Portal Hypertension Due to Alveolar Echinococcosis and Recanalized Paraumbilical Vein

Ömer Yılmaz[1], Mustafa Koplay[2], Ömer Onbaş[3], Bulent Aydınlı[4]

[1,4]Atatürk University, Medical Faculty, Departments of Gastroenterology[1] and General Surgery[4] Erzurum, Turkey

[2]Selcuk University, Selcuklu Medical Faculty, Department of Radiology, Konya, Turkey

[3]Duzce University, Medical Faculty, Department of Radiology, Duzce, Turkey

Eur J Gen Med 2012;9 (Suppl 1):39-42

Received: 20.04.2010

Accepted: 04.09.2010

***Correspondence**: Mustafa Koplay M.D. Department of Radiology, Selcuk University, Selcuklu Medical Faculty The Central Campus, Konya, Turkey*
E-mail: koplaymustafa@hotmail.com

Abstract

Hepatic alveolar echinococcosis is an uncommon parasitic disease caused by Echinococcus multilocularis. It carries high fatality rates and poor prognosis if not diagnosed early and managed correctly. We report a 29-year-old female patient who admitted to our clinic with fatigue, weight loss and abdominal bloating in her right upper quadrant. Physical examination revealed the presence of a large palpable mass in her right upper quadrant, ascites and bilateral pedal edema. Computed tomography revealed a large non-homogeneous mass located in the right lobe of the liver and a dilated paraumbilical vein arising from the left branch of the portal vein. Echinococcus serology assessed by enzyme-linked immunosorbent assay was noted to be positive. Histopathologic examination of the hepatic mass was consistent with alveolar echinococcosis. The patient's complaints were determined as being related to cirrhosis due to alveolar echinococcosis. Alveolar echinococcosis must be considered in patients with portal hypertension of uncertain etiology, especially in endemic regions.

***Key Words**: Alveolar echinococcosis, portal hypertension, paraumbilical vein, imaging, CT*

Alveolar Ekinokok Nedeniyle Oluşan Portal Hipertansiyon Ve Rekanalize Paraumblikal Ven

Özet

Hepatik alveolar ekinokok, ekinokokkus multilocularis tarafından sebep olunan yaygın olmayan parazitik bir hastalıktır. Erken tanı ve doğru tedavi yapılmazsa prognozu kötüdür ve fatal seyreder. Biz, sağ üst kadranda şişkinlik, kilo kaybı ve yorgunluk şikayetleriyle gelen 29 yaşındaki kadın hastayı rapor ettik. Fizik muayene sağ üst kadranda büyük bir kitleyi, asiti ve bilateral pedal ödemi ortaya çıkardı. Bilgisayarlı tomografi'de karaciğer sağ lobta non-homojen büyük bir kitle ve portal venin sol dalından köken alan dilate paraumblikal ven tespit edildi. Ekinokokkus serolojisi pozitif bulundu. Hepatik kitlenin histopatolojik muayenesi alveolar ekinokokla uyumluydu. Hastanın şikayetlerinin alveolar ekinokok nedeniyle oluşan sirozla ilişkili olduğu tespit edildi. Alveolar ekinokok, özellikle endemik bölgelerde etiyolojisi belli olmayan portal hipertansiyonlu hastalarda düşünülmelidir.

***Anahtar kelimeler**: Alveolar ekinokok, portal hipertansiyon, paraumblikal ven, görüntüleme, BT*

INTRODUCTION

Alveolar echinococcosis (AE) is a highly pathogenic and potentially fatal chronic infestation that is characterized by a tumor-like infiltrating constitution (1). The liver is the most frequent site (95% of cases) (2). Invasion of biliary and vascular walls is characteristic of this severe disease (3). Invasion of the bile ducts and vessels can lead to very severe complications, such as cholangitis, portal hypertension and biliary cirrhosis (4). The development of portosystemic collaterals occurs often in portal hypertension (5), and a patent umbilical vein is one of its symptoms (6). The specificity of the sign is not well known (7). In the present case, we report a female patient with portal hypertension and AE, which manifested with a calcified mass occupying the right hepatic lobe and a dilated paraumbilical vein (PUV) arising from the left branch of the portal vein.

