

Case Report

Multisystem involvement of alveolar echinococcosis in a child

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Abstract

Alveolar echinococcosis (AE) is a chronic progressive infestation inducing a slowly progressing, life-threatening tumor-like growth in the liver. It may spread to other organs by regional extension or hematogenous or lymphatic metastasis. Herein, we report a fifteen-year-old patient diagnosed with AE of the liver and simultaneous lung and brain metastasis with a literature review.

Key words: Alveolar echinococcosis, liver, lung, brain.

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Introduction

Alveolar echinococcosis (AE) is a rare parasitic disease resembling slow-growing malignant tumor on imaging. Initially, it is located in the liver and then may spread to any other organ by regional extension or distant metastases. The most commonly involved organs via metastatic spread are the lungs and the brain [1]. Clinical diagnosis is based on patient history including epidemiological data, clinical findings, morphological assessment of imaging studies, such as ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI), and immunodiagnostic tests, using purified *Echinococcus multilocularis* antigen (Em2-ELISA) as a diagnostic marker [2,3]. Hepatic AE associated with simultaneous lung and brain metastases is a rare clinical entity. We herein report a 15-year-old patient diagnosed with AE of the liver and simultaneous lung and brain metastasis and review current literature.

Methodology

A 15-year-old boy was admitted to the pediatric department of Atatürk University hospital with a history of epilepsy and headaches for last two months. Neurological examination was completely normal. Upon MRI of the brain, the left frontotemporal mass appeared iso-hypointense on T1 weighted images and hypointense on T2 weighted images with lobulated

contours, peripheral signal void areas (calcification), ring-shaped contrast enhancement, and peripheral edema causing the shift of the midline (Figure 1 A-D). Multidetector CT (MDCT) of the chest showed a thick-walled cavitory lesion with a mural hypodense component measuring 5,5×3,5 cm with well-defined borders which was detected in the posterobasal segment of the right lung (Figure 2. A,B). MDCT of the abdomen revealed a peripherally calcified mass lesion measuring 18×11 cm with cystic and necrotic areas in the far left lobe, partially right lobe of liver invading the portal vein and inferior vena cava, causing mild biliary dilatation (Figure 3). Based on the imaging findings, a diagnosis of alveolar echinococcosis was performed. Histopathological examination of ultrasound-guided liver biopsy confirmed the diagnosis of AE (Figure 4).

The hepatic lesion was considered as nonresectable due to invasion of the portal vein and inferior vena cava. Liver transplantation was avoided because of the brain metastasis. Thus, the patient will receive life-long medical treatment, namely albendazole at a dose of 10 mg/kg/day. He is currently under-observation.

Figure 1. T1 weighted non-enhanced (A, M: mass) FLAIR axial image (B, arrows: rim, M: mass), T1 weighted contrast enhanced (C, M: mass) axial image, and sagittal T2 weighted image (D, M: mass).

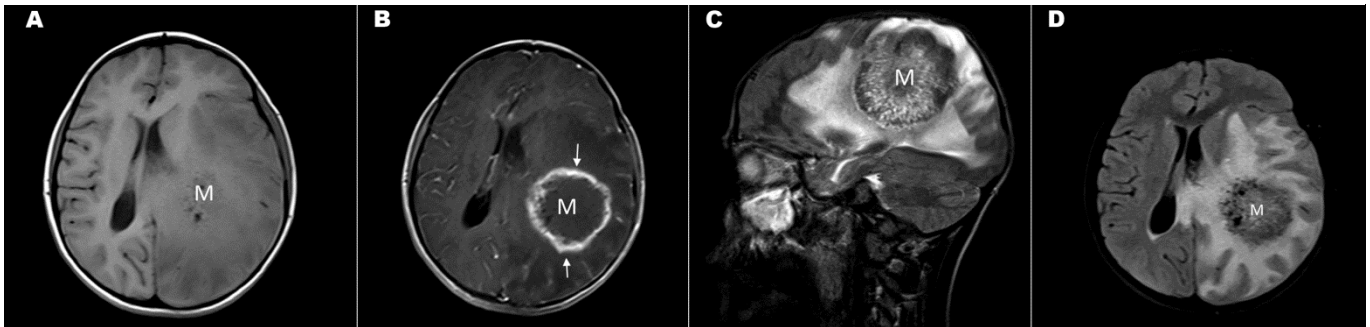


Figure 2. Contrast enhanced thorax CT mediastinal window (A), parenchymal window (B) (arrow: calcification)

