CASE REPORT

A case of Budd-Chiari syndrome associated with alveolar echinococcosis

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Abstract

Alveolar echinococcosis of the liver is caused by the tapeworm Echinococcus multilocularis. Budd-Chiari Syndrome (BCS) is a hepatic venous outflow tract obstruction and involves abdominal pain, hepatomegaly and high-gradient ascites. A low-gradient ascites in connection with the Syndrome rarely occurs in case of alveolar echinococcosis of the liver. Here we report a 20-year-old man with the syndrome and low-gradient ascites due to a huge liver mass. Further diagnostic examination revealed alveolar echinococcosis of the liver. Diuretic agents and albendazole were used as palliative therapy in the case.

Keywords: Echinococcus multilocularis, Budd-Chiari Syndrome, Alveolar echinococcosis.

Introduction

Human echinococcal disease can be classified in three forms according to morphologic appearance and the behaviour of the disease; cystic echinococcosis caused by Echinococcus granulosus, alveolar echinococcosis (AE) caused by Echinococcus multilocularis; and polycystic echinococcosis caused by Echinococcus vogeli or Echinococcus oligarthrus. Carnivores, mainly foxes, are the major reservoirs for the tapeworm Echinococcus multilocularis (E. multilocularis) which is mainly found in the northern hemisphere, including the eastern part of Turkey.[1]

The duration between the infection of this tapeworm and the onset of symptoms may take several years. The metacestode (larval) form of E. multilocularis almost always causes massive liver destruction and may also invade vascular structures of the liver, including the hepatic vein. The disease closely mimics primary liver cancer.[2]

Budd-Chiari Syndrome (BCS) is a hepatic venous tract obstruction characterised by hepatomegaly, abdominal pain, high-gradient ascites and rarely acute liver failure. Myeloproliferative diseases are responsible for the majority of the cases. The hepatic venous system may be mechanically compressed by large extraluminal masses, including alveolar cystic disease. The obstruction of the hepatic venous outflow causes peritoneal fluid accumulation due to elevated portal venous pressure.[3] Though BCS by E. Multilocularis is a rare disease, there have been some reports similar to the one presented.[4,5]

Case Report

A 20-year-old Turkish man presented to the emergency department (ED) with a two-week history of fever,

Table: Age and laboratory parameters of patient on admission.

Variable	level of laboratory parameters
Age (year)	20
Ascites protein (mg/dl)	5.6
Ascites albumin (mg/dl)	2.1
Ascites WBC (/mm^3)	800
Ascites lymphocyte (/mm^3)	560
Ascites neutrophils (/mm^3)	240
Ascites Glucose (mg/dl)	104
Ascites LDH (U/L)	21
Eosinophils (/mm^3)	1168
SAAG (g/dl)	1.4
Serum ascites-albumin gradient(g/dl)	1.0
WBC (/mm^3)	7300
Haemoglobin x 10 (g/dl)	11.2
PLT x 100 (/mm^3)	235
Serum albumin (g/dl)	3.1
Serum globulin (g/dl)	6.3
Serum total protein (g/dl)	9.4
Prothrombin time (seconds)	17.9
CRP (U/L)	30.5
Aspartate aminotransferase (U/L)	35
Alanine aminotransferase (U/L)	26
Alkaline phosphatase (U/L)	1458
Serum sodium(mmol/L)	131
Serum potassium (mmol/L)	3.8
Serum creatinine (mg/dl)	0.7
Serum glucose (mg/dl)	102
Gammaglutamyl transferase (U/L)	130
Direct bilirubin(mg/dl)	1.75

WBC: White Blood Count. LDH: Loctate Dehydrogenaase. SAAG: Serum Ascites-Albumin Gradient. PLT: Platelet. CRP: C-Reactive Protein.

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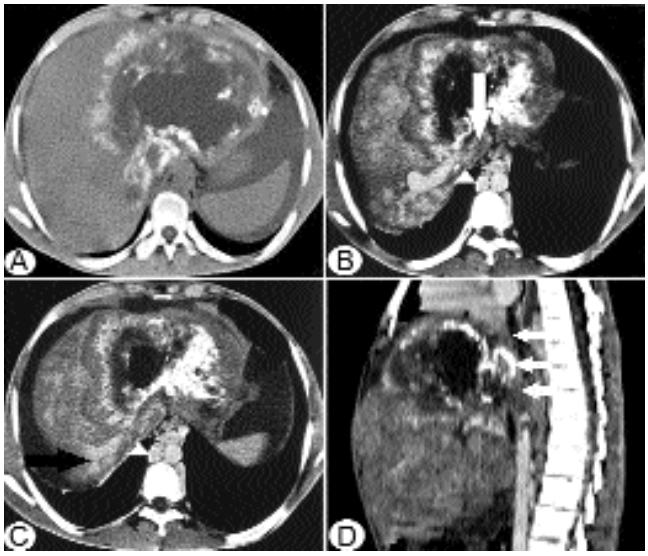

