JBR–BTR, 2011, 94: 21-23.

HEPATIC ALVEOLAR ECHINOCOCCOSIS: A DIAGNOSTIC CHALLENGE

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We report a case of alveolar echinococcosis involving the liver in a 61-year-old male. Alveolar echinococcosis is a rare chronic and progressive disease, which can involve mostly liver and in rare cases lung and brain. It is caused by Echinococcus multilocularis. In this report we describe the imaging findings of hepatic involvement of alveolar echinococcosis by ultrasonography, computed tomography and magnetic resonance imaging.

Key-word: Echinococcosis.

Case report

Alveolar echinococcosis is a rare, but potentially fatal, chronic and progressive disease, caused by infestation of the liver by Echinococcus multilocularis. Humans are the intermediate host that become infected by ingestion of the wild berries, plants or water which is contaminated with eggs of the parasites or by direct contact with the definitive host, which is the wild canine (foxes, wolves). The disease is usually seen in central and northern Europe, North America, Canada, China and Japan (1). It is endemic in eastern Turkey (2). An invasive tumor-like multi-vesiculated lesion develops during a long, asymptomatic period which is characteristic for the disease. In this study we present a case of Echinococcus multilocularis infestation, forming a tumor-like lesion in the liver.

A 61- year-old man was referred to our clinic with right-upper quadrant pain, weight loss and fatigue of almost 1 year duration. Physical examination revealed no significant finding. Abnormal laboratory findings were as follows: erythrocyte sedimentation rate 52 mm/h (5-25 mm/h), gamma-glutamyl transferase 122 IU/L (8-61 IU/L), lactate dehydrogenase 500 IU/L (240-480 IU/L), serum aspartate transaminase 52 IU/L (0-37 IU/L), white blood cell count 10.75 x10^3/UI (3.8-10.0 x 10^3/UI), neutrophil percentage 32.8 (45-78%), eosinophil percentage 34.8 (0.8- 7.0%), haemoglobin 11.6 g/dL (13.0-17.5 g/dL), CA 125 level 48.2 U/ml (0-30.2 U/ml), CA 19-9 level 71.4 U/ml (0-37 U/ml). Hepatitis B surface antigen level was 287.8 mIU/ml in the serum (positive: > 10 mIU/ml).

Ultrasonography (Aplio XV, Toshiba, Tokyo, Japan) revealed a large heterogeneous liver mass almost completely filling the right lobe with isoechoic and hyperechoic areas relative to the normal liver parenchyma. There was a small hypoechoic cystic space with irregular margins on the right upper peripheral region (Fig. 1A). The mass had irregular, indistinct borders. It demonstrated no significant finding by colored Doppler ultrasonography. Because the use of sonographic contrast agents were not allowed by the ministry of health in Turkey, contrast enhanced ultrasonography could not be performed.

Unenhanced abdominal computed tomography (Siemens Somatom Sensation 16-detector row CT, Erlangen, Germany) revealed an area of heterogeneous density in the right lobe of the liver with some millimetric calcifications located centrally (Fig. 1B). The margins were indistinct and ill-defined. There was no prominent contrast enhancement after the iodinated contrast medium injection (Fig. 1C). Magnetic Resonance imaging (Signa Excite HD 1.5T; GE Healthcare, Milwaukee, WI, USA) of the liver revealed a heterogeneous hypointense mass with irregular margins on both unenhanced T1 and T2- weighted images (Fig. 2A-B). The lesion measured 16 cm x 12 cm; it was almost totally filling the right upper lobe and extending into the left lobe. It had a right peripheral cystic necrotic area. After the administration of hepatocyte-specific contrast agent (Primovist 0.25 mmol/ml - Bayer Schering, Berlin,Germany), the lesion revealed no contrast enhancement in arterial (Fig. 2C) and portal venous phases. There was no contrast enhancement in the late hepatobiliary phase (Fig. 2D), indicating that the mass does not contain any hepatocytes.

