

Undifferentiated embryonal sarcoma of liver in an adult masquerading as complicated hydatid cyst

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ABSTRACT

Benign or malignant hepatic lesions may rarely mimic inflammatory lesions on imaging. We describe a case of young adult male presenting with pain abdomen and fever of short duration. Imaging revealed large complex multiseptated lesion in the right lobe of liver. Complicated hydatid cyst was kept as first differential diagnosis depending on clinic-radiologic findings. However, due to few atypical imaging features, FNAC of the lesion was performed which confirmed the lesion as UESL.

Key words. Undifferentiated embryonal sarcoma. liver. Hydatid cyst.

INTRODUCTION

Undifferentiated embryonal sarcoma of liver (UESL) is a rare primary malignant tumour of liver usually presenting in the paediatric population and rarely seen in the adults. Radiologically it appears as a solid mass with or without cystic areas and can mimic complicated hydatid cyst or an abscess.^{1,2} In doubtful cases, fine needle aspiration cytology (FNAC) of the mass is problem solving to avoid inadvertent intervention.

CASE REPORT

A 24 year old male presented with pain in the right hypochondrium and low grade fever for 2 weeks. There was no history of cough, jaundice or diarrhoea. On examination, the liver was palpable 4 cms below the right costal margin and was firm and tender. Routine laboratory investigations revealed Hb 10.9 gm/dL and TLC 8,400 /mm³. Renal function tests were normal but the liver enzymes were mildly elevated. Alfa feto protein (AFP) level of the patient was 5 mg/L (normal 0-15 mg/L). The viral markers (HB-

sAg, Anti HCV) were negative. Abdominal ultrasonography of the patient showed a large, complex cystic lesion in the right lobe of the liver with multiple internal septations and low level internal echoes (Figure 1). Depending on the clinical and USG findings the possibilities of a complicated hydatid cyst and a liver abscess were kept. Hydatid serology of the patient was negative. CECT abdomen revealed a large, approximately 14 X 12 cm, well defined hypodense mass in the right lobe of liver with few enhancing internal septations (Figure 2). On MRI the mass appeared heterogeneously hyperintense on T2 weighted



Figure 1. Transverse ultrasound scan of upper abdomen showing a large, complex cystic lesion in the right lobe of the liver with multiple internal septations and low level internal echoes.

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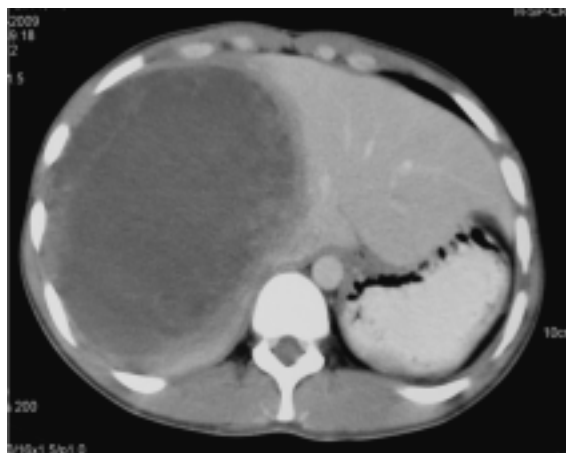


Figure 2. Contrast enhanced CT axial section of abdomen revealing well defined hypodense mass in the right lobe of liver with few enhancing internal septations.

images with hypointense septations (Figure 3A). Areas of haemorrhages were seen as T1 hyperintense areas within the lesion (Figure 3B). Post gadolinium MR images showed enhancement of the internal septations (Figure 3C). Based on the radiological appearance the diagnosis of complicated hydatid cyst was given. Neoplastic lesions like biliary cystadenocarcinoma, embryonal sarcoma and haemangiosarcoma were kept in the differential diagnosis. USG guided fine needle aspiration cytology (FNAC) was done from the lesion, which did not show any evidence of *E. histolytica* or scolices of *E. granularis*. FNAC smears of the lesion were confirmatory of UESL.

