

Child with hepatic hydatid cyst: A diagnostic uncertainty

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ABSTRACT

Many parts of India are endemic for hydatid cyst, and hence, the most common diagnosis of a hepatic cyst in such regions is hydatid cyst. Undifferentiated embryonal sarcoma of the liver (UESL) is a rare differential diagnosis for hepatic hydatid cyst as the clinical and radiological features of hydatid cysts and UESL overlap. Here, we report a 4-year-old boy with hepatic cyst, who was initially diagnosed as hepatic hydatid cyst, which was later confirmed as UESL. The child was successfully treated with chemotherapy. This case report highlights the need to spread awareness about UESL, as prompt diagnosis and treatment improves prognosis of the same.

Key words: *Hydatid Cyst, Undifferentiated embryonal sarcoma of the liver, Hepatic cyst, Differential diagnosis*

Hepatic hydatid cyst though more common in adults can affect children also [1]. In India, hydatid disease (HD) is endemic in Andhra Pradesh, Tamil Nadu, and Rajasthan [2]. In such regions, the most common diagnosis of any hepatic cyst is hydatid cyst. However, one should be aware of undifferentiated embryonal sarcoma of the liver (UESL) as a rare differential diagnosis (DD) for cystic hepatic lesions. UESL is the third most common malignant tumor affecting the liver and accounts for 13% of hepatic malignancies in children [3]. Here, we describe a child diagnosed with hepatic hydatid cyst but were later proved to have UESL and were successfully treated.

CASE REPORT

A 4-year-old Indian boy from lower socioeconomic status presented with fever, jaundice, and pain abdomen for 1 month. Clinically, he was icteric with tender hepatomegaly; his blood investigations revealed elevated serum bilirubin. The complete blood count, liver enzymes, alkaline phosphate (ALP), and lactate dehydrogenase (LDH) were found to be normal. Ultrasonography (USG) abdomen showed multicystic mass with variable solid matrix while the computed tomography (CT) of the abdomen revealed hypodense encapsulated mass in the right lobe, suggestive of hydatid cyst (Fig. 1).

As he did not respond to Albendazole, cystopericystectomy was planned. Intraoperatively, the lesion was found to be solid and frozen section examination done was suggestive of sarcoma; hence, the right hepatectomy was done. Histopathological examination of the resected sample showed pleomorphic

spindle-shaped cells with high mitotic rate, suggestive of UESL (Fig. 2).

Further, immunohistochemistry (IHC) was positive for vimentin, CD 10, and CD 68. Fluorodeoxyglucose positron emission tomography scan was negative for metastasis (Fig. 3).

He received adjuvant chemotherapy with injection vincristine, injection doxorubicin, injection cyclophosphamide, injection Ifosfamide, and injection etoposide. He is currently 3 years post-treatment and is doing well.

DISCUSSION

UESL occurs predominantly in children with the peak incidence between 6 and 10 years [4]. Similar to the child in this study, it commonly involves the right lobe of the liver [3]. The pathogenesis of UESL is not well established; it is said to be of mesenchymal origin and was first reported as a mesenchymoma. Few authors opine that UESL can arise within mesenchymal hamartomas [3,5].

Cao *et al.* reported that while ALP and LDH may be elevated, cancer antigen 125 (CA125), carcinoembryonic antigen, and CA19 are non-specific for UESL [3]. Similarly Lee *et al.* also reported that serum tumor markers are usually normal in UESL [6]. Zhang *et al.* observed that alpha-fetoprotein was occasionally elevated in UESL. They reported one case of UESL with increased CA-125 [7]. There are previous reports of UESL with peripheral eosinophilia and few authors consider UESL as a DD for eosinophilia accompanying hepatic cysts [7]. This is similar to the eosinophilia seen in HD. In our child, however, the peripheral

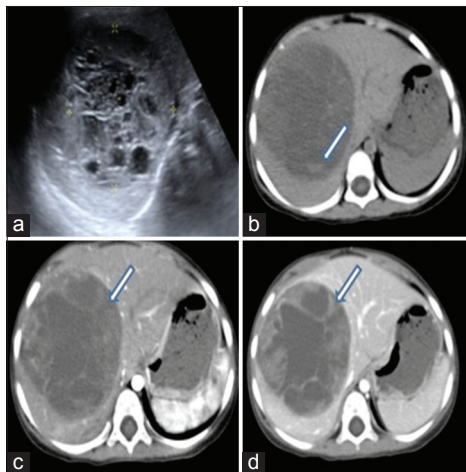


Figure 1: (a) Sonographic image of the liver reveals multicystic, multilocular mass lesion in the right lobe of liver with variable solid matrix. (b) Unenhanced axial computed tomography (CT) image of the liver reveals hypodense well-encapsulated mass in the right lobe. Few dependent hyperdense areas (arrow) are seen, which may represent debris or hemorrhagic areas. (c and d) Contrast-enhanced axial CT image in arterial phase shows peripherally enhancing cystic mass lesion with multiple peripherally intralobular cystic areas and septations in c and d (arrow). The matrix appears predominantly cystic with variable solid areas

