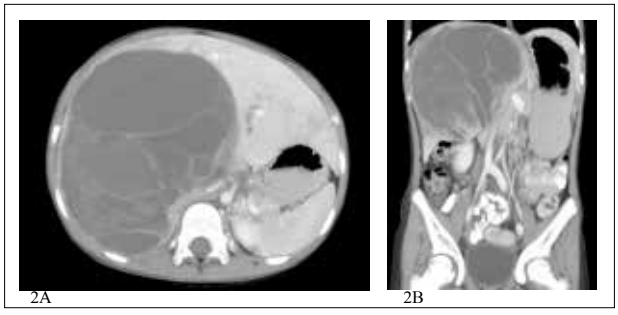

Fig 2A and B. Axial and coronal reformatted CT images of the abdomen demonstrate a large hypodense mass in the right lobe of the liver. Internal septations were present. Minimal peripheral and septal enhancement was seen.

No calcification was seen within the mass. Findings were likely consistent with UES of the liver, given the age of the patient and amount of solid appearance on sonographic findings. (Fig 2A, B)

The patient had an extended right hepatectomy. Gross pathology of the tumor revealed an encapsulated mass with multiloculated cysts, varying in size from 5 mm -10 cm, containing hemorrhagic products. No solid components were found. The final pathologic diagnosis was UES of the liver. The patient had post-operative chemotherapy and at the most recent follow up was doing well.

CASE 2

A 13 year-old female presented to a local hospital with fever, abdominal pain and distention. The patient underwent an abdominal CT scan, which showed a large cystic mass in the liver. An initial diagnosis of a liver abscess was made. The lesion was aspirated and bloody contents were found. The patient was then referred to our hospital. Basic laboratory results were normal other than for a mildly increased alkaline phosphatase (207 units). Serum AFP was 0.67 IU/ml. Hepatitis serologies were negative.

A plain radiograph demonstrated a large soft tissue mass in the right upper quadrant of the abdomen. Bowel gas was displaced to the left side. No abnormal calcifications were noted. Bony structures were normal. The patient underwent a CT scan which demonstrated a large hypodense mass in the right lobe of the liver. A few areas of mild hyperdensity were seen within the mass. No internal calcification was detected. (Fig 3A, B) The presumptive diagnosis of UES was made. The patient underwent surgery with tumor removal. The patient was doing well after surgery at the most recent follow up visit.

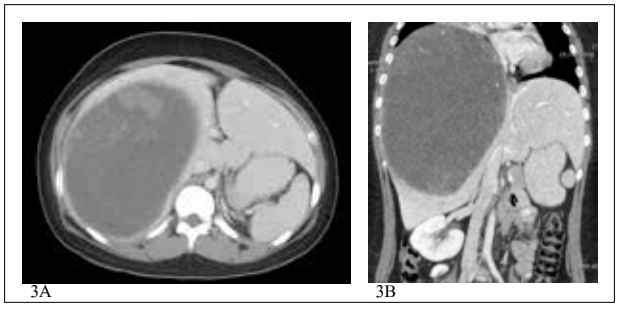

Fig 3A and B. Axial and coronal reformatted CT images of the abdomen demonstrate a large hypodense mass in the right lobe of the liver. Focal hyperdensity was noted inside the mass.

TABLE 1. The immunohistochemical findings.

Antibody	Expression	
	Case 1	Case 2
Vimentin	+ diffuse	+ diffuse
AE1/AE3	n.a.	+ faint focal
HePar1	-	n.a.
S-100 protein	-	-
SMA	+ faint focal	-
Desmin	+ faint focal	+ faint focal
HHF-35	na	-
Myogenin	na	-
Factor VIII	na	-
CD31	+ focal	n.a.
CD34	+ faint focal	-
CD68	+ focal	n.a.
CD117	n.a.	-

Note: + positive, - negative, n.a. not applicable, SMA smooth muscle actin

Pathologic findings

Microscopically, these tumors were composed of compact and loosely arranged spindles, and stellate and bizarre multinucleated giant cells. They showed moderate to marked nuclear pleomorphism. The nuclear chromatin varied from hyperchromatic to vesicular. Mitoses and atypical mitoses were common in both cases. Eosinophilic globules were present within tumor cells and extracellularly. These globules were PAS positive.

