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# Living Donor Right Lobe Liver Transplantation as a Treatment for Hepatic Alveolar Echinococcosis: Report of Three Cases

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## SUMMARY

*Echinococcus alveolaris* is a parasite from tenia family which causes tumor-like lesions in the livers of infected people. If it is not diagnosed in the early stage of the disease, it frequently causes multiple cysts in the liver. The clinical importance of the disease is rapid progression, infiltration into different tissues like a malignant tumor and capacity of creating metastatic masses. The disease could be treated either by surgical resection or liver transplantation. The resection of

the cystic disease is the preferred treatment method. In cases where resection is not possible, liver transplantation is the choice of treatment. Here we present three cases which were admitted to the hospital with unresectable hepatic alveolar echinococcosis and treated by liver transplantation successfully. Patients for whom surgical resection is not possible, we recommend liver transplantation as the treatment method.

**Key Words:**  
*Echinococcus alveolaris*, Liver transplantation.

## INTRODUCTION

Hepatic alveolar echinococcosis is a parasitic infection caused by the larval stage of *echinococcus alveolaris*. The infection is endemic in some parts of the world like Eastern Europe, North America, Turkey, and Eastern Asia. The prevalence of *Echinococcus alveolaris* is nearly 3 % in all cystic liver diseases.(1)

The clinical importance of *echinococcus alveolaris* infection is the infiltrative pattern of the lesions in the liver.(2) If not diagnosed in the early stage, the disease progression might occur rapidly and cause a spectrum of changes including giant hepatic tumor-like lesions. The patient might suffer from jaundice as a result of biliary obstruction. Besides due to portal hypertension caused by intrahepatic venous outflow obstruction, ascites, esophageal variceal bleeding could be observed. The treatment method is surgical resection of the tumor-like lesion. When the liver lesion is too infiltrative and surgical resection is not feasible, liver transplantation has to be considered as the treatment method.

In this article we aimed to present three cases which were treated successfully by living donor liver transplantation and followed without any complications.

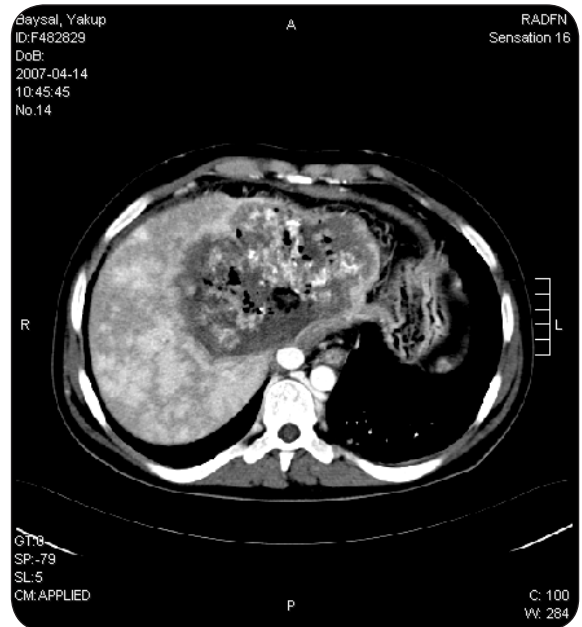
## CASE 1

Twenty-six year-old male patient was admitted to the hospital with the complaint of intermittent abdominal pain which was present for two months. He was an inhabitant from a city in Eastern Anatolia. He complained also about nausea and vomiting. The physical examination revealed 2 cm hepatomegaly below the

