



Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Advanced alveolar echinococcosis disease associated with Budd–Chiari syndrome



Vural Soyer^{a,*}, Cengiz Ara^a, Faik Yaylak^b, Barış Sarıncı^a, Mustafa Ozsoy^c, Okay Koç^d, Sezai Yılmaz^a

^a Inonu University, Faculty of Medicine, Department of General Surgery, Malatya, Turkey

^b Dumlupınar University, Faculty of Medicine, Department of General Surgery, Kutahya, Turkey

^c Afyon Kocatepe University, Faculty of Medicine, Department of General Surgery, Afyon, Turkey

^d Diyarbakir Research and Training Hospital, Diyarbakir, Turkey

ARTICLE INFO

Article history:

Received 31 October 2014

Received in revised form

25 December 2014

Accepted 6 January 2015

Available online 9 January 2015

Keywords:

Alveolar echinococcal disease

Budd–Chiari syndrome

Living donor liver transplantation

Clinical imaging

Computed tomography

ABSTRACT

INTRODUCTION: Alveolar echinococcal disease of the liver is rare. *Echinococcus multilocularis* is responsible for the development of the related clinical conditions. Advanced disease may result with serious complications such as end stage liver disease and Budd–Chiari syndrome.

PRESENTATION OF CASE: In this presentation, a 28 years–old woman who was a case with advanced alveolar echinococcosis complicated with a Budd–Chiari syndrome and was performed successful living donor liver transplantation, has been demonstrated with clinical and radiological images.

DISCUSSION: Initially there may be no clinical evidence of the disease in humans for years. Severity and fatality are the significant characteristics of the natural history. Extension to the surrounding tissues and metastasis of the parasitic mass may be observed. Prevention is essential in disease control. Serologic assay may identify the parasite. However, early diagnosis is rare. Staging is based on radiologic imaging. Some patients with advanced disease may require surgery. Hepatic resection and liver transplantation are accepted procedures in selected patients.

CONCLUSION: The importance of early diagnosis to prevent advanced complications such as development of Budd–Chiari syndrome and metastasis has been underlined.

© 2015 The Authors. Published by Elsevier Ltd. on behalf of Surgical Associates Ltd. This is an open access article under the CC BY-NC-SA license (<http://creativecommons.org/licenses/by-nc-sa/4.0/>).

1. Introduction

Alveolar echinococcus (AE) is a rare parasitic disease of the liver [1]. *Echinococcus multilocularis* (*E. multilocularis*) is responsible for the development of the related clinical conditions. The natural history of *E. multilocularis* has been documented and was reported previously [2]. After ingestion of eggs the parasite is carried to the liver, where it settles, via portal system. Humans are intermediate hosts and larval stage of the parasite causes a slow progressive necrosis and fibrosis in the host tissue. AE is diagnosed with various clinical manifestations due to parenchymal liver destruction, biliary and hepatic venous involvement and local expansion/invasion or distant spread [1].

Budd–Chiari syndrome (BCS) is defined due to AE as the occlusion of major hepatic veins (HV) and retro hepatic inferior vena cava (IVC) [3,4]. This complication of the AE may be difficult to treat with advanced surgical techniques [5]. Early recognition and diagnosis of the AE is essential in preventing and/or treatment of complications.

In this case report, the aim was to present progressive clinical findings of AE disease complicated with Budd–Chiari syndrome.

2. Presentation of case

A 28 years–old woman with a Child C Score and MELD score eight was scheduled for living donor liver transplantation for chronic liver disease and Budd–Chiari syndrome. WBC: 18.6 10³/ML; HB: 10.3 g/dL; HTC: 32.8%, eosinophil: 0.5%, PLT: 547 10³/ML, INR: 1.3 D-dimer: 451 ng/dL, creatinine: 0.44 mg/dL, albumin: 2.6 g/dL; total/direct bilirubin: 1.5/1.1 mg/dL, AST/ALT: 15/8 U/L ALP: 162 U/L, GGT 88 U/L. Budd–Chiari syndrome was diagnosed with preoperative computed tomography (CT), which has demonstrated hepatomegaly, caudate lobe hypertrophy, occlusion of the hepatic veins and the presence of extensive ascites (Fig. 1a). A cystic lesion (21 × 10 × 14 cm) was observed in segments 6, 7, 8 and 4 with a solid component. Peripheral and central calcifications were observed. Extension of the lesion to the right diaphragm and compression of retro hepatic segment of vena cava was observed (Fig. 1b). Previous CT obtained one year before was comparable with the size of mass in segment 6 (10 × 7 × 11 cm). CT evaluation

* Corresponding author. Tel.: +90 5337785793.

E-mail address: hvuralsoyer@yahoo.com (V. Soyer).