Following the radiological work-up, the patient was referred to the liver biopsy. The biopsy was completed without any complication. The histopathologic work-up, as the most reliable method to have the definitive diagnosis, showed hepatic alveolar echinococcosis.

Discussion

Hepatic infestation of echinococcus multilocularis appears as an ill-defined infiltrative disease usually consisting of cystic and solid components. Microcalcifications are commonly seen (3). Necrotic areas frequently develop due to ischemia as the lesion size increase. The surrounding vascular structures and biliary system may be infiltrated by the disease or they may even be displaced due to the mass effect of the lesion, which imitates a slow-growing tumor (3).

In most cases, lesions are hyperechoic with heterogeneous echopattern and indistinct margins on ultrasonography (USG) (1). Cavitary or cystic to vesicular appearance can be seen within the masses. Color Doppler USG shows the absence of vascular flow in the solid components of the lesions (4).

CT imaging of the liver infestation shows heterogeneous, ill-defined, hypodense areas which have poor or no enhancement after intravenous contrast administration (1). In 90% of the infected cases, calcifications are found (1). Pseudocystic necrotic areas and dilatation of the intrahepatic bile ducts can also be identified (3).

The fibrotic changes of the disease process and extension of the lesion within the liver parenchyma can be demonstrated by MRI (3). Fibrosis and parasitic tissue have low signal intensity on T1-weighted images (3). On T2-weighted images, lesions may be hypointense, hyperintense or isointense (1). Central necrotic zones and cystic peripheral extensions are better identified on T2-weighted images (3).

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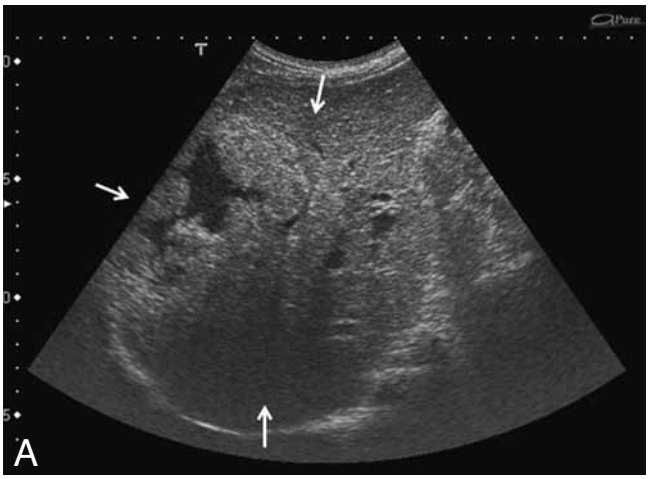