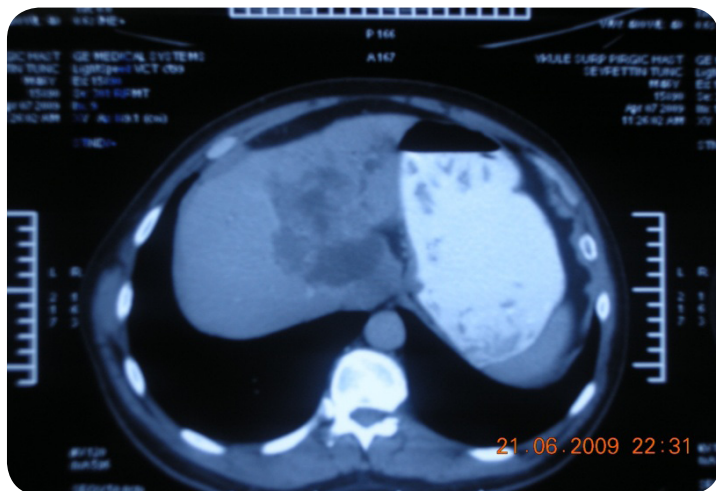
costal margin. Biochemistry and complete blood count was normal. The abdominal computerized tomography demonstrated diffuse infiltrative cystic and solid lesions (largest was 9 x 7 cm) which was located in the left lobe of the liver extending to the right lobe and invaded intrahepatic vascular structures and biliary tree. (Figures 1, 2). The thorax computerized tomography and cranial MR showed no extrahepatic manifestation of the disease. The result of indirect hemagglutination test for echinococcosis were positive at 1/32. The patient was scheduled for surgical resection. Since the lesions were diffuse and involve both the biliary and vascular system, live donor liver transplantation was planned if the lesion was evaluated as unresectable preoperatively. Then, his elder brother who was 38 years old was also prepared for possible living liver donor operation. The total liver volume of the donor was 1440 grams; the left lobe was 460 grams and the right lobe was 980 grams. The remnant liver volume was 32% and the graft recipient weight ratio was 1.43. During the preoperative period, the patient was treated with 15 mg/kg/day albendazole for two weeks. The patient was operated. In the exploration of the abdomen, there were no extrahepatic manifestation of the parasitic disease. The dissection of vena cava and the hepatoduodenal ligament was impossible due to invasive tumor-like lesion. Total hepatectomy was done after clamping suprahepatic vena cava, infrahepatic vena cava and hepatoduodenal ligament to prevent massive hemorrhage. On back table left hepatectomy was performed. During this process, aortic graft interposition was done between superior and inferior



**FIGURE 1.** Demonstrating the liver lesions in the arterial phase of abdominal computerised tomography.



**FIGURE 2.** The cystic and solid liver lesions that can be seen in the venous phase of the abdominal computerised tomography.



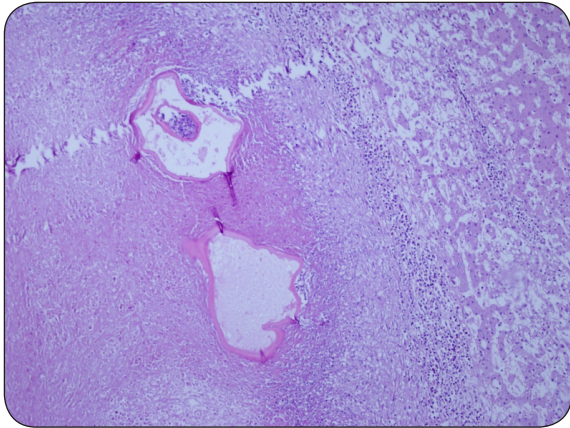
**FIGURE 3.** The infiltrative cystic and solid lesion could be seen in the abdominal computerised tomography



