Presentation of an unusual case of hepatic alveolar echinococcosis : multi-detector CT and US findings

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A 40 year old male farmer admitted to internal medicine clinic with post prandial dyspnea and excessive sweating lasting for the last two months. In his physical examination, hepatosplenomegaly and right upper quadrant tenderness were detected. He had no jaundice or pruritus. His liver function tests were elevated with normal bilirubin levels. He was referred to our department for Ultrasound (US) examination which revealed multiple well defined, nodular, homogeneous lesions isoechoic with the liver parenchyma and with surrounding hypoechoic rims (Fig. 1a). Further evaluation with abdominal Computed Tomography (CT) demonstrated a total of six masses in both lobes of the liver. The largest of them located at segment 7 was 10 cm to 7.5 cm. All lesions were solid, with well defined, lobular borders and all had a central calcification. Densities of the lesions were ranging from forty to forty- five Hounsfield Units. Lesions had no contrast enhancement in arterial, portal venous or late phases. Although they were compressing the vascular structures, there was no sign of invasion. Compression to biliary system produced intrahepatic bile duct dilatation which was more evident in the left lobe while common bile duct was within normal limits (Fig. 1b). Lymphadenopathy at distal paraesophageal and portal hilus regions were present, largest of them measuring 3 cm in length. CT examination was otherwise unremarkable. A primary diagnosis of metastasis was made though hepatocellular carcinoma could not be excluded. Cranial and thoracic CT scans were also obtained to find out a primary neoplasm but they were unremarkable. Histological diagnosis was recommended because it would affect the choice of treatment. However, the biopsy specimen obtained revealed alveolar echinococcal lesion, a diagnosis that was not considered. The serologic test applied afterwards was positive for Echinococcus multilocularis and diagnosis was made to be alveolar hydatid disease of the liver. The patient was accepted as inoperable and liver transplantation following albendazole treatment was decided. By the time of this writing, he had had a successful transplantation from a living donor and no complications occurred for two months until now.

Alveolar hydatid disease is a rare parasitic disease with a mortality rate of 90% within 10 years in untreated cases (1,2). It has the characteristics of a slow growing cancer infiltrating the liver and surrounding structures, especially the vascular and biliary system. Central necrosis frequently develops as the mass increases in size as well as calcifications, a common finding (3). So far, many case reports or series were published to state the characteristic imaging features of the lesion. Although different presentations are possible, described main features were heterogeneous mass with ill defined borders, scattered or peripheral calcifications and sometimes central necrosis (1-9). Four main patterns were defined by US ; granular strong echo with or without acoustic shadowing (coarse calcifications), irregular heterogeneous areas (inflammatory changes or necrosis), small hypoechoic areas (vesicles) or large hypoechoic areas (liquefied necrosis or cysts) (2). Reports until now describe the most common morphological CT profile as a heterogeneous, infiltrative and destructive mass with irregular outlines and non-vascular necrotic center, appearing as a low density structure of 0-25 HU often (83%) with scattered coarse calcifications (2,4-6). Lesions are usually solitary and invasive nature is an important feature (2,7). Calcifications are the characteristic sign of the lesion and may be visible in plain radiographs, either as cyst wall calcifications or clusters (4,7). Ogasawara and colleagues categorized CT appearance in four aspects : focal calcifications, irregular areas of low attenuation, small low attenuation cysts smaller than two centimeters or low attenuation cysts larger than two centimeters. They also stated that cysts might be accompanied by low attenuation areas resembling necrosis (8).

Our case revealed imaging features almost contrary to what has been reported until now ; homogeneous well defined masses including no cystic or necrotic areas and single calcifications at their centers instead of scattered calcifications. The only feature that resembles the alveolar hydatid cases published before is that there was no contrast enhancement of the masses on CT. With these findings, our case is unique in literature and may lead the radiologists to think of alveolar hydatid disease as an alternative in differential diagnosis of masses with such characteristics.

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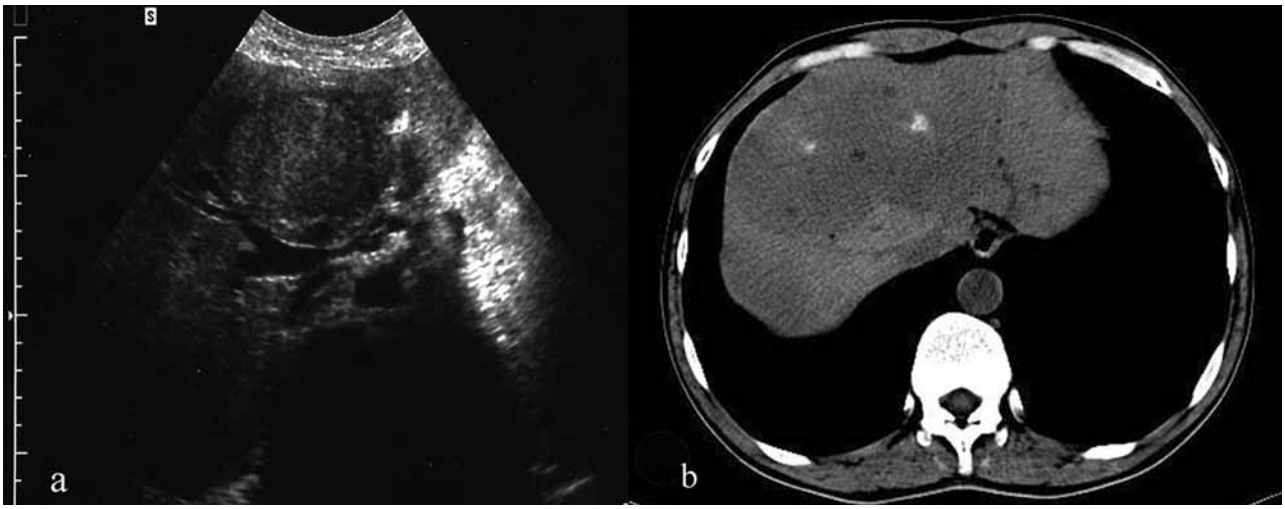

Fig. 1. — (a) US demonstrates a homogenous, well defined mass, isoechoic with the liver parenchyma and a hypoechoic rim. Note the compressed portal vein and dilated intrahepatic bile duct ; (b) Non-enhanced CT defines two homogenous hypodense lesions with densities of 45 HU located at the dome region of the liver. Lesions have distinct irregular borders. Note the central calcifications within and accompanying bile duct dilatation.

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