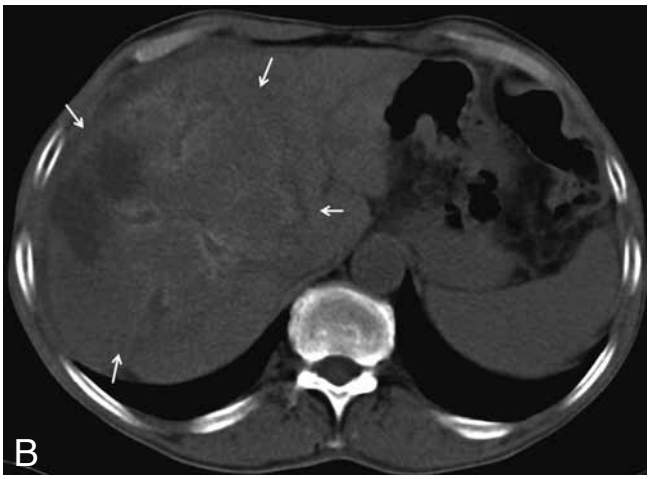

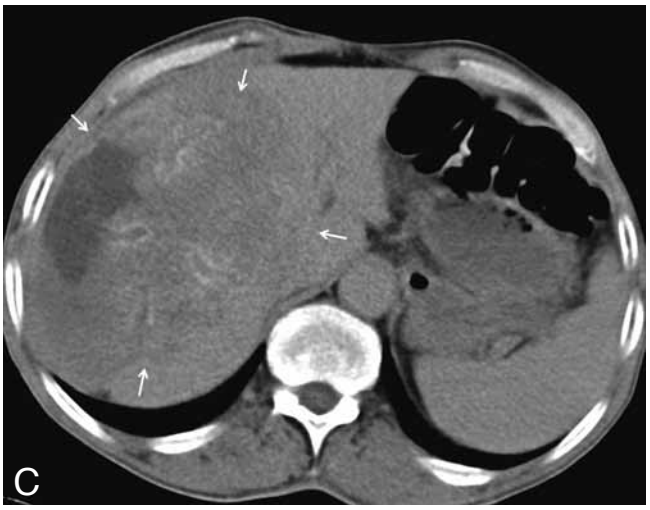

Fig. 1. — A: Axial grey-scale ultrasound image reveals a large hepatic mass with heterogenous echogenity. B: On unenhanced CT image the mass shows heterogeneous hypodensity and contains calcific areas. C: After intravenous administration of the iodinated contrast material, the mass does not show any enhancement.

There is poor or no enhancement after intravenous administration of gadolinium (1).

CT and MRI findings of alveolar echinococcosis can overlap with those seen in malignant liver neoplasms such as hepatocellular carcinoma (HCC), intrahepatic cholangiocarcinoma (IHCC), solitary metastasis and primary lymphoma of the liver.

On unenhanced CT scans, HCC is usually demonstrated as a large, hypodense mass, often with central areas of low attenuation representing areas of necrosis (5). On MRI, HCC lesions typically appear hypointense on T1-weighted sequences; hyperintense on T2-weighted sequences. On both CT and MRI, on contrast-enhanced images, increased arterial enhancement and a rapid washout is observed (6).

Intrahepatic cholangiocarcinoma appears as a homogenous, hypodense mass on unenhanced CT images. On MRI it is seen as a hypointense mass on T1-weighted images and as a hyperintense mass on T2-weighted images (7, 8). After the intravenous contrast agent administration, the enhancement patterns depend on the size of the lesion (8). Larger lesions (> 4 cm) show peripheral enhancement which progress centripetally, while smaller lesions (2-4 cm) enhance homogenously (8).

On unenhanced CT scans, primary lymphoma of the liver is often a poorly defined mass of low attenuation. Necrosis and calcification may be observed (9). On T1- weighted MR images lesions are hypointense. On T2- weighted images, low to moderately high signal intensity is seen. After intravenous contrast administration, enhancement is predominantly peripheral (10).

Most of the solitary liver metastases appear hypodense relative to liver parenchyma on unenhanced CT images (11). On T1-weighted MR images the lesions are usually hypointense. Lesions frequently show heterogeneously moderate to moderately high signal intensity on T2-weighted images. Since the majority of the metastatic liver lesions are hypovascular, they show a diminished late enhancement on contrast-enhanced dynamic imaging (12, 13).

Unlike E. granulosus infection, which has typical radiological patterns such as daughter cysts and detached membranes, alveolar echinococcosis is hard to diagnose in most cases (14). Unfortunately, no pathognomonic feature of hepatic alveolar echinococcosis is identified. The clinical course may resemble that of a slowly developing tumor (15). The definite diagnosis can be reached by correlating radiological findings with the clinical and laboratory data, and in some cases by the histopathologic work-up. Even the cyst aspiration for diagnosis was considered as a possible risk of anaphylaxis or recurrence due to the spillage, USG guided fine needle aspiration biopsy is used to confirm the diagnosis in some cases (16, 17). Especially in endemic regions, if a complex appearing liver lesion is detected, alveolar echinococcosis must be definitely included in the differential diagnosis.