DISCUSSION

UESL is amongst the three most common malignant childhood liver tumours, 88% of cases occur below the age of 15 years.³ The usual age of presen-

tation is between 6 to 10 years with rare occurrence in the adult population.¹ UESL was first reported as a unique clinic-pathologic entity by Stanely, *et al.*⁴ Rarely it occurs in the adult population, and till now approximately 60 adult cases have been reported in the English literature to the best of our knowledge.⁵ Macroscopically it appears as a large, well-circumscribed solitary mass showing variable areas of necrosis, cystic degeneration and haemorrhage.⁶ Although there is no specific clinical feature of UESL, the patients usually present with upper abdominal mass with or without pain and weight loss. Intraperitoneal haemorrhage may occur due to spontaneous rupture of tumour. Unlike hepatocellular carcinoma, UESL is not associated with hepatitis, liver cirrhosis, and alteration of hepatic function or elevation of AFP.¹

Radiologically, UESL appears as a large, encapsulated solitary mass, often reaching a size greater than 10 cm. Abdominal radiograph may show a large soft tissue shadow in the right upper quadrant, with displacement of the bowel loops. It has a variable appearance on sonography, ranging from a complex cystic lesion with or without echogenic mural nodules to a predominantly echogenic mass.⁷ On CT, the UESL manifests as a well-defined, predominantly hypodense mass with multiple septations with or without areas of haemorrhage or necrosis. However, it may contain solid components with attenuation value of muscle. Although most of them do not show enhancement, the tumour rim and internal septations usually enhance. 50% of the tumours are hypovascular on angiography, 17% are hypervascular, and 33% are avascular.⁷ UESL shows hyperintense signal on T2 weighted MR images with low signal intensity septations.⁴

The differential diagnosis of a solitary predominantly cystic or multicystic hepatic lesion in an

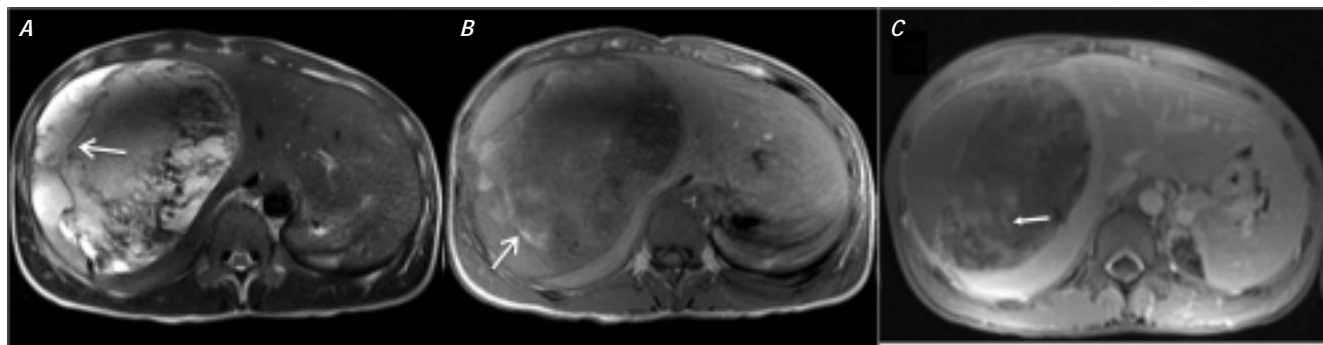


Figure 3. Axial T2 weighted (A) MRI showing heterogeneously hyperintense mass in the right lobe with hypointense internal septations (arrow). Axial T1 weighted (B) image shows areas of haemorrhages within the mass (arrow). Post gadolinium (C) images showing enhancing septations (arrow).

adult includes hydatid cyst, cavernous haemangioma, necrotic primary or metastatic tumour, and hemorrhagic hepatocellular adenoma. Although rare in the adult population, UESL has been found in patients as old as 70 years. Therefore, it should be kept in the differential diagnosis of a large solitary well-circumscribed hepatic mass in an adult.⁴ The treatment option of UESL in an adult includes combination of chemo-radiotherapy followed by radical resection of the tumour. It has got a very poor prognosis and majority of the patients die within 2 years of initial surgery due to tumour recurrence or metastasis.¹

Due to atypical presentation and non-specific imaging features, it may be difficult to differentiate neoplastic hepatic lesions from inflammatory lesions. In such cases FNAC of the lesion should be performed to avoid inadvertent delay in the diagnosis and management.

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