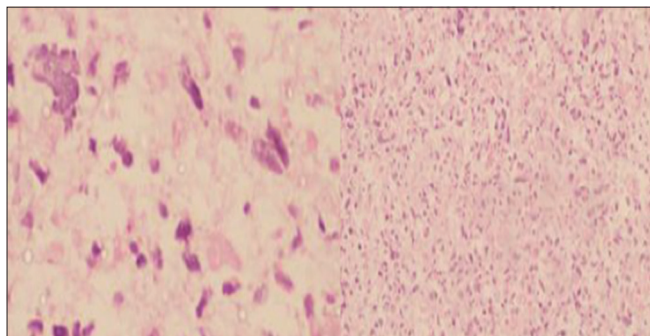


Figure 2: Histopathological features suggestive of undifferentiated embryonal sarcoma of the liver

eosinophil count was normal. The immunohistochemical markers of UESL are non-specific. As in our child, UESL is consistently positive for vimentin, other markers such as CD 68 and CD 10 show variable expression [6]. IHC is more helpful to exclude other tumors rather than to prove UESL [7].

The diagnosis of UESL is delayed in 23.5% of cases because a large cystic hepatic mass is usually suggestive of a benign lesion [4]. It is often misdiagnosed as amebic liver abscess, intrahepatic hydatid cyst, polycystic liver disease, etc. [5,7]. Faraj *et al.* described an adult with UESL who was misdiagnosed as HD [4]. Cao *et al.* described one case of UESL where the initial diagnosis was teratoma/echinococcosis [3]. Likewise, Yildiz *et al.* reported three cases of UESL, of which two were radiologically diagnosed as HD [8].

UESL is often misdiagnosed as hydatid cyst due to the overlap in the radiological features of UESL and hydatid cyst. All modalities of imaging in hydatid cyst reveal the fluid character within the cyst [1], whereas UESL appears predominantly solid in USG and cystic in CT/magnetic resonance imaging [4].

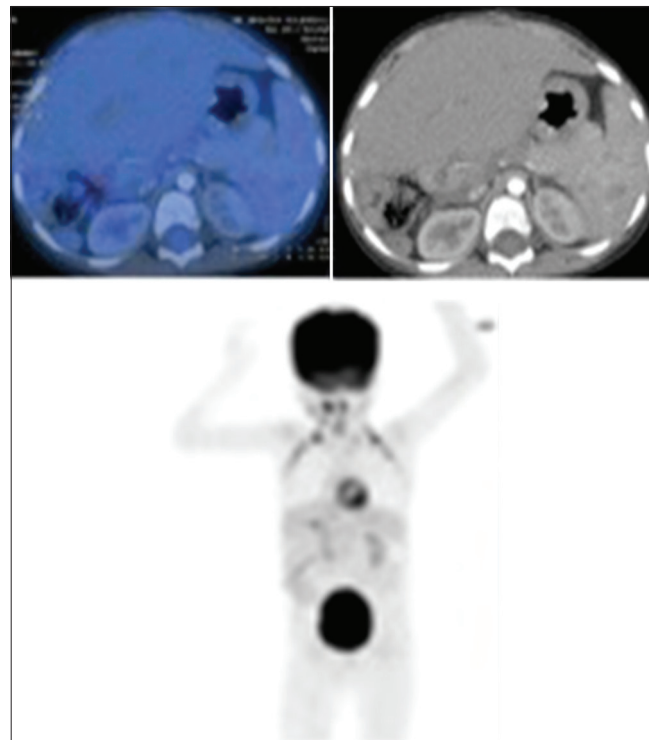


Figure 3: Fluorodeoxyglucose positron emission tomography scan showing the right lobe hepatectomy with no residual lesion and no evidence of pulmonary metastases

Therefore, if there is a difference between the USG and CT finding of a hepatic lesion, then UESL should be considered as a DD [9]. However, sometimes, both cystic and solid components may be found in USG [10]. This is similar to our child where even the ultrasound revealed cystic areas with variable solid matrix complicating the diagnosis further.

Complete resection followed by adjuvant chemotherapy is the current standard of care for UESL [4]. Hepatic transplantation is effective in unresectable tumor and post-operative recurrence [7]. Previously, the prognosis of UESL was poor with overall survival of <37.5% at 5 years [3]; however, with multimodality treatment, the prognosis has improved to 70–100% [7].

CONCLUSION

UESL must be considered as a DD in the diagnosis of a hepatic cyst, especially if there is a discrepancy between the USG and CT features. However, a definite diagnosis depends on the pathological examination of the specimen as the USG may also show a cystic lesion which was the case in our studies child.

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