CASE

A 29-year-old female patient was admitted to the hospital with fatigue, weight loss and abdominal bloating and pain in her right upper quadrant. Physical examination revealed the presence of a large palpable mass in her right upper quadrant, ascites, cachexia and bilateral pedal edema. The patient had huge venous collaterals on the abdominal wall, around the umbilicus. There was no evidence of cardiovascular or pulmonary disease. The liver profile disclosed normal values for aspartate aminotransferase and alanine aminotransferase but bilirubin, alkaline phosphatase and gamma-glutamyl transpeptidase were increased. Serum albumin was decreased but the g-globulin was elevated. All additional routine laboratory tests were normal. Contrast enhanced computed tomography (CT) revealed a large non-homogeneous mass located in the right lobe of the liver in close vicinity to the liver hilum. Postcontrast CT scans did not reveal any contrast enhancement of the involved area. The left lobe of the liver appeared heterogeneous, with compensatory hypertrophy (Figure 1A). The inferior vena cava and right portal vein were invaded. The umbilical vein measured 15 mm and had a patent lumen (Figure 1B). Tests for hepatitis markers and for tumor markers were negative. In upper gastrointestinal endoscopy, grade 3 esophageal varices were observed. Serum sample was investigated for E. multilocularis antibodies using Em2plus enzyme-linked immunosorbent assay (ELISA), and serology was noted to be positive. Liver biopsy was performed. Histopathologic examination of the hepatic mass was consistent with AE (Figure 2). Our patient had a disease stage 3b (P4N0M0) based on PNM staging (P, hepatic localization of parasite; N, extrahepatic involvement of neighboring organs; and M, absence or presence of distant metastases) (8). Liver transplantation was considered but the patient did not accept surgical treatment. Propranolol (to decrease portal pressure) and mebendazole (200 mg four times daily) were prescribed, and the patient was discharged.

DISCUSSION

AE is an uncommon parasitic disease caused by growth of the taenia Echinococcus multilocularis larvae (9). Hepatic alveolar type appears as a gradually developing tumor-like mass (2). It can differ from small foci a few millimeters in size to large (15-20 cm in diameter) areas of infiltration (10). The portal veins are totally involved in the parasitic tissue and the portal hilum is encircled by parasitic disease. After long terms of latent and asymptomatic stages, this vascular involvement leads to portal hypertension or progresses to cirrhotic stage, as in our case (4, 11). Early diagnosis can improve the prognosis and treatment in affected patients. Early clinical symptoms are lacking and the majority of cases are diagnosed at advanced stages, as with the present case (1, 12). In our case, the increase in bilirubin and cholestasis enzymes was probably related to involvement of the main hepatic ducts in the hilar region in association with secondary biliary cirrhosis.

After birth, the umbilical vein atrophies and is called the ligamentum teres. A few small collapsed PUVs form the falciform ligament. In contrast to popular belief, the atrophied umbilical vein itself does not become enlarged but rather the recanalized PUV, which consists of a portosystemic collateral circulation. The PUV is a frequent portosystemic collateral, occurring in up to 43% of patients with portal hypertension (6). Saddekni et al. (7) recognized a cutoff diameter of -3 mm to differentiate the PUV of patients with portal hypertension from that of normal individuals. This cut-off diameter is consistent with the finding of Obel (13), who established in an autopsy series that in normal individuals no umbilical vein exceeded 3 mm. In our patient, contrast-enhanced CT demonstrated patent and dilated PUV with a measured diameter of 15 mm. In our case, portal hypertension documented by huge venous col-

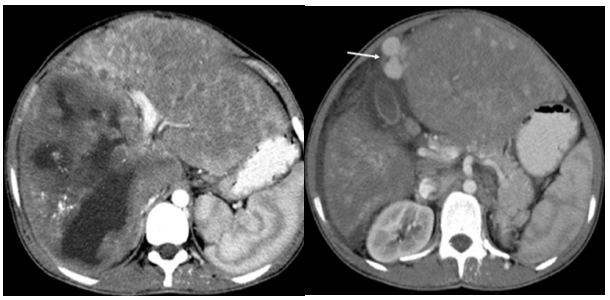

***Figure 1**. A. Postcontrast computed tomography scans did not reveal any contrast enhancement of the involved area. Liver left lobe appears heterogeneous, with compensatory hypertrophy. B. Contrast-enhanced computed tomography demonstrated a dilated paraumbilical vein arising from the left branch of the portal vein. It invaded the inferior vena cava and right portal vein (arrow: paraumbilical vein).*

