AN UNUSUAL CAUSE OF CIRRHOSIS: ALVEOLAR ECHINOCOCCOCCOSIS

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ABSTRACT

Alveolar echinococcosis (AE) is an uncommon but potentially fatal disease.

We report a 49-year-old female patient who was admitted to our clinic with a large palpable mass in her right upper quadrant and ascites. Contrast enhanced axial computed tomography (CT) showed a calcified mass lesion filling the left hepatic lobe with extension to the right hepatic lobe and ascites. Echinococcus serology assessed by ELISA was noted to be positive. Histopathologic examination of the hepatic mass was consistent with alveolar echinococcosis. The diagnosis was cirrhosis due to alveolar echinococcosis. AE must be considered in the differential diagnosis in a patient with ascites in cirrhosis, especially in endemic regions.

Key Words: Alveolar echinococcosis, liver, cirrhosis Nobel Med 2011; 7(1): 106-108

OLĂĞAN DIȘI BİR SİROZ NEDENİ:
ALVEOLAR EKİNOKOKKOZİS

ÖZET

Alveolar ekinokokkozis (AE) fazla yaygınındır, fa-
kat potansiyel olarak öldürük bir hastalıktır.

Biz burada asıt ve sağ üst kadranı dolduran büyük palpabl külesi mevcut olan 49 yaşında, kliniğimizde takip ettiğimiz bir hastayı sunduk. Kontrastla kuvvet-

lendirilmiş aksiyel komputerize tomografi, sol hepatik lobu dolduran ve sağ hepatik loba uzanan, kalsifiye küüleyi ve asıtı gösterniyordu. ELISA ile değerlendirilen ekinokokkozis serolojisi pozitif idi. Hepatik küt-
lenin histopatolojik değerlendirme AE ile uyumlu geldi. Hasta AE ye bağlı siroz olarak değerlendirildi. Özellikle endemik bölgelerde asıtı ve sirozlu hastaların teşhisinde AE göz önünde bulundurulmalıdır.

Anahtar Kelimeler: Alveolar ekinokokkozis, karaci-
INTRODUCTION

Alveolar echinococcosis is an uncommon parasitic disease caused by Echinococcus multilocularis. In the human host, the parasite involves primarily the liver. It proliferates like a tumor and may induce poor-prognosis disease, with a high death rate of approximately 90% within 10 years of the diagnosis. Echinococcus alveolaris (EA) grows with external vesiculation, leading to stenosis of the intrahepatic bile ducts, hepatic veins, and portal vein branches. One-third of cases present with cholestatic jaundice, one-third present with epigastric pain, and the remainder present with vague symptoms like weight loss or fatigue or are diagnosed incidentally during radiological examinations. EA has a widespread geographic range, including an endemic area in central Europe, parts of North America, and most of central Eurasia (parts of Turkey, Iran, India, and Japan). We report a female patient with ascites and non-resectable alveolar echinococcosis which manifested with a calcified mass that occupied the left hepatic lobe with extension to the right hepatic lobe.