**FIGURE 4.** Macroscopic image of the resected specimen. Solid and cystic tumor like lesions could be visualised.

vena cava. Portocaval shunt was done. The right lobe of the liver was brought to the surgical area and right hepatic vein was anastomosed to aortic graft and right portal vein was anastomosed to recipient portal vein and reperfusion was maintained. However, bleeding from remnant liver cut surface could not be stopped so the liver donor of the patient was prepared for surgery. The superior and inferior vena cava was clamped; portal vein was clamped and the right liver lobe was resected. Consequently, portocaval shunt was done again. Meanwhile right lobe living donor hepatectomy operation was performed and right lobe liver graft was perfused on the back table. The recipient aortic graft was prepared for triangular veno-venous anastomosis and hepatic vein anastomosis was done using 4/0 prolene sutures with continuous intraluminal evertting technique. The portocaval shunt was clamped. The liver graft had two portal veins. The portal vein orifices was sutured on the back-table and portal vein anastomosis was done using 5/0 prolene using continuous intraluminal evertting technique. The right hepatic artery of the graft was anastomosed to right hepatic artery of the recipient with 8/0 interrupted sutures. The bile duct anastomosis was done with 6/0 prolene in duct-to-duct fashion.

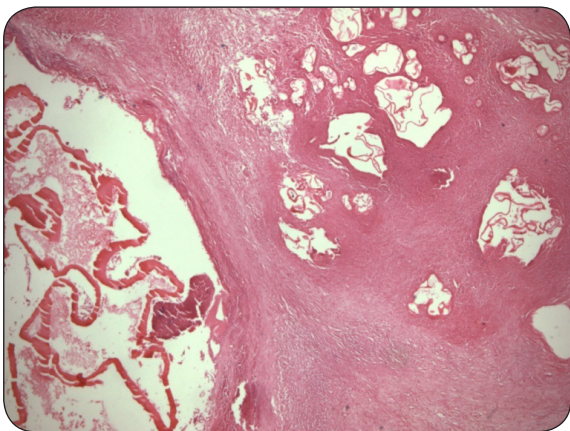
The operation was performed successfully lasting 12 hours. The patient was discharged from the intensive care unit on the fourth day and from the transplant service on the 15th day of the operation. The pathology report revealed liver abscess and granulomatous inflammatory tissue response due to echinococcus alveolaris infection. The immunosuppressive regimen comprised methylprednisolone 5 mg/kg, tacrolimus, 0.1 mg/kg. The steroid dosage was gradually decreased and discontinued within 3 months after the operation. Albendazole therapy of 15 mg/kg/day was prescribed and continued for two years. Now, he has been followed up from the outpatients' clinic with well status and no recurrence for 43 months.



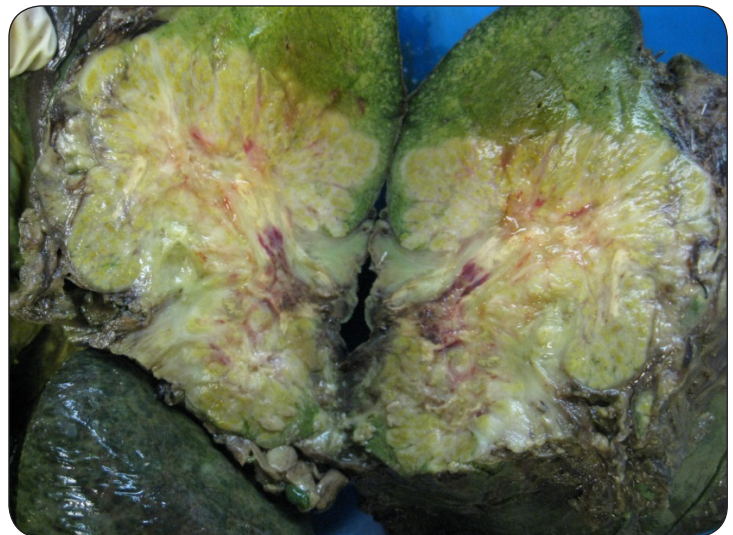
**FIGURE 5.** Fibrotic and hyalinized tissue surrounding small cysts were observed HEX200.