Figure: Contrast-unenhanced axial computed tomography (CT) image (A) revealed a huge calcific mass as well as massive ascites. Contrast-enhanced axial CT images in the portal venous phase (B, C). The middle and left hepatic vein could not be identified due to invasion. The right hepatic veins were not draining into the inferior vena cava because of the invasion of inferior vena cava (white arrow). The intra-hepatic collateral vessels (black arrow) associated with right hepatic vein could be seen. Dilatation of azygos vein was evident (white arrow head). Contrast-enhanced re-constructed sagittal CT image (D) shows an invasion of the vena cava inferior (white arrows).

jaundice and dull abdominal pain. He was born in a forest village and was working in a shipyard in eastern Turkey.

He looked ill with temperature of 38.2°C, blood pressure of 80/50mmHg, and a heart rate of 86/min. His conjunctiva were subicteric. There was tender hepatomegaly with tense ascites. There was also moderate splenomegaly. The remainder of the physical examination was normal.

Serum ascites-albumin gradient (SAAG) was calculated as 1.0g per decilitre (Table).

At the time of admission, an abdominal ultrasonography showed a huge hepatic mass, ascites and lack of the visualisation of the hepatic veins. An abdominal computer tomography (CT) scan also demonstrated an extremely large hepatic mass in the left lobe of the liver. It was 18x14cm in diametre, and there was non-uniform contrast enhancement of the liver parenchyma (Figure), an enlarged caudate lobe, and hepatic vein compression with massive ascites.

The histopathological examination of the liver mass revealed wide areas of necrosis and shadow alveolar vesicles in the biopsy material. The Enzyme-Linked Immunosorbent Assay (ELISA) test for E.multilocularis was also positive. A serum sample from the patient was tested using Em18-ELISA and was positive for E. multilocularis. The patient was considered to have BCS due to alveolar echinococcosis of the liver and was given spironolactone with low molecular weight heparin. At this time, albendazole (15 mg./kg. per day in divided doses) was also started as adjunctive therapy. In the second week of the therapy, the patient's ascites level had gradually reduced. He was referred for a liver transplantation and he is still waiting for the procedure at the hepatology unit.

Discussion

Alveolar echinococcosis is a zoonotic illness caused by infection with E. multilocularis which is generally found in the northern hemisphere. Liver disease caused by echinococcosis results from the significant destruction of the hepatic parenchyma in the larval stage of tapeworm. According to the PNM (P= parasitic mass in the liver; N= involvement of neighbour organs; and M= metastasis) classification of alveolar echinococcosis, stage III and IV diseases are associated with hepatic venous involvement as well as BCS.[6]

The majority of patients suffer right upper abdominal pain, jaundice or constitutional symptoms like fatigue or weight-loss. The most frequent complications of the disease are biliary cholangitis and sepsis. The laboratory findings of an E. multilocularis infection of the liver include elevation of liver-related transaminases, higher cholestasis enzymes, hyperglobulinaemia, eosionophilia, higher C-reactive protein levels and a positive result to the serum ELISA test.[7] In the current case, all the laboratory findings were compatible with echinococcal liver disease.

Radiologic studies remain the gold standard for the diagnosis of E. multilocularis of the liver, especially for hepatic venous tract involvement. It is reported that in patients with an E. multilocularis infection the ultrasonography of the liver usually shows a space-occupying mass with necrotic fluid as well as granular echogenic area with an acoustic shadow.[8]

The characteristic features of the abdominal CT reveals scattered calcifications, central necrosis and small peripheral cysts.[9]

For SAAG, the following formula is used: Corrected SAAG= uncorrected SAAG x 0.16 x (serum globulin [g/dL]+2.5).[10] In patients with BCS, the SAAG is higher than 1.1 g/dL, with the total protein level in the ascitic fluid usually being more than 2.5g per decilitre.[11]

However, the presented case had a low SAAG (<1,1 g/dl.) which was not compatible with BCS.

According to PNM classification,[6] our patient was in the surgically incurable stage. A long-term medical therapy with albendazole can provide palliation in patients with advanced stage disease.[12] As such, we did not adopt a surgical approach for the case, and started a long-term palliative treatment with albendazole (15 mg/kg in divided doses).

Conclusion

A diagnosis of BCS must always be considered in all patients together with alveolar echinococcosis of the liver, especially eosinophilia and cystic liver masses. A corrected SAAG may provide an effective diagnostic tool for patients who have both ascites and hyperglobulinaemia.

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