Undifferentiated Embryonal Sarcoma of the Liver in Adult Mimicking Hepatic Abscess

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ABSTRACT

Undifferentiated embryonal sarcoma is an uncommon malignant tumor of liver of mesenchymal cell origin, usually observed in children. We report a case in a 34 years old male who presented with right upper quadrant pain. Ultrasonography showed a complex solid cystic lesion. However, on computed tomography it appeared as predominantly cystic with small solid components within. It was initially diagnosed as hepatic abscess. After failure to improve on conservative treatment, the patient underwent surgical resection and histopathology revealed undifferentiated embryonal sarcoma. Here we describe the typical clinical and radiologic features of this rare tumor.

KEY WORDS

Adult, CT scan, Undifferentiated embryonal sarcoma of liver, Ultrasonography

INTRODUCTION

Undifferentiated embryonal sarcomas of the liver (UESL) are rare, aggressive, and malignant liver tumors seen in the pediatric population, mostly in children. UESL is a very rare neoplasm and even rarer in adult age group.[1] Cases in adults generally present once a large mass develops and may be mistaken for other tumor or abscess.[2] The diagnosis is difficult because of the similarity in epidemiological, clinical, and radiological findings with those of other liver tumors.[3,4] Since the clinical and radiological findings are often nonspecific, the diagnosis of UESL is based on its histology and immunophenotyping. In the current study, we present a case of UESL which clinically and on initial investigations mimicked hepatic abscess.

CASE REPORT

A 34 years old male was admitted to the Department of Surgery, BPKIHS, Dharan on 4th of May, 2017 with complain of fever along with pain and swelling in right upper abdomen for 2 weeks. There was a palpable mass on physical examination in right hypochondriac region. White blood cell (WBC) count was elevated (14.2 x 109/L). Blood culture was sent which showed no growth. Alfa-fetoprotein (AFP) on lab analysis was normal. On ultrasonography (USG), there was a hypoechoic lesion with predominant solid component in left lobe and also partly involving medial segments of right lobe of liver (Fig. 1). It was initially diagnosed as a case of liver abscess. On ultrasound guided aspiration from the lesion, blood mixed aspirate was

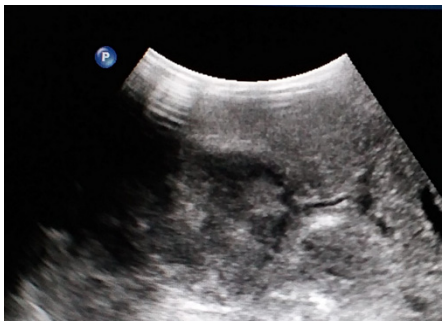

Figure 1. USG abdomen in a case UESL showing a heterogeneous predominantly hypoechoic mass in left lobe of liver

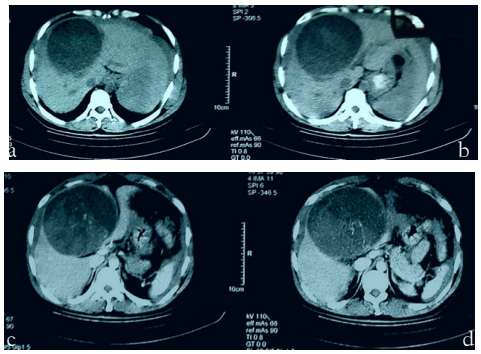

Figure 2. Axial NCCT (a,b) and CECT (c,d) images of a case of UESL. The tumor is located predominantly in left lobe of liver with enhancing peripheral and solid component and large non enhancing necrotic component within, mimicking an abscess.

detected. On suspicion of alternate diagnosis, contrast enhanced Computed tomography (CT) was done on next day which showed hypodense lesion of size ~7 x 6 cm in left lobe and partly extending into the medial segments of right lobe (Fig. 2). The mass showed peripheral and few small solid areas of enhancement with predominant non enhancing necrotic area within. Compression of the adjacent biliary duct and hepatic vessels was noted. The case was reported on CT as malignant hepatic mass. Patient subsequently underwent open laparotomy with resection of the mass. The postoperative histopathological diagnosis was undifferentiated embryonal sarcoma of liver (Fig. 3).

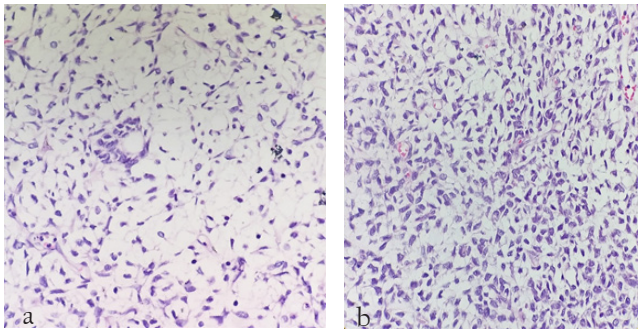

Figure 3 a. Hematoxylin and eosin stained section showing hypocellular area of tumor with entrapment of bile duct (200X). b. Hematoxylin and eosin stained section showing proliferation of spindle to stellate shaped tumor cells (400x)

DISCUSSION

UESL is a rare hepatic tumor of mesenchymal origin that was first described by Stocker et al. in 1978.[2] UESL occurs almost exclusively in children, with peak incidence in age range of 6 to 10 years. There is no gender predilection. UESL is an extremely rare neoplasm in adulthood as seen in our case.[5,6] Review of the various literatures showed that there were only 70 cases of UES in adults reported worldwide till 2008.[7] The tumor is mostly located in right lobe of liver while rarely it occurs in left lobe, as noted in our case. Hemorrhage and necrosis along with cystic changes are frequently observed, while clinical manifestations include abdominal mass, pain and fever mimicking liver abscess.[8]

All the clinical and laboratory findings are nonspecific. The combination of presenting symptoms like fever, patient's age and elevated blood counts and normal AFP levels can point towards infective pathology. The fever is likely due to the hemorrhage and necrosis occurring within the tumor, as seen in our case, which may lead to preliminary diagnosis of an underlying infective process.[9]

USG predominantly shows solid nature of the tumor while CT findings can resemble those of cystic lesions. The predominant cystic appearance on CT is related to the high water content of the myxoid stroma within the tumor. So findings of a large hepatic lesion with predominantly cystic appearance on CT or Magnetic Resonance Imaging (MRI) images and a paradoxically solid appearance on USG are highly suggestive of this tumour.[9,10] Similar paradoxical USG and CT findings were also noted in present case.

A definite diagnosis of UESL can rarely be determined preoperatively. The diagnosis depends on postoperative pathological findings and immune-histopathological results. UESL must be differentiated from hepatic abscess, hepatocellular carcinoma and hydatid cyst. Clinical history combined with imaging findings of USG and CT has been beneficial for the differentiating UESL from other lesions of liver. Previous studies have shown that UESL is often misdiagnosed as liver abscess or hydatid cyst in view of the history of fever and raised serum inflammatory markers, however, cultures and serology tests were negative for hydatid cyst and abscess.[2,8,9,11]

UESL can be difficult to diagnose and both the clinical features and imaging findings of this rare tumor may be misleading. Any discordance between CT and USG images findings, that is if the lesion appears solid in USG but predominantly cystic on CT images, one should suspect the possibility of UES as the diagnosis. Since the clinical and radiological findings are often nonspecific, the diagnosis of UESL depends upon the histologic examination and immunologic evaluation. Early and prompt diagnosis and therapy are important for better prognosis and long term survival of the patient.

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