**FIGURE 6.** Macroscopic appearance of Case 2 in hepatectomy specimen. Huge mass with small cysts is seen.



**FIGURE 7.** Microscopic appearance of cysts and buds of the germinal layer in hyalinized tissue. HEX200.

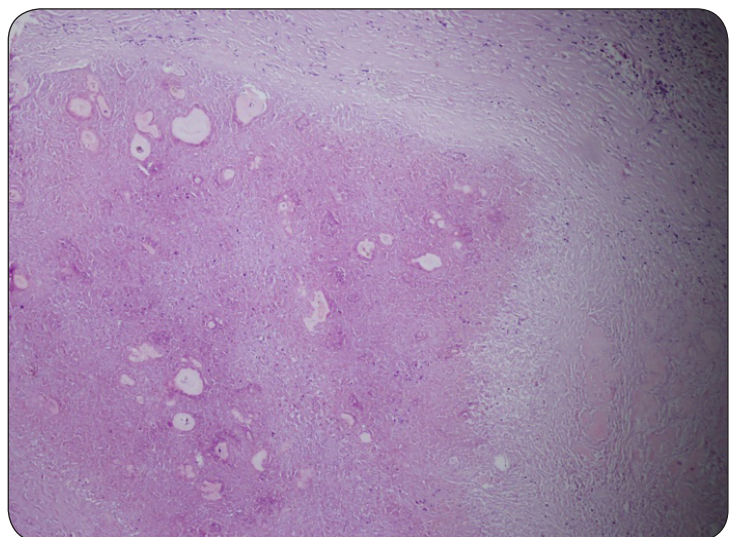


**FIGURE 8.** Macroscopic appearance of 3<sup>rd</sup> case that has large mass with small cysts.

**CASE 2**

Forty-six year-old male patient was admitted to the hospital with the complaints of abdominal pain, abdominal distention and fatigue which were present for three months. He was from a village in the northeast Anatolian region. The physical examination of the patient revealed hepatomegaly 3 cm below the right costal margin. Biochemical tests were normal except GGT and albumin. (GGT was 209 IU and albumin level was 2.4 mg/dl.) Serologic markers were found negative for hepatitis B and hepatitis C virus. The abdominal Magnetic Resonance Imaging (MRI) revealed a cystic and solid lesion which located in the left lobe extending to right and caudate lobes and surrounding the inferior vena cava which was approximately 10 x 8 cm. (Figure 3) The thorax computerized tomography and cranial MR showed no extrahepatic manifestation of the disease. The result of indirect hemagglutination test for echinococcosis were positive at 1/32.

As the liver lesion was located in the left lobe we attempted left lobe liver resection. However, during exploration it was observed that the cystic lesion surrounded the vena cava-hepatic vein junction that was preventing the dissection of it. It was not possible to put a clamp to the inferior vena cava and hepatic vein. Therefore decision of performing liver resection was technically impossible. So, the patient was scheduled for living donor liver transplantation. His nephew who was 23 years



**FIGURE 9.** Microscopic features of 3<sup>rd</sup> case. Small cysts in hyalinized tissue. HEX100.

old was prepared for the donor operation. The total liver volume of the donor was 1228 grams; the left lobe was 423 grams and the right lobe was 805 grams. The remnant liver volume was 34% and the graft recipient weight ratio was 1.16. During the preoperative period, the patient was treated with 15 mg/kg/day albendazole. After 7 days medical therapy, the patient was operated. During the exploration, there were no extrahepatic manifestation of the parasitic disease. As the cystic lesions in the liver was found to surround the hepatic veins and vena cava inferior, sternotomy was performed. The hepatic vein anastomosis was performed to right atrium-inferior vena cava junction with end-to-end technique using 4/0 prolene sutures. End-to-end portal vein anastomosis was performed with 5/0 prolene sutures. Graft right hepatic artery was anastomosed to recipient right hepatic artery with interrupted sutures. The bile ducts were anastomosed in duct to duct fashion with 6/0 prolene sutures using interrupted technique.

