

Research Paper

Undifferentiated Embryonal Sarcoma of Liver Mimicking Liver Abscess: A Case Report and Review of Literature

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ABSTRACT

Undifferentiated embryonal sarcoma is a rare primary malignant tumor of the liver. It occurs almost exclusively in childhood and should be considered in the differential diagnosis of a pediatric liver mass, particularly in older children with characteristic discordant findings on ultrasonography and computed tomography. Very few cases are reported in adult age group. Here, we report a case of hepatic undifferentiated embryonal sarcoma in an adult male mimicking a liver abscess with review of literature.

KEYWORDS

Introduction:
Undifferentiated embryonal sarcoma of liver (UESL) is rare malignant mesenchymal tumor that shows no evidence of differentiation (1). Before the definition by Stocker and Ishak, it had been described under other names, such as embryonal sarcoma, malignant mesenchymoma, fibromyxosarcoma or simply sarcoma (2, 3). It accounts for 9% to 15% of all liver malignancy in pediatric age with an equal sex incidence (4); ranking fourth after hepatoblastoma, infantile haemangioendothelioma and hepatocellular carcinoma (5). Less than 60 cases of UESL in adults have been reported in the literature (1). It has characteristic discordant findings on ultrasonography (USG) and computed tomography (CT). On CT the appearance of the tumor may simulate that of an abscess or other cystic lesion which may mislead the treating physician into attempting a percutaneous drainage. On USG, however, the tumor appears as a predominantly solid mass lesion (3). Here, we report a case of UESL presenting in the adult age group mimicking an abscess.

Case report:
A twenty-year-old male was referred to us with complaints of right upper quadrant pain, low grade fever, and abdominal distension since four months. He had been diagnosed as a case of liver abscess based on his clinical features and a CT scan done outside. On examination, a tender protruding mass was palpable in the right upper quadrant. Laboratory values included mild leucocytosis; serum total protein 6.8 g/dL, serum albumin 2.3 g/dL; normal serum aspartate aminotransferase and alanine aminotransferase, elevated serum alkaline phosphatase, normal serum bilirubin and serum alfa-fetoprotein (AFP) levels. Enzyme linked immuno-sorbent assay for Echinococcal antigen (Ig G) in serum was negative. On CT scan, arterial (Figure 1a) and portal venous phase (Figure 1b) showed a well-defined, large, hypodense mass lesion predominantly of water attenuation in liver with peripheral thin septations. The mass was located in segment VIII, V and IV of the liver and was splaying the right and left hepatic arteries and displacing the gall bladder and portal vein; although the spleno-portal axis was patent. After admission, an abdominal USG followed by guided drainage of the abscess was decided. On USG (Figure 2a), the lesion appeared predominantly solid and heterogeneously hyperechoic situated in the medial segment of left lobe. An USG guided true cut needle biopsy was done (Figure 2b) from the lesion and the histopathology (Figure 3) demonstrated a tumor composed of mixed population of pleomorphic oval to elongated epithelioid and spindle cells, scattered loosely in the myxoid ground substance and hemorrhagic tissue. Near total surgical resection of the tumor was performed. On immunohistochemical analysis the tumor cells were positive for vimentin, alpha-1-antitrypsin, alpha- 1-antichymotrypsin and cytokeratin suggesting a diagnosis of UESL.

Discussion:
UESL is a rare, aggressive mesenchymal neoplasm that occurs in older children and adolescent age group and usually presents with non-specific symptoms such as an abdominal mass, fever, and pain (6). Laboratory data are non suggestive. Of possible significance is the absence of jaundice and normal serum AFP in these patients as was seen in our case (2, 7).