CASE REPORT

A 49-year-old woman was admitted to Ataturk University Hospital with the complaints of abdominal bloating, weight loss, epigastric pain and fatigue. The patient had no history of alcohol consumption. Physical examination revealed cachexia, jaundice, a large palpable mass in the right upper abdomen, and massive ascites but no splenomegaly. The patient had venous distention on the abdominal wall, around the umbilicus. Bilateral pedal edema was noted. Examinations of the respiratory and cardiovascular systems were normal. Laboratory findings revealed an increase in alkaline phosphatase, lactate dehydrogenase, bilirubin, and gamma-glutamyl transpeptidase levels. Laboratory studies showed no evidence of eosinophilia in the differential count. Serum albumin was 2.6 g/dl. However, the γ-globulin was markedly elevated. Prothrombin time was lengthened. All additional routine laboratory tests were normal. Tests for antihepatitis C virus antibody, hepatitis B surface antigen (HBsAg), and anti-HBsAg antibody were negative. Tests for tumor markers were negative. In upper gastrointestinal endoscopy, grade 2 esophageal varices were observed. We performed paracentesis, which yielded a large volume of ascitic fluid. Total albumine concentration of the ascites fluid was 0.8 g/dl. The serum ascites-albumine gradient was 1.8 g/dl. Serum sample was investigated for E. multilocularis antibodies using Em2plus enzyme-linked immunosorbent assay (ELISA), and serology was noted to be positive. Contrast enhanced axial computed tomography (CT) showed a calcified mass lesion filling the left hepatic lobe with extension to the right hepatic lobe and ascites. The left and middle hepatic veins were invaded. The right and left portal veins were invaded as from portal hilum (Figure 1). The alveolar mass resulted in compression of the retrohepatic vena cava and thrombosis of this vessel (arrow). Ultrasound-guided liver biopsy was performed. Histopathologic examination of the hepatic mass was consistent with alveolar echinococcosis (Figure 2). Our patient had a disease stage 3b (P4N0M0) based on PNM staging (P, hepatic localization of parasite; N, extrahepatic involvement of neighboring organs; M, absence or presence of distant metastases). The lesion in the liver was accepted as nonresectable. Liver transplantation was considered but the patient did not accept surgical treatment. Albendazole was administered at 10 mg/kg daily and her follow-up is continuing.
DISCUSSION

Echinococcus multilocularis larvae aggravate a prominent granulomatous reaction and the lesions are never restricted by a fibrous defense. In humans, as an aberrant intermediate host, a gradually progressive malignant tumor-like growth in the liver occurs, delaying the diagnosis for several years. Invasion of the bile ducts and the vessels leads to severe complications, such as cholangitis, portal hypertension, and biliary cirrhosis. Portal hypertension and secondary biliary cirrhosis are the most frequently observed clinical features. This disease can progress to the cirrhotic stage after long terms of latent and asymptomatic stages. The increase in bilirubin and alkaline phosphatase in our case was probably related to involvement of the hilar region in association with secondary biliary cirrhosis.

Early clinical symptoms are absent and most patients are diagnosed at late stages with unresectable hepatic lesions, as observed in the present case. Early diagnosis can improve the prognosis and treatment of infected patients, and it relies on complementary procedures such as evaluation of clinical symptoms, radiologic imaging methods and serologic diagnosis. CT is always performed because it has a high sensitivity (94%). Classically, CT demonstrates an infiltrating tumor-like hepatic mass with irregular margins and a calcified or cystic component. On CT, delineating the hypodense lesion with calcifications and discerning the lack of contrast enhancement are helpful in differentiating EA from the liver neoplasms. CT in our case revealed a large non-homogeneous mass located in the liver in close vicinity to the liver hilum and exhibiting a structure indicative for AE.

At present, the treatment is still under debate. Though drug therapy may stabilize the lesions in some cases, surgical treatment has been the mainstay of therapy, but radical surgery is occasionally not feasible, as in this case. Liver transplantation (LT) should only be considered in patients with severe hilar extension, leading to symptomatic secondary biliary cirrhosis with ascites or severe esophageal varical hemorrhage due to portal hypertension. We wanted to transfer our patient to another hospital for liver transplantation, but she did not accept surgical treatment.

At this time, there is no parasitocidal drug available to treat AE. Some studies provide clear evidence that long-term drug therapy of AE in humans with benzimidazole derivatives (albendazole, mebendazole) produces a decrease or arrest of parasite proliferation in the majority of patients (i.e. 84%). Thus, in cases in whom surgery as a therapeutic option is impossible, chemotherapy should be used for long time (about 10 years). In our case, albendazole was administered at 10 mg/kg daily and the patient’s follow-up is continuing.

In conclusion, alveolar echinococcosis should be differentiated from other hepatocellular masses. We believe that in endemic regions, alveolar echinococcosis disease, as a rare cause of cirrhosis, should be remembered in the differential diagnosis. An expansion of screening programs for alveolar echinococcosis in endemic regions would help to prevent delayed diagnosis and provide the opportunity for curative treatment of the disease.

REFERENCES