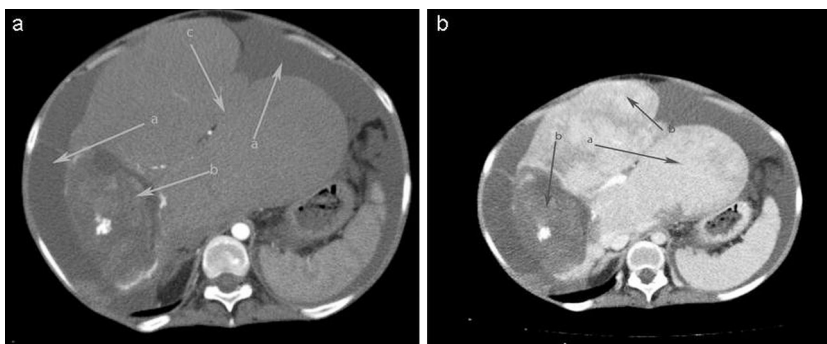


Fig. 1. (a) Contrast enhanced computed tomography demonstrating a – massive ascites; b – a calcified mass in the right liver; with c – the absence of hepatic veins and d – occlusion of the vena cava. (b) a – Significant hypertrophy of the caudate lobe of the liver and b – cystic mass in contrast enhanced computed tomography.

for thorax was not significant. CT angiography for celiac and hepatic arterial system was not significant. During recipient operation ten liters of ascites was aspirated. Perioperative management was intense with volume replacement (including transfusions of blood products and albumin transfusions), cardiopulmonary and renal support. Liver was enlarged and significant caudate lobe hypertrophy was noted (Fig. 2a). Dissection was difficult due to extensive perihepatic fibrosis (Fig. 2b). Total hepatectomy was only possible after infra and supra hepatic clampage and partial anterior wall resection of retro hepatic cava with meticulous surgical technique (Fig. 3). Retro hepatic vena cava was resected and replacement was with a cryopreserved IVC graft. Right lobe living donor liver transplantation was completed. Postoperative course was not fair.

3. Discussion

Within the last decade, various reports have emerged indicating the role of urbanization and increased interaction of the human and wilderness in development of AE. Currently cases from Europe have been reported. However, AE was previously considered as a rural disease and most cases were primarily from Asia. In order to improve epidemiological control of the disease World Health Organization (WHO) has supported consensus studies on the diagnosis, classification and treatment of AE [2]. Thus, demographic features are important for the suspicious patients of alveolar echinococcosis. In this case report, the patient was presented from a region where

a close relation of human and wilderness is well known (southeast of the country).

Clinical features of the overt disease are similar to the malignant liver tumors and commonly related to the extension of parenchymal destruction, involvement of the biliary and venous systems. Abdominal pain, weight loss, abdominal mass, icterus may be observed. Portal hypertension and ascites may indicate the development of Budd–Chiari syndrome in these patients [5]. However, there is no specific or characteristic clinical finding of the diseases. In our case and insidious progression of the disease was observed. Clinical presentation was with an advanced and complicated liver disease due to parasitic invasion. Increased eosinophil counts and elevated liver transaminases and cholestatic enzymes may be observed. Even, a serologic assay (an enzyme linked immunosorbent assay-ELISA) with high sensitivity rate (range 84–90%) is available to detect the worm. However, diagnosis is only possible when the clinical findings are significant [6]. Imaging techniques including ultrasonography (US), computed tomography (CT) and magnetic resonance imaging (MRI) has significant role in the diagnosis. The WHO staging system depends on the radiological imaging features. WHO classification system stages the disease based on parasite neighboring metastasis [7,8].

The primary of treatment is early diagnosis. Albendazole has critical importance in medical therapy. Hepatic resection and liver transplantation have been accepted surgical techniques in therapy [9,10]. However, development of BCS in some patients may complicate the clinical course further. In such conditions, additional advanced surgical techniques and procedures such as vena cava



Fig. 2. Intraoperative image of (a) the parasitic mass in the right liver and (b) caudate lobe hypertrophy.

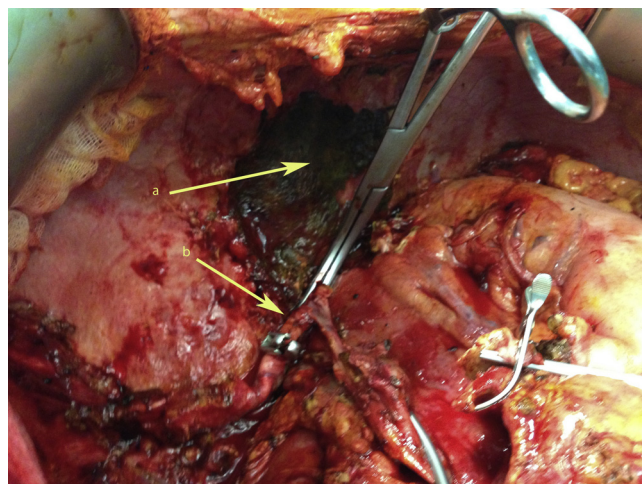


Fig. 3. Operative field demonstrating (a) the invasion side of the diaphragm; and the (b) the resected vena cava.

resection and reconstruction with various vascular grafts may be required [5]. However, LDLT for AE with BCS may be a challenging procedure with several difficulties [11]. It is clear that advanced diseases and surgery may result in increased morbidity and mortality. In this case the patient was listed for cadaveric liver waiting list. However, limited organ donation causes long waiting periods. A living donor for LDLT was the only alternative for this patient.