On US, UESL appears as a large, predominantly solid, well-defined mixed echogenicity liver mass with few small anechoic spaces reflecting its pathologic features (8). The right lobe of the liver is the site most frequently affected. However, this tumor usually has a misleading cyst like appearance on CT and MRI. CT may show a large mass lesion predominantly of water attenuation with thin septations in the periphery as we have seen here and T2-weighted MR images show high signal intensity almost equal to that of cerebrospinal fluid (9, 10). This discrepancy in the appearance is considered to be due to the high water content of the abundant myxoid stroma within the tumor. The myxoid components have been shown to contain acid mucopolysaccharides. These are negatively charged moieties that are hydrophilic and draw water osmotically into the ground substance and are characteristic of UESL (10). Therefore, US may be helpful to differentiate this tumor from predominantly cyst like masses on CT and MRI, such as biliary cystadenoma and cystadenocarcinoma, hepatoblastoma, mesenchymal hamartoma, cystic degeneration of hepatocellular carcinoma, cholangiocarcinoma, abscess, metastatic disease and post traumatic resolving hematoma (3,8). This unusual discordance is also helpful in preventing an erroneous attempt to drainage of the lesion (10).

To conclude, UESL presents in older children or in the adolescent age group usually with a right upper quadrant mass and no other specific symptoms. On CT the tumor appears paradoxically as a fluid attenuation mass lesion which appears to be predominantly solid on USG. In a young adult with a liver mass lesion, this finding is useful in making a prospective diagnosis of UESL and avoiding erroneous attempts at drainage. Histopathological examination and immunohistochemistry are used to confirm the diagnosis.

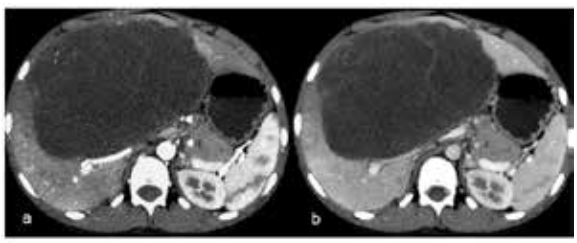


FIGURE: 1

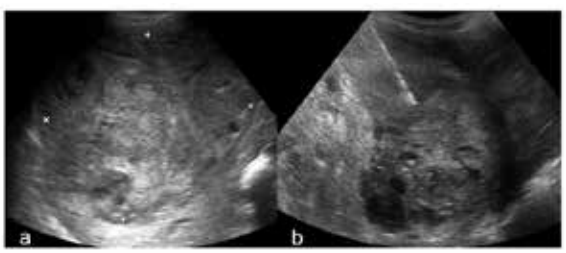


FIGURE: 2

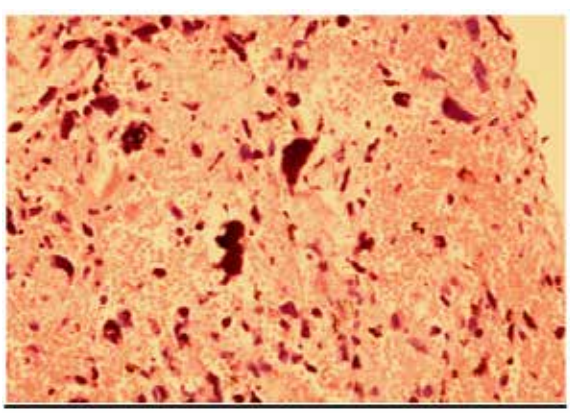


FIGURE: 3

Legends:

Figure 1: Axial contrast-enhanced CT scan in arterial (a) and portal venous (b) phases show a large lesion of predominantly low attenuation close to that of water in the liver. Thin septations are noted at the periphery of the lesion.

Figure 2: USG image (a) shows a well-encapsulated, heterogeneously hyperechoic solid mass with few small scattered anechoic foci suggesting cystic cavities. True cut needle biopsy (b) was done from the solid portion.

Figure 3: Histopathology shows tumor composed of mixed population of pleomorphic oval to elongated epithelioid and spindle cells, scattered loosely in the myxoid ground substance and hemorrhagic tissue. The tumor cells displayed irregular dense hyperchromatic nuclei and scant cytoplasm.

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