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Alveolar Echinococcosis of Liver Presenting with Neurological Symptoms due to Brain Metastases with Simultaneous Lung Metastasis: A Case Report

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SUMMARY: Alveolar echinococcosis (AE) is a chronic and serious, even lethal, parasitic infection caused by the helminth *Echinococcus multilocularis* (EM). AE is an endemic disease in Turkey and it is particularly common in people living in the eastern Anatolia Region. In addition to various clinical presentations, symptoms which lead to diagnosis, however, are usually associated with the metastatic lesions. We herein reported a 62-year-old man who had liver alveolar hydatid disease with simultaneous lung and brain metastasis. We think there was only one therapeutic option, namely medical treatment with albendazol, which is the usual treatment for patients living in eastern Anatolia and who are admitted late resulting in a subsequent inoperable situation. Thus, radiological screening studies for the public in this region may increase the possibility of surgical treatment for alveolar hydatid disease.

Key Words: Alveolar echinococcosis of the liver, brain metastasis

Beyin Metastazına Sekonder Nörolojik Semptomlarla Başvuran Eş Zamanlı Akciğer Tutulumlu Hepatik Alveolar Ekinokokkozis: Olgu Sunumu

ÖZET: Alveolar Echinococcosis (AE), bir helmint olan *Echinococcus multilocularis*'in (EM) sebep olduğu ciddi ve kronik bir paraziter enfeksiyondur. AE Türkiye'de endemik olmakla birlikte özellikle Doğu Anadolu'da yaygın olarak saptanmaktadır. Klinik tablo çok çeşitli olmakla beraber teşhisin konulmasında yardımcı olan semptomlar genellikle metastatik lezyonlarla ilişkilidir. Biz bu olgu sunumunda 62 yaşında eş zamanlı akciğer ve beyin tutulumu da saptanan hepatik AE teşhisli bir hastayı değerlendirdik. Bu olguda tek tedavi seçeneği, sıklıkla Doğu Anadolu'da yaşayan hastalarda geç başvuru ve sonrasında alınan inoperabl kararı nedeniyle olduğu gibi, albendazol ile medikal tedavi oldu. Bu nedenle endemik olan bu tür bölgelerde radyolojik görüntüleme yöntemleri ile yapılan taramalar cerrahi rezeksiyon imkanı artırılabilir.

Anahtar Sözcükler: Hepatik alveolar ekinokokkozis, beyin metastazı

INTRODUCTION

Human alveolar echinococcosis (AE) is a potentially fatal, chronically progressive parasitic infection characterized by a long asymptomatic period and development of an invasive tumor-like lesion throughout this period (7, 14). Early diagnosis of AE is very difficult because of long latent or asymptomatic period which may be as long as 20 years (7, 12). The clinical diagnosis is based on patient history including epidemiological data, clinical findings, morphological lesions detected by imaging studies such as ultrasonography (US), computerized tomography (CT), magnetic resonance imaging (MRI), and immunodiagnostic tests such as the enzyme-linked immunosorbent assay, using purified *Echinococcus multilocularis* antigen (Em