In this case cadaveric liver graft was not available for the patient. With availability of a living donor, LDLT was considered to be acceptable for this young patient. Meticulous and intensive preoperative preparing, surgical technique and intensive care support were available for the patient. However, due to severe hemodynamic alterations and significant technical difficulties were observed due to advanced and complicated disease. Especially, the need of inferior vena cava replacement may require a prolonged total vascular clampage (of the vena porta and inferior vena cava) and anhepatic phase during transplantation surgery. Significant effects advanced surgery and interventions should be weighted with patient toleration and response individually. Thus, this case once again underlines the importance of early diagnosis of AE disease to lower the need for surgery and prevent surgical morbidity and mortality.

4. Conclusion

In this case report an advanced AE has been presented. Outcomes may be worse even with advanced surgical techniques. Early diagnosis is essential to prevent severe complications such as development of BCS and metastasis.

References

- [1] M. Piarroux, R. Piarroux, R. Giorgi, J. Knapp, K. Bardonnnet, B. Sudre, J. Watelet, J. Dumortier, A. Gérard, J. Beytout, A. Abergel, G. Mantion, D.A. Vuitton, S. Bresson-Hadni, Clinical features and evolution of alveolar echinococcosis in France from 1982 to 2007: results of a survey in 387 patients, *J. Hepatol.* 55 (2011) 1025–1033.
- [2] WHO/OIE, Manual on Echinococcosis in Humans and Animals: a Public Health Problem of Global Concern, WHO, 2001.
- [3] H. Bedioui, K. Nouira, S. Ayadi, A. Daghfous, M. Bakhtri, R. Ksantini, F. Chebbi, F. Fteriche, M. Jouini, M. Kacem, M. Emna, Z.B. Safta, Budd–Chiari syndrome secondary to hepatic echinococcosis, *Gastroenterol. Clin. Biol.* 31 (2007) 721–724.
- [4] I.A. Rossi, D. Delay, S.D. Qanadli, A. Jaussi, Inferior vena cava syndrome due to *Echinococcus multilocularis*, *Echocardiography* 26 (7) (2009) 842–846.
- [5] R. Mamedov, N. Novruzov, A. Baskiran, F. Yetisir, B. Unal, C. Aydın, N. Bayramov, C. Kayaalp, S. Yilmaz, Living donor liver transplantation with replacement of vena cava for echinococcus alveolaris: a case report, *Int. J. Surg. Case Rep.* 5 (3) (2014) 169–171.
- [6] E. Brunetti, P. Kern, D.A. Vuitton, Writing panel for the WHO-IWGE. Expert consensus for the diagnosis and treatment of cystic and alveolar echinococcosis in humans, *Acta Trop.* 114 (2010) 1–16.
- [7] P. Kern, H. Wen, N. Sato, D.A. Vuitton, B. Gruener, Y. Shao, E. Delabrousse, W. Kratzer, S. Bresson-Hadni, WHO classification of alveolar echinococcosis, *Parasitol. Int.* 55 (Suppl) (2006) S283–S287.
- [8] K. Deniz, S. Nazlım, T.E. Patiroglu, E. Deniz, T. Artış, A. Karaman, S. Yazar, Retrospective evaluation of the alveolar echinococcosis cases between 1980–2010 in Erciyes University Hospital, *Türk. Parazitol. Derg.* 36 (1) (2012) 33–36.
- [9] A.C. Dülger, M.E. Küçüköğlü, H. Akdeniz, S. Avcu, O. Kemik, Case report: Budd–Chiari syndrome and esophageal variceal bleeding due to alveolar echinococcosis, *Türk. Parazitol. Derg.* 34 (3) (2010) 187–190.
- [10] O. Miman, S. Yazar, Alveolar echinococcosis in Turkey: in the light of the literature, *Türk. Parazitol. Derg.* 36 (2) (2012) 116–120.
- [11] S. Hatipoglu, B. Bulbuloglu, T. Piskin, C. Kayaalp, S. Yilmaz, Living donor liver transplantation for alveolar echinococcus is a difficult procedure, *Transplant. Proc.* 45 (3) (2013) 1028–1030.

Open Access

This article is published Open Access at sciedirect.com. It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.