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CASE REPORT

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Portal hypertension and cholestasis due to Echinococcus alveolaris: A case report

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Abstract

Alveolar ecchinococcosis (AE) is a rare but potentially life-threatening serious parasitic disease. The parasit may involve liver and, most importantly, expand vessels and biliary tract. We present here a case of esophageal variceal bleeding due to portal hypertension and cholestasis secondary to AE. We present the case of a 30-year-old female patient who was admitted to emergency service with hematemesis, melena and icter. Upper gastrointestinal endoscopy revealed gastroesophageal varices and portal gastropathy. Contrasted computerized tomography examination showed a solid calcific lesion (cross dimension 6x11 and 5x9 cm.) of biliary tracts causing dilatation. An USG guided biopsy from the mass was performed and histological examination revealed AE. We also review here the literature on portal hypertension and cholestasis due to Echinococcus alveolaris. AE is rare a disease and may cause severe complications. Endoscopic and percutaneous drainage for biliary obstruction caused by hepatic AE has a palliative effect even in the late stages. Liver transplantation was recommended but this operation was not performed for living donor shortage. Liver transplantation should be considered in the management of advanced liver alveolar echinococcosis.

Keywords: Portal hypertension, cholestasis, Echinococcus alveolaris

Introduction

Alveolar echinococcosis (AE) results from infection by the metacestode (larval) form of Echinococcus multilocularis [1]. Hepatic AE is a rare but potentially life-threatening serious parasitic disease. Larvae of AE produce fibroinflammatory response in host cells by building multilocular cysts in liver. The parasitic tissue is surrounded by an intense fibroinflammatory reaction [2]. The parasit may involve several liver segments and, most importantly, expand along large liver vessels and biliary tract, particular the hilum. AE may cause portal hypertension and cholestasis. Later, the larva infiltrates neighbouring organs. Cells of the larval tissue frequently detach and disseminate via lymph nodes and blood vessels, thus leading to metastasis in large parenchymatous organs. Therefore, AE is often compared with a slow-growing liver cancer [3]. We present here a case of esophageal variceal bleeding due to portal hypertension and cholestasis secondary to AE.

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Case report

A 30-year old female patient was admitted to emergency service with hematemesis and melena. Blood pressure was 90/60 mmHg and pulse 110/minute. Physical examination revealed icteric sclera and ascites. The spleen and liver was palpable below the subcostal margin. Complete blood count showed; hemoglobin: 5,7 gr/dl, hematocrit: 16%, white blood cells: 22500 /mm^3 and platelet: 227000 /mm^3. Biochemical parameters: Aspartate aminotransferase (AST): 106 U/L, Alanine aminotransferase (ALT): 50 U/L, Alkaline phosphatase (ALP): 432 U/L, Gamma glutamyl transpeptidase (GGT): 125 U/L, Lactate dehydrogenase (LDH): 226 U/L, Total bilirubin: 14.2 mg/dl, Direct Bilirubin: 10,9 mg/dl, Albumin: 1,8 g/dl, Prothrombin time: 20 seconds. Hepatitis markers were negative and tumors markers were within normal ranges. Urgent upper gastrointestinal endoscopy revealed gastroesophageal varices and portal gastropathy without bleeding. Abdominal ultrasonography (USG) showed ascites and a partially calcific liver mass of 10 cm diameter. Contrasted computerized tomography examination showed a solid calcific lesion (cross dimension 6x11 and 5x9 cm.) with lobulated contour in segment 4-8 which extended to the bifurcation of

biliary tracts causing dilatation in intrahepatic biliary tracts (Figure 1). The mass was surrounding the left-right portal vein and right inferior hepatic vein. An USG guided biopsy from the mass was performed and histological examination revealed EA (Figure 2). Albendazole treatment was initiated following biopsy diagnosis. Endoscopic retrograde cholangiography was unsuccessful and percutaneos biliary drainage performed for cholestasis. Liver transplantation was recommended in general surgery consultation but this operation was not performed for living donor shortage.