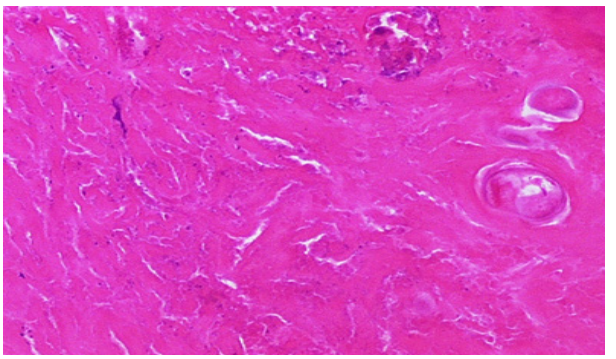

***Figure 2**. Histological examination of the biopsy material at high magnification (hematoxylin eosin stain, 40X) demonstrates calcifications and a few areas of necrosis.*

laterals on the abdominal wall, recanalized PUV and esophageal varices indicated that an obstruction of the portal vein was probably related to involvement of the hilar region. To our knowledge, this is the first case with dilated PUV concomitant with cirrhosis due to AE. The choice of treatment (medical therapy, hepatectomy and transplantation) is still controversial and depends on the extent of infection (14). Radical surgical resection of the parasite in an early phase of infection provides good results for cure, but most cases are diagnosed in a progressed stage, as seen in our case (15). Liver transplantation can only be considered in patients with extreme hilar extension, leading to symptomatic secondary biliary cirrhosis with ascites or variceal hemorrhage due to portal hypertension. We hoped to transfer our patient to another hospital for liver transplantation, but she did not accept surgery. Medical treatment (mebendazole, albendazole) is given in inoperable cases, but these drugs are only parasitostatic, and lifetime therapy is required (4). Some studies seem to strongly show that these compounds are able to delay parasite growth, and reductions in the size of lesions have been reported (9). We also prescribed life-long mebendazole (200 mg four times daily) in this patient.

In conclusion, the finding of a patent or recanalized umbilical vein has frequently been incidental; in endemic regions, AE should not be neglected in the differential diagnosis in patients who have hepatic mass and portal hypertension findings.

REFERENCES

1. *Persat F, Pariset C, Gottstein B, Ninet J, Picot S. Alveolar echinococcosis in a patient without hepatic disturbance and with unusual humoral immune response. Eur J Clin Microbiol Infect Dis 2004;23:859-60.*
2. *Proietti S, Abdelmoumene A, Genevay M, Denys A. Echinococcal cyst. Radiographics 2004;24:861-5.*
3. *Bresson-Hadni S, Delabrousse E, Blagosklonov O, et al. Imaging aspects and non-surgical interventional treatment in human alveolar echinococcosis. Parasitol Int 2006;55:267-72.*
4. *Bresson-Hadni S, Koch S, Miguet JP, et al. Indications and results of liver transplantation for Echinococcus alveolar infection: an overview. Langenbecks Arch Surg 2003;388:231-8.*
5. *Lewis CP, Murthy S, Webber SM, Chokhavatia S. Hemorrhage from recanalized umbilical vein in a patient with cirrhosis. Am J Gastroenterol 1999; 94: 280.*
6. *Horton KM, Fishman EK. Paraumbilical vein in the cirrhotic patient: imaging with 3D CT angiography. Abdom Imaging 1998; 23: 404-8.*
7. *Saddekni S, Hutchinson DE, Cooperberg PL. The sonographically patent umbilical vein in portal hypertension. Radiology 1982;145: 441- 3.*
8. *Aydinli B, Kantarci M, Polat KY, et al. Stereological evaluation of treatment response in patients with non-resectable hepatic alveolar echinococcosis using computed tomography via the Cavalieri method. Liver Int 2006;26: 1234-40.*
9. *Bresson-Hadni S, Koch S, Beurton I, et al. Primary disease recurrence after liver transplantation for alveolar echinococcosis: long-term evaluation in 15 patients. Hepatology 1999;30:857-64.*
10. *McManus DP, Zhang W, Li J, Bartley PB. Echinococcosis.*

Lancet 2003:18;362:1295-304.

11. Sekiya C. Parasitic cirrhosis of the liver. Nippon Rinsho 1994;52:234- 9.
12. Koroglu M, Akhan O, Gelen MT, et al. Complete resolution of an alveolar echinococcosis liver lesion following percutaneous treatment. Cardiovasc Intervent Radiol 2006;29:473-8.
13. Obel W. The umbilical vein in different periods of life in man. Folia Morphol 1976;35:73- 9.
14. Harman M, Arslan H, Kotan C, Etlik O, Kayan M, Deveci A. MRI findings of hepatic alveolar echinococcosis. Clin Imaging 2003;27:411-6.
15. Ammann RW, Fleiner-Hoffmann A, Grimm F, Eckert J. Long-term mebendazole therapy may be parasitocidal in alveolar echinococcosis. J Hepatol 1998;29:994-8.