The patient was taken to the intensive care unit after the surgery and was followed for three days and taken to the transplant service afterwards. The immunosuppressive regimen comprised methylprednisolone 5 mg/kg, tacrolimus, 0.1 mg/kg. The steroid dosage was gradually decreased and discontinued within 3 months after the operation. Albendazole treatment was continued 15 mg/kg/day postoperatively. On the 14 day of the operation the liver function tests were within normal levels and the patient was discharged from the hospital. The patient is still followed by polyclinic controls and no recurrence has been detected.

In pathologic examination of the hepatectomy specimen, grossly there was large infiltrative mass in the left lobe (**Figure 6**). Typical cysts and protrusions ("buds") of the germinal layer were detected in hyalinized and necrotic background in the samples of this mass microscopically, as more solid structures growing by infiltration and destruction. In some areas, the daughter cysts bud outwardly from the parent cyst. (**Figure 7**)

### CASE 3

Thirty-one-year-old-male patient was admitted to our hospital with the complaints of abdominal pain, fever, jaundice and abdominal distention attacks which were present for three years. The patient was an inhabitant from eastern rural part of the country. His physical examination revealed 10 cm of hepatomegaly below right costal margin. The biochemical tests revealed AST:103 IU/L, ALT:103 IU/L, GGT:307 IU/L, Alp:554 IU/L, total bilirubin:9.3 mg/dL, direct bilirubin 9.17 mg/dL, albumin: 3.1 g/dL. Serologic markers were found negative for hepatitis B and hepatitis C virus. The abdominal ultrasound revealed a cystic and solid mass which was nearly 10 cm in diameter in the right lobe of the liver. The abdominal CT revealed a multiloculated cystic mass which was in 10 x7 cm in diameter, and filling the right liver lobe. The immune hemagglutination test by ELISA for EA was positive at 1/32.

The patient was scheduled for liver transplantation because the patient was suffering from recurrent cholangitis attacks. Secondly it was impossible to perform a liver resection to the patient. The third and last reason for performing liver transplantation was the absence of extra-hepatic alveolar echinococcosis. His wife who was 24 years-old was prepared for the donor operation. The total liver volume of the donor was 1836 grams; the left lobe was 605 grams and the right lobe was 1231

grams. The remnant liver volume was 32% and the graft recipient weight ratio was 1.16. During the preoperative period, the patient was treated with 15 mg/kg/day albendazole. After 7 days medical therapy, the patient was operated.

During the exploration, there were no extrahepatic manifestation of the parasitic disease. The hepatoduodenal ligament was dissected and the right and the left hepatic arteries were ligated and cut. The posterior accessory inferior hepatic veins were ligated and the liver was devascularised by Piggy-back technique over the vena cava. The right hepatic vein, middle and left hepatic veins and the portal vein were clamped and the recipient hepatectomy was completed. The right lobe graft was brought to the surgical area after the perfusion on the back table. The hepatic vein anastomosis was performed to right hepatic vein stump with end-to-end technique using 4/0 prolene sutures. End-to-end portal vein anastomosis was performed with 5/0 prolene sutures. Graft right hepatic artery was anastomosed to recipient right hepatic artery with interrupted sutures. The bile ducts were connected as duct to duct with 6/0 prolene sutures using interrupted technique.

In the gross examination of the hepatectomy specimen, a large infiltrative mass like a tumor was found in the left lobe (**Figure 8**). In microscopic examination, a large hyalinized and necrotic tissue was present with destruction of normal liver. In many areas, small cysts with germinal layer were seen in this hyalinized tissue (**Figure 9**)

No blood was transfused to the patient. The anhepatic phase lasted 31 minutes. The patient was taken to the intensive care unit and was followed one day and taken to the transplant service afterwards. Albendazole treatment was continued 15 mg/kg/day postoperatively. On the 11th day of the operation the liver function tests were within normal levels; all the drains were taken out and the patient was discharged from the hospital. The patient is still followed by polyclinic controls and no recurrence has been detected.

