Case Report

Hepatic Alveolar Echinococcosis

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Abstract

Alveolar hydatid disease is a highly malignant form of echinococcosis caused by the larvae of the cestode echinococcus multilocularis. Alveolar hydatid disease always affects the liver and can metastasise to the lung and brain. Early diagnosis and precise evaluation of the localization as well as the extent of lesions are essential for treatment. In this report, we present ultrasound and computed tomography findings in a patient with hepatic alveolar echinococcosis. The patient, who was presented with hepatomegaly, jaundice, and an infiltrative solid tumor, diagnosed by ultrasound and computed tomography. In contrast to hydatid cyst caused by echinococcus granulosus, this is a rare disease in Iran.

Keywords: Echinococcosis, echinococcus multilocularis, hepatic, Iran

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Introduction

Alveolar echinococcosis (AE) which is caused by echinococcus multilocularis (EM) is a chronic, critical and sometimes fatal parasitic infection. Alveolar hydatid disease is endemic to North America, Alaska, Central Europe, and Turkey. The cyst of EM grows by external buckling of the germinal membrane with a progressive infiltration of the surrounding tissue which differs from that of E. granulosus.[1,2] The liver is the most common site of involvement (95%), but the parasite may disseminate from there to other organs, such as the lung and brain.[3,4] Clinical symptoms include epigastric pain, fever, jaundice, and hepatomegaly. Diagnosis depends on imaging examination, serologic tests, and pathologic findings, because the clinical presentation is usually non-specific.[1,5,6] ultrasonography (US), computed tomography (CT) scan, and magnetic resonance (MR) imaging are the most useful methods of diagnostic imaging. Without timely diagnosis and therapy the prognosis is worse.[4] Surgery is the treatment of choice and combined with parasite static chemotherapy.[4,6] There are a few reports of hepatic AE especially from the northern part of Iran. In this article, we report a case of hepatic AE, who presented with hepatomegaly and jaundice from Khorasan.

Case Report

A 40-year-old woman from the North-East of Iran referred to our hospital with a history of abdominal pain, hepatomegaly and jaundice for one year. In addition, she complained of weight loss, and anorexia, without any fever. Physical examination revealed a firm and tender hepatomegaly with a hepatic mass. Abdominal ultrasound demonstrated a large liver mass with heterogeneous echogenicity and irregular margins. There was evidence of a mild biliary duct dilatation. Precontrast abdominal CT scan revealed an irregular hypodense mass with multiple foci of calcification that occupied most of the left liver lobe. CT images demonstrated multiple foci of calcification in the hepatic lesion. CT did not delineate perihepatic extension of the disease (Figure 1). Post contrast CT scan did not demonstrate any contrast enhancement of the involved area. The lesion appearance in CT scans occasionally demonstrates an infiltrative pattern (Figure 2). The patient laboratory results were as follows: alkaline phosphatase, 90u/liter; aspirate aminotransperase, 23u/liter; lactate dehydrogenase, 304u/liter; bilirubin, 12umol/liter; and white blood cells, 5.2×10^9/liter with 68% neutrophil. The patient underwent hepatic surgery. In the histological examination of the biopsy specimen, irregular cysts containing laminated membranes were seen, but neither nucleated germinal membranes nor protoscolices or hooklets were noticed. The host immune reaction to cysts comprised a peripheral fibrosis with inflammatory cells. Pathologic examination revealed hepatic alveolar echinococcosis (Figure 3).

Discussion

Hepatic alveolar echinococcosis is a rare, but possibly lethal parasitic disease, which develops due to the growth of the EM larvae in the liver. The characteristic features of this infection are its exogenous, tumor like, multi vesicular, and infiltrating structures that consist of numerous small vesicles embedded in a stroma of connective tissue. Humans are accidental intermediate hosts, as rodents are the most common intermediate host. Carnivores, such as foxes are the final hosts and can transmit the parasite to humans via their feces, and the larva develops in the liver. AE is an endemic disease in North America, Alaska, central Europe (such as Austria, Germany, Eastern France, and Switzerland), Balkan states and parts of Asia. However, the majority of human cases of AE, have been reported in the Peoples of Republic of China.[1,2,4] Although, Echinococcous granulosus is an endemic disease in Iran, but there is a little reported data about the AE. The liver is the initial site of infestation by EM, but the infection may disseminate from there to other organs of the body, such as the