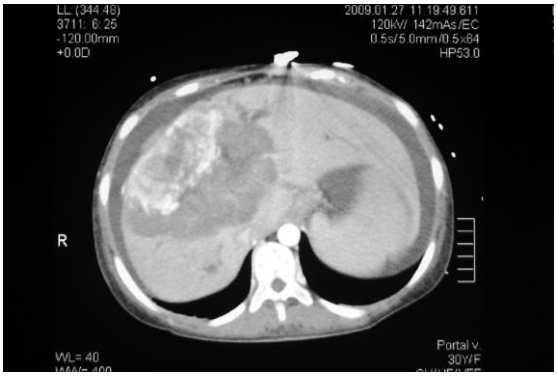

Figure 1. Solid calcific lesion with lobulated contour in segment 4-8 of liver in upper abdominal computerized tomography

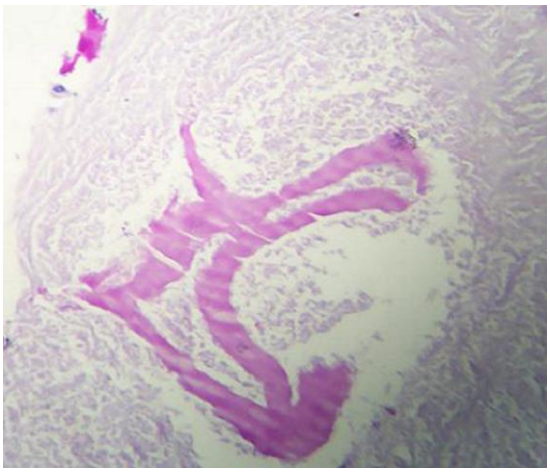

Figure 2. Periodic acid Schiff (PAS) positive magenta colored acellular lameller structr ues of E M among necrotic tissue obtained by tru-cut biopsy (PAS stain X200)

Discussion

Echinococcus alveolaris is a rare parasitic disease due to the intra-hepatic development of the larva of the small taeniid cestode Echinococcus multilocularis. This disease is only observed in the Northern Hemisphere. The main endemic regions for human AE are Central Europe, Russia, Turkey, Japan, China and North America [2]. Eastern part of Turkey is an endemic region for EA [4]. Cases of AE are characterized by an initial asymptomatic incubation period of 5 to 15 years and a subsequent chronic course. The right lobe is most frequently involved [5]. Parasitic lesions in the liver can vary from small foci a few millimeters in size to large (15–20 cm in diameter) areas of infiltration. A very dense and serious fibrosis may develop in the liver [6]. There is a broad clinical spectrum in this disease. Some patients may have no clinical signs for years, some experience biliary complications, upper gastrointestinal bleeding related to portal hypertension, Budd-Chiari syndrome, which all can conclude in death [7]. In our case have portal hypertension, ascites, esophageal varices and cholestasis.

The definitive diagnosis of AE in most cases depends on routine radiological examinations such as US, CT, or MRI, specific antibody determination in the serum and pathological examination. On sonography, lesions are heterogeneous with indistinct margins and in most cases hyperechoic. Didier et al. described the so-called hailstorm pattern. Masses also can appear cavitary or cystic to vesicular [8]. Color Doppler sonography shows the absence of vascular flow in the solid components of the lesions; this finding simplifies and clarifies diagnosis in many patients [9]. CT and MR imaging displays multiple irregular, ill-defined lesions scattered throughout the involved liver tissue [10,11]. On MR imaging, multiple small round cysts with a solid component or solid component surrounding large and/or irregular cysts with multiple round cysts are thought to be characteristic of and highly specific for alveolar echinococcosis of the liver [10]. When radiological examinations do not help to distinguish hepatic AE from a tumor, histopathology is decisive. Histopathologic findings play an important role in differential diagnosis of the liver mass.

Main therapeutic strategies for hepatic AE are chemotherapy, surgery, and percutaneous drainage. Radical surgical treatment should be applied to all appropriate cases. In contrast to curative surgical management, partial resection and radiologic interventions, which are directed toward treating complications, significantly impair the effectiveness of liver transplantation. Furthermore, the transplantation procedure is more difficult and more complicated because hepatic artery thrombosis and biliary leakage occur frequently [12]. Therefore, in patients with symptomatic and advanced disease, liver transplantation should be considered as a treatment option [12,13]. Other important treatments are continuous anti-biotherapy and percutaneous drainage which are required in unsuitable cases for radical surgery. In our case percutaneus biliary drainage performed prior to liver transplantation.

In conclusion, AE is rare a disease and may cause severe complications. Therefore, early diagnosis is

crucial. Clinical suspicion and relevant radiologic, serologic, and histopathologic findings are important. Liver transplantation should be considered in the management of advanced liver alveolar echinococcosis Endoscopic and percutaneous drainage for biliary obstruction caused by hepatic AE has a palliative effect even in the late stages.

Conflict of Interest

All authors declare that no conflict of interest

Informed Consent

It was recevied the informed consent form from the patient.

Funding Information

There are no Funding Sources in this case report.

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