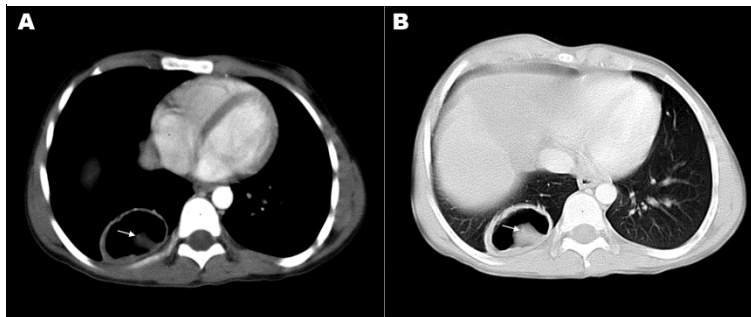


Figure 3. Contrast enhanced abdomen CT (arrows: calcification, M: mass)

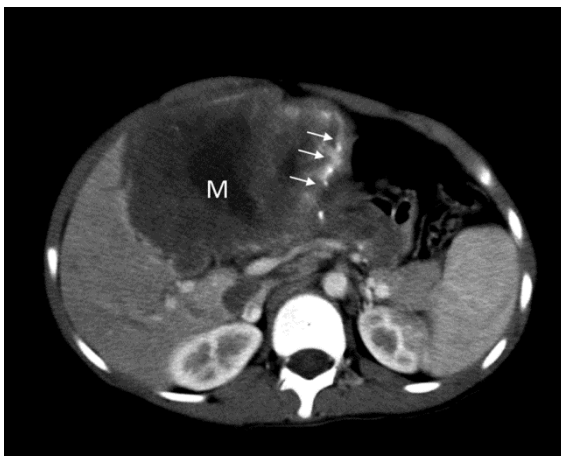
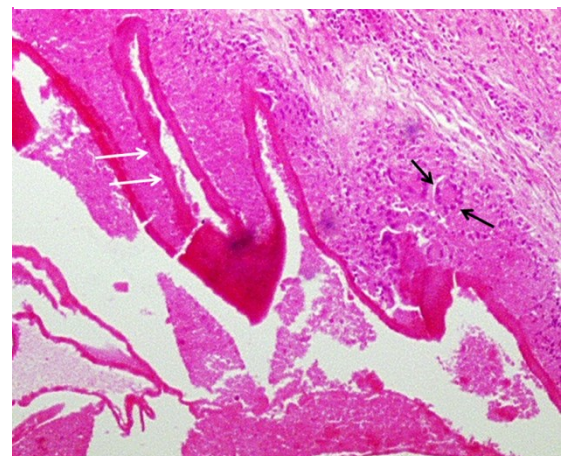


Figure 4. Photomicrograph (original magnification, $\times 100$; hematoxylin-eosin stain) shows multinucleated giant cells (black arrows) metacestodes (white arrows)



Discussion

Alveolar echinococcosis (AE) is caused by infection, with the larval stage of *Echinococcus multilocularis*. AE is found across the globe and is especially prevalent in the northern latitudes of Europe, Asia, and North America. The adult tapeworm is normally found in foxes, coyotes, and dogs. Infection in the larval stages is transmitted to people through ingestion of food or water contaminated with tapeworm eggs. Alveolar echinococcosis is a rare parasitic disease that is caused by the larva of *Echinococcus multilocularis* [3, 4]. The diagnosis of echinococcosis is based on clinical findings, lesion morphology as determined by imaging techniques, immunodiagnostic, and other laboratory tests [5].

Although the majority of AE cases are initially located in the liver, the larvae may spread to other organs by regional extension (e.g. diaphragm, perirenal space, abdominal lymph nodes, peritoneum, pancreas or peripancreatic space, inferior vena cava, lung, gall bladder, retroperitoneal space, abdominal wall, and spleen) [3,5]. The most commonly involved organs via metastatic spread are the lungs and the brain, as in our patient. Liver lesions are seen as a heterogeneous cystic-necrotic mass with calcifications on US, CT and MRI. Mass with irregular margins, scattered calcification, central necrosis, no significant enhancement, but maybe slight long-lasting enhancement of the peripheral fibroinflammatory component, areas of low signal intensity corresponding to fibrotic or collagenous components on T2-weighted sequences and hypointensity on DW images with high b values and high ADC values are suggestive of AE [6].