### DISCUSSION

Hepatic alveolar echinococcosis is a parasitic insidious disease which may cause serious cystic and solid lesions in the liver. It might resemble malignant diseases and metastasize to distant organs. The clinical findings could include jaundice, liver abscess, cholangitis, sepsis, Budd-Chiari syndrome. Many complications might be seen as a result of the disease requiring radiological and surgical interventions. Some of them are cholangitis attacks as a result of drainage of cysts into bile ducts; cavitory infections; cholestasis, invasion of distant organs and portal hypertension.(3,4) If liver resection is not possible, liver transplantation should be considered as an alternative method of treatment. (5,6,7)

In preoperative assessment, there was no extrahepatic involvement in our patients. The clinical importance of the parasitic disease caused by echinococcus alveolaris is the aggressiveness of the disease and the metastasis of the parasitic lesions to other tissues. The liver, lungs, brain, bones, muscle, spleen, heart and lymph nodes are the major organs that cystic lesions could be infiltrate.(8,9) Without treatment, it was reported that 90% of the patients would die within 10 years.(10,11,12)

Surgical radical resection is the usual treatment for hepatic alveolar echinococcosis. Before resection medi-

cal treatment with benzimidazole derivatives (Albendazole 2x400 mg/day) should be started at the time of diagnosis with the dose of 10-15 mg/kg/day. This medicine prevent the maturation of the larval stage of the parasite and do not completely treat the disease. (13,14,15) According to Sato et al.(14) radical resection could be practiced in these situations:

#### Lesions that do not invade vena cava, portal vein or hepatic artery

Lesions that invade less than 3 segments of the liver

Distant metastases should not be present

The age of the patient should be smaller than 75

The residue liver tissue should be sufficient for the patient

Patients for whom surgical resection is not possible, partial resection and radiological interventions could be used but these methods are temporary and do not solve the main problem. Also, these interventions might impair the effectiveness of better methods like liver transplantation. The conditions for a patient with the diagnosis of echinococcus alveolaris for LT are: (a) recurrent life-threatening cholangitis or severe liver insufficiency, (b) inability to perform radical liver resection and (c) absence of extra-hepatic locations.(17)

The stage of the disease at the time of admission is an important factor affecting treatment. All of our cases were admitted to the hospital with irresectable lesions. After assessing with CT angiography, it was decided that resection would not be possible and the patients were scheduled for liver transplantation from living liver donors.

The resection of the diseased liver might be hard as a

result of invasion of surrounding tissues. Radical resection of the entire parasitic lesion should follow the rules of tumor surgery. Non-radical surgery does not offer advantages over conservative treatment. (18)

Similarly, venovenous bypass procedure might be helpful to minimize the risk of bleeding during the operation.(5)

According to Bresson-Hadni et al.(19) the postoperative recurrence is influenced by three factors; extrahepatic dissemination of the disease preoperatively, medical treatment preoperatively and postoperatively and high dose immunosuppression. The evaluation of distant organs should be done precisely with conventional computerized tomography. None of our patients had distant metastasis.

The rate of recurrence seems high in patients receiving high doses of immunosuppression. Both of our patients were given calcineurin inhibitors (tacrolimus) and steroids and the doses were adjusted according to blood levels of the medicine. The steroid dosage was decreased slowly in 6 months and the patient received only tacrolimus thereafter. In order to prevent disease recurrence, albendazol was given 10 mg/kg/day and continued for two years. During the follow-up period, cranium, thorax and abdominal CT should be done in case of high index of suspicion.

As a result living donor or cadaveric donor liver transplantation should be considered for patients with advanced hepatic echinococcosis. The cranium, thorax and abdominal imaging must be done preoperatively. The medical therapy should be done during the preoperative and postoperative period and the dosage of immunosuppression should be kept in low levels.

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