Alveolar echinococcosis with portal vein thrombosis: An unusual cause for liver transplantation

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Alveolar echinococcosis (AE), caused by larval stage of *Echinococcus multilocularis* (*E. multilocularis*), is a very aggressive and potentially fatal infestation which affects primarily the liver and can metastasize to any part of the body through hematogenous spread. It displays a prolonged latency period before clinical presentation, behaving like a slowly growing liver cancer with subsequent invasion of liver tissues and metastatic dissemination. The disease produces liver necrosis invading not only the hepatic and biliary tissues, but also the vessels of the liver, inferior vena cava, and diaphragm. Surgical resection of the parasite at an early stage of infection may provide favorable prospects for cure, but due to the long clinical latency, many cases are diagnosed at an advanced stage, so that partial liver resection can be performed in only 40% of patients [1]. Live-donor liver transplantation (LT) can provide patients with advanced-stage (AE), the best chance for long term disease-free and overall survival [1,2]. We herein report a case of hepatic echinococcosis who successfully underwent live-donor LT in our institution.

A 31-year-old female from Kyrgyzstan came to our center with complaints of vague discomfort and right upper quadrant abdominal pain since one year which increased since last two months with history of anorexia, weight loss and jaundice. On physical examination, her vital signs were within normal limits with yellowish discoloration of skin and visible mucosa. On examination of the abdomen, the liver was enlarged up to 12 cm below right costal margin with firm and irregular surface. Laboratory biochemical analysis revealed total bilirubin, direct bilirubin, serum aspartate aminotransferase, alanine aminotransferase and alkaline phosphatase concentrations of 1.1 mg/dL, 0.4 mg/dL, 75 U/L, 104 U/L and 1 448 U/L, respectively. Tumor markers were found to be negative. Abdominal ultrasonography with doppler revealed an enlarged liver showing heterogeneous echotexture with multiple space occupying lesions. Many ill-defined echogenic areas were seen distributed in the right lobe of liver forming a conglomerate mass like lesion and another echogenic space occupying lesions measuring 51 mm × 65 mm in the left lobe. The right main portal vein showed low level echoes without any uptake of color suggestive of thrombus. Computed Tomography of the abdomen revealed a diffusely infiltrative geographical hypodense lesion within the liver parenchyma with multiple amorphous areas of calcification almost completely replacing the left lobe of liver (Figure 1A). Multiple other similar infiltrative nodular areas were seen further in the posterior segment of the right lobe, the largest lesion measuring 66 mm × 52 mm. This lesion was encasing, attenuating and occluding the arterial, hepatic venous, the portal venous divisions and its territory. The main portal vein at the level of the liver hilum was significantly occluded. In the meanwhile, keeping the diagnosis of echinococcus in mind, a serum sample was sent for *E. multilocularis* antibodies using enzyme-linked immunosorbent assay, and serology was noted to be positive. Based on serology and imaging studies, hepatic Alveolar echinococcosis was diagnosed and due to extensive liver involvement with portal vein thrombosis, live donor LT was offered as a curative treatment choice. The patient underwent live donor related LT. Gross examination from the explanted liver, revealed smooth and focally bossedated external surface with whitish areas. Nearly half of the cut surface of the right lobe was replaced by whitish firm areas, largest measuring 105 mm × 95 mm × 80 mm, surrounding the major portal tracts and ducts (Figure 1B). Focal areas of necrosis were identified. Left lobe was almost completely replaced by firm areas. On histopathological examination, extensive areas of necrosis were seen with multiple necrotising granulomas around small, dead and viable ectocysts with scolices (Figure 1C). Strong periodic acid-schiff (PAS) positive ribbon like and lamellated structures were seen (Figure 1D). Adjacent hepatocytes showed atrophy with moderate macro and microvesicular steatosis. Sections from gall bladder wall showed focal necrosis and granulomas. Hepatectomy specimen was compatible with diffuse *Echinococcus alveolaris* infection. The postoperative period was unremarkable, and the patient was discharged from the hospital 18 d after undergoing live-donor LT. The immunosuppressive
regimen consisted of methylprednisolone, 5 mg/kg and tacrolimus, 0.1 mg/kg. Albendazole 400 mg/d was also prescribed. The patient is doing well 6 months follow-up.

Hepatic Alveolar echinococcosis is a rare slow growing tumor like lesion, primarily involving liver and has a potential to infiltrate adjacent organs and distant sites as brain, lungs through hematogenous spread. It is a rare cause of LT. Correct and timely diagnosis of the hepatic AE will allow for appropriate treatment and thereby improving overall survival. Abdominal ultrasonography is the first-line imaging modality for evaluation of patients, in whom AE is suspected and is also used to monitor efficacy of medical therapy [3]. CT is always performed because it has a high sensitivity (94%) [4]. Radical surgery is recommended for treatment, whenever feasible in early stage AE. Living donor or cadaver donor liver transplantation should be considered in the management of advanced hepatic alveolar echinococcosis. However, surgical procedures and radiologic interventions which are directed toward treating complications may pose technical difficulties for future liver transplantation [1]. Therefore, in patients with symptomatic and advanced disease, liver transplantation should be considered as a preferred treatment option. Extrahepatic invasion by the lesion must be carefully sought during the preoperative period. In addition, the dosages of immunosuppression medications should be kept at low levels, and medical therapy must be administered before and after surgery. Even after the LT, for most patients this is still a chronic disease requiring long-term, mostly lifelong chemotherapy under medical supervision. The possibility of ongoing disease must be kept in mind. Regular follow-up of patients should include a systematic radiological evaluation, monitoring of anti–E. multilocularis by enzyme-linked immunosorbent assay for assessment of disease status and evaluation of drug side effects.

To conclude, we report a case of hepatic echinococcosis which successfully underwent live-donor LT. It is therefore a feasible option in end-stage cases that are considered intractable.

**Conflict of interest statement**

We declare that we have no conflict of interest.

**References**