Immunohistochemical studies have been summarized in Table 1. Most tumor cells in both cases expressed vimentin. UES was diagnosed in both cases. Microscopic findings from both cases have been shown. (Fig 4 and 5 A, B)

DISCUSSION

Undifferentiated embryonal sarcoma (UES) of the liver is a malignant neoplasm of mesenchymal origin. It represents about 9-15% of all hepatic tumors in children, with approximately 150 cases reported in the literature.[2] UES of the liver is most commonly seen in children, with the peak incidence between the ages of 6-10 years, but also occurs in adults.[1,3]

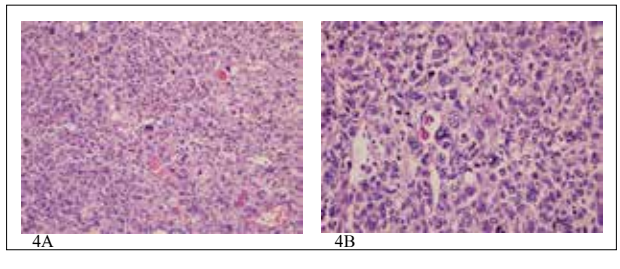

Fig 4A and B. Histologic findings from Case 1
A. Tumor cells exhibit marked nuclear pleomorphism. Atypical mitoses are common. (H&E, 200x)
B. Eosinophilic globules of varied size are present within tumor cells and in the extracellular matrix. (H&E, 400x)

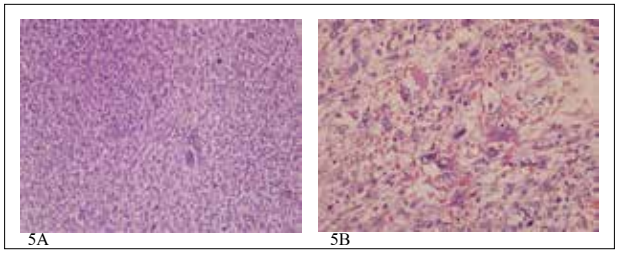

Fig 5A and B. Histologic findings from Case 2
A. Mitotically active atypical spindle cells and multinucleated giant cells. (H&E, 200x)
B. Higher magnification illustrates multinucleated tumor giant cells containing eosinophilic globules of varied size in a loose edematous myxoid matrix. (H&E, 400x)

Patients usually present with an abdominal mass and/or abdominal pain. Other symptoms include fever, weight loss, anorexia, malaise, nausea and vomiting. Diagnosis of UES is challenging because the presenting symptoms are nonspecific, no serological markers exist, and the tumor is rare. Our patients presented with abdominal masses and abdominal distention, and palpable masses were discovered on physical examination.

Plain radiographs demonstrate a large soft tissue mass in the right upper quadrant of the abdomen. The sonographic appearance varies from a multiseptated cystic mass with or without mural echogenic nodules to a predominantly echogenic mass.[4-5] This is in contrast to CT and MRI, which typically demonstrate a large mass with largely cystic attenuation. The majority of these tumors are hypointense on T1W and hyperintense on T2W images.[3] It has been theorized that the cystic appearance of this tumor is related to abundant myxoidfibroma.[6] The discrepancy in the appearance of the internal architecture between sonography and CT has been described as an important identifying characteristic of UES.[7-8]

Macroscopically, UES is usually a large, well-defined mass. The cut surface has a variegated, predominantly yellowish to tan, glistening appearance and often has cystic areas with necrotic debris, hemorrhagic fluid or gelatinous materials.[4]

Differential diagnoses include hepatocellular carcinoma (HCC), hydatid cyst, liver abscess and mesenchymalharmatoma. HCC is the most common primary malignant hepatic tumor in children over 5 years old. It usually occurs in association with chronic liver disease such as cirrhosis, and serum alpha fetoprotein is usually elevated. However, HCCs do not usually present as a multiloculated cystic mass.

Hydatid disease is one of the main differential considerations for UES. Prior reports have described UES as resembling hydatid cysts.[3,9] Hydatid cysts are common in children living in endemic areas. These lesions can be seen on ultrasound and CT as unilocular or multilocular cystic lesions. Daughter cysts can be demonstrated within a dominant cyst. Mesenchymalharmatoma may be difficult to radiographically differentiate from UES. However, these are usually found in younger age groups, with the peak incidence occurring at approximately 18-24 months of age.

Although there is no standard treatment, surgery and chemotherapy remain the most common means of therapy. Positive resection margins and iatrogenic rupture of the tumor are associated with early recurrence and death.[10] Local recurrence and distant metastases are common, especially to the peritoneum, pleura and lungs.

As there is no serum marker to predict recurrence, post-treatment imaging surveillance, with abdominal ultrasound or CT examination is useful.

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