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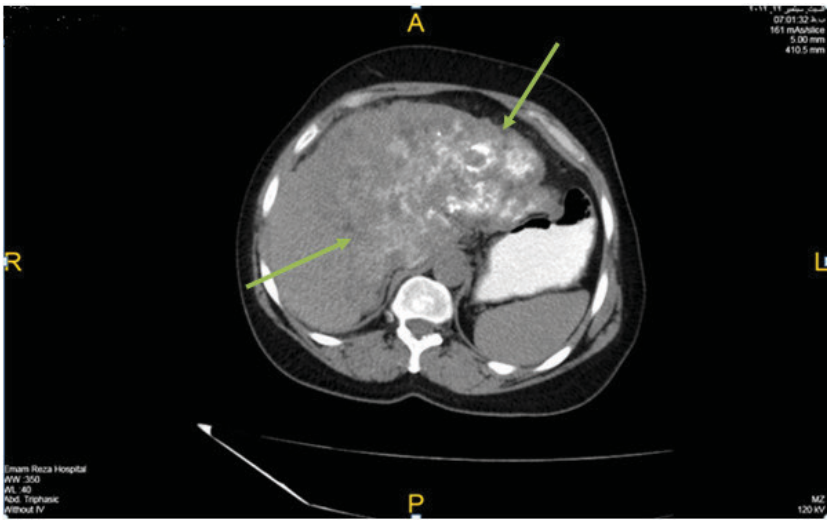

Figure 1. Abdominal CT without I.V contrast injection: hypo-dense liver mass with in distinct, irregular margins and dense calcification (arrows).

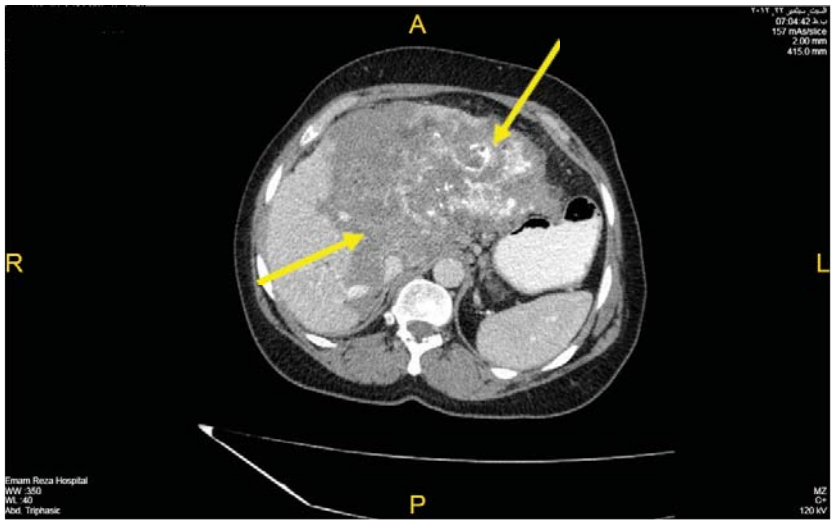

Figure 2. Abdominal CT with I.V contrast injection: hypo-dense lesion with calcification and no evidence of a significant enhancement after i.v bolous injection of contrast material (arrows).

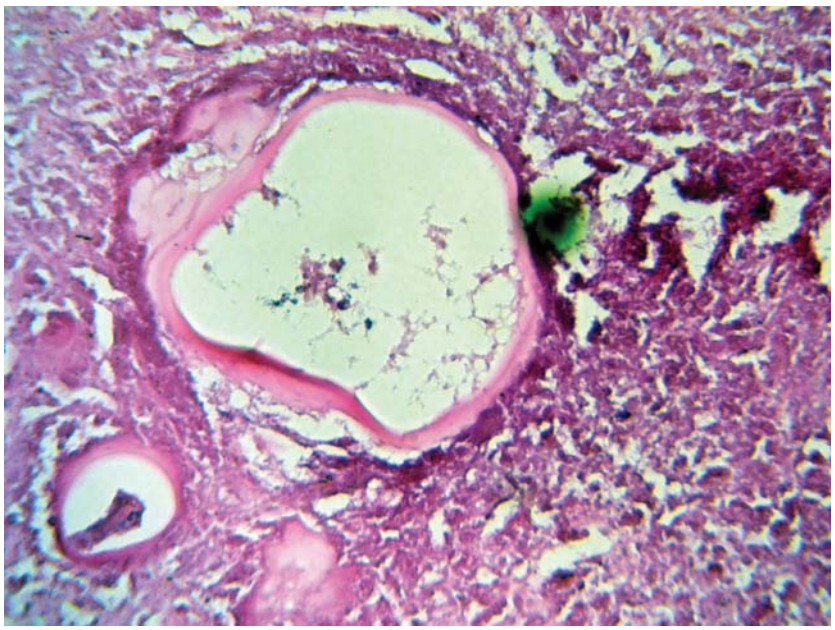

Figure 3. Histopathology of liver lesion. Cystic mass with budding daughter cyst with a tri layered membrane wall is visible. No protoscolices or calcaneous carpuseles are seen. (H&E, magnification: 400×).

lung, heart, brain, bones, and soft tissues.[1,3,4] Liver involvement has been observed in 90% of the patients. The disease usually reaches other organs by secondary hematogeneous dissemination, but it may also spreads contiguously to adjacent organs by direct extension. It also grows like a slow tumoral lesion.[1,7] The most common presenting symptoms of liver involvement are RUQ pain, fever, jaundice, itching, and portal hypertension. Our patient presents with abdominal pain, hepatomegaly and a mild jaundice. Early diagnosis is imperative to the patient in order to prevent complication.[1,5] Diagnosis depends on imaging methods, serology, and pathologic findings. The basic imaging methods for the diagnosis of AE are Us and CT scan.[6,8] Us is the screening method of choice. Typical ultrasound findings are large sized irregular cysts, with heterogeneous and hypoechoic contents.