Treatment choices include benzimidazoles and surgical resection or liver transplantation. The conditions which qualify a patient for liver transplantation are: [1] severe liver insufficiency (secondary biliary cirrhosis or Budd-Chiari syndrome) or recurrent life-threatening cholangitis, [2] inability to perform radical liver resection and absence of extra-hepatic AE locations: cases with residual AE in lung or abdominal cavity should be regarded as exceptional indications, balancing all the pros and cons [3].

Metastasis to the lung occurs relatively frequently and is observed in 7–20% of patients [6-8]. The clinical symptoms of pulmonary AE are hemoptysis, chest pain, cough with expectoration, and exertional dyspnea. There were no pulmonary symptoms in our case. Lung lesions of AE on CT scans appear as low density masses with lobulated or irregular contour. Intra-lesional and wall calcification may be present.

The lesions may be single or multiple, unilateral or bilateral, and of various sizes. In our case the lung lesion was solitary, thick walled and well-defined cavitory lesion with a mural hypodense component [6].

Brain metastasis is considered as a sign of the terminal phase of AE and has been reported in only 1–3% of patients with the disease [7]. Increased intracranial pressure, epilepsy, skull deformity, and cranial nerve palsies have been reported [8]. Our patient presented with epilepsy and headache. On CT or MRI, intracerebral AE lesions exhibit relative characteristic features, showing cystic and often multilobular, grape-like masses with definite margins and surrounding contrast enhancement within the pericystic inflammatory reaction. In our case the lesion was iso-hypointense on T1 weighted- and hypointense on T2 weighted images with lobulated contours, peripheral calcification areas, ring-shaped contrast enhancement, peripheral edema and subfalxian shift. Tuberculosis, bacterial abscesses, fungal infections, invasive brain tumors and metastases should be considered in the differential diagnosis of these infections. Histopathological examination may be necessary for definite diagnosis [8,9].

In conclusion, although AE associated with simultaneous lung and brain metastases is a rare clinical entity; this infection should be kept in mind, especially in patients from endemic areas.

References

1. Craig P (2003) *Echinococcus multilocularis*. *Curr Opin Infect Dis* 16: 437-444.
2. Bresson-Hadni S, Koch S, Beurton I, Vuitton DA, Bartholomot B, Hrusovsky S, Heyd B, Lenys D, Minello A, Becker MC, Vanlemmens C, Mantion GA, Miguet JP (1999) Primary Disease Recurrence After Liver Transplantation for Alveolar Echinococcosis: Long-Term Evaluation in 15 Patients. *Hepatology* 30: 857–864.
3. Polat KY, Balik AA, Celebi F (2002) Hepatic alveolar echinococcosis: clinical report from an endemic region. *Can J Surg* 45: 415–419.
4. Li HT, Tuerganaili, Ayifuhanahan, Shao YM, Zhao JM, Ran B, Wen H (2010) Clinical experiences in the treatment of human multi-organ alveolar echinococcosis by surgery and drugs. *Zhonghua Yi Xue Za Zhi* 90: 2839–2842.
5. Kayacan SM, Vatansever S, Temiz S, Uslu B, Kayacan D, Akkaya V, Erk O, Saka B, Karadag A, Turkmen K, Yakar F, Guler K (2008) Alveolar echinococcosis localized in the liver, lung and brain. *Chin Med J Engl* 121: 90–92.
6. Jiang C (2002) Two cases of liver alveolar echinococcosis associated with simultaneous lung and brain metastases. *Chin Med J* 115: 1898–1901.
7. Kantarci M, Bayraktutan U, Karabulut N, Aydinli B, Ogul H, Yuce I, Calik M, Eren S, Atamanalp SS, Oto A (2012) Alveolar echinococcosis: spectrum of findings at cross-sectional imaging. *Radiographics* 32: 2053-2070.

8. Hattingen J, Uerlings A, Jaussi R (1996) Alveolar echinococcosis of the liver with lymphogenous, pulmonary and cerebral metastasis. *Rofo* 165: 91–93.
9. Kantarci M, Ogul H, Bayraktutan U (2012) Intracerebral alveolar echinococcosis. *Headache* 52: 1296-1297.

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