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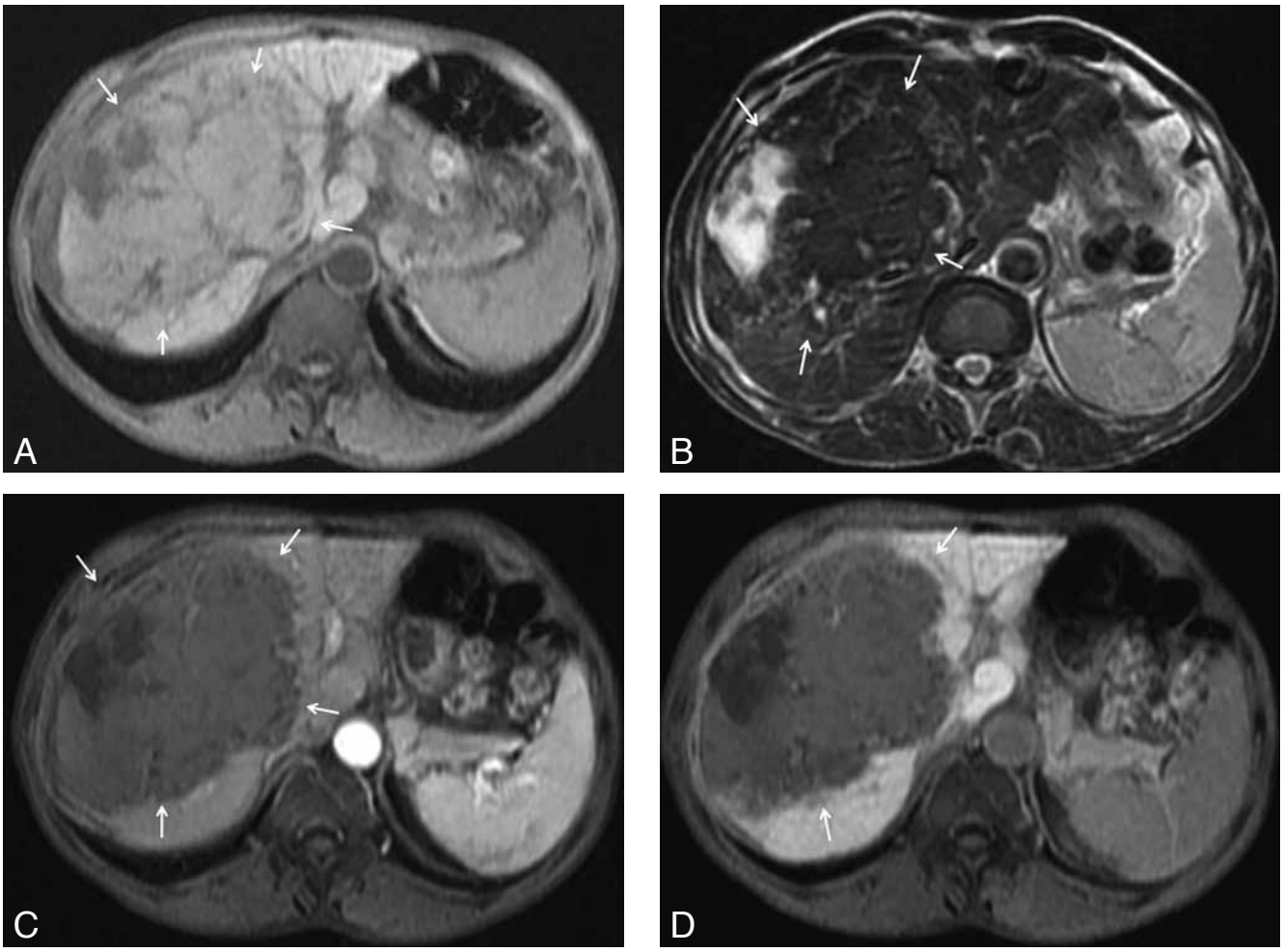

Fig. 2. — On T1-weighted (A) and T2-weighted (B) MR images the mass shows heterogenous signal intensity. On T1-weighted MR images obtained after intravenous administration of gadolinium-containing hepatocyte specific contrast material (C), the mass does not enhance in the arterial phase. On images obtained during the hepatobiliary phase (D), there is no enhancement. neither.

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