The liver mass may be associated with hypoechoic central areas of pseudo-liquid necrosis and hyper echoic areas due to the reactionary fibrotic tissue. Many lesions may have irregular calcifications, which may be micronodular, nodular, or en plaque.[6] In our patient, US showed a hepatic mass with heterogeneous echogenicity, irregularity and internal calcifications. Color doppler ultrasound may be helpful and the absence of vascular flow in a solid hepatic mass is a diagnostic feature when associated with the above-mentioned sonographic features.

CT scans should be always performed, because it has a high sensitivity (95%). CT scan determines the number, size, localization and vascular, biliary, and extra-hepatic extension of the hepatic lesions. In severe infestation, the walls of the bile ducts and blood vessels may be invaded.[8] Involvement of abdominal lymph node,[9] and budd-chiary syndrome secondary to vascular invasion had been reported in patients with hepatic alveolar AE.[10,11] Cross-sectional imaging is crucial in differentiating echinococcosis from malignant processes. CT scans have usually demonstrated a mass like hepatic lesion with irregular borders, which may be accompanied by cystic or calcified components. Calcification is usually observed at the periphery of the lesions, but it may also occur in the central part of the lesion. Calcification is a common finding on CT scan and usually appears either punctuate or amorphous in shape. Microcalcifications have been reported in hepatic EA. The reticular fibrotic tissue may enhance a little in contrast to necrotic areas with no change in enhancement. Hilar involvement is a frequent finding. In severe infestation, the infiltrative nature of the lesion causes stenosis of the intrahepatic bile ducts and involvement of the hepatic and portal venous branches. Inferior vena cava invasion may also occur. There are multiple cases of budd-chiary syndrome due to the hepatic EA in the literature. In our patient, CT scan revealed a hypodense and heterogeneous mass with irregular margin and multiple amorphous calcifications, mainly at the peripheral of the lesion. There was no evidence of a significant enhancement or vascular invasion after I.V contrast injection in our patient. Follow up by US and CT scans can display the increase of primary lesions, the occurrence of new foci, and local or regional extensions.[9,12]

MRI with standard and diffusion-weighted sequences, all provides useful information and plays complementary rules in detecting and diagnosing hepatic echinococcal lesions.

In uncertain cases, MR imaging may be performed to demonstrate a cystic component. Hepatic EA may show various signals intensity on T1 and T2–weighted images, and demonstrate slight enhancement on gadolinium –enhanced images. The cystic, necrotic, and fibrotic components appear as a hypointense area within the liver parenchyma in T1-weighted images. T2-weighted images are useful to detect small cystic areas. We did not perform MR imaging for our patient. MR imaging can help in evaluation of the biliary duct and vascular involvement as well as an extra hepatic extension of AE.[13]

F-18-flouro deoxyglucose (FDG) is a sensitive and specific adjunct in the diagnosis of suspected AE and can help in differentiating AE from echnococcus granolusus.[14] Although, a specific diagnosis can be made by examination of aspirated lesion contents, serodiagnostic tests are often preferred because aspiration may result in either dissemination of infection or anaphylactic reactions. Serologic test for EM is based on the detection of a specific antibody response by using an enzyme-linked immunosorbant assay with a possible Cross-reaction for E granolusus. Polymerase chain reaction technology is now widely used because it is more specific and don't involve cross-reactions.[15] Treatment of hepatic AE is extremely critical because once the clinical symptoms of these patients appear; liver lesions have already become infiltrative and extensive which making it hard to operate. Surgery is the treatment of choice in early stages of the disease and various types of hepatectomy have been performed.[2,4,16] When the hepatic lesion is very extensive and infiltrative. Liver transplantation can be performed.[4,16] Medical therapy is also important in AE involving use of benzimidazole carbonate derivates, such as albendazole and mebendazole. Albendazole is more effective than mebendazole, and chemotherapy must be instituted and maintained for at least six months.[4,5]

In conclusion, although hydatid disease due to infestation with echinococcus granolosus is known to be endemic in Iran, hepatic AE is not common. Cross-sectional imaging is crucial in differentiating AE from malignant lesions and this entity should be included in the differential diagnosis, especially in countries where the disease is endemic. We would also like to emphasize that when imaging studies revealed an extensive, infiltrating and tumor like lesion in the liver, AE should be kept in mind as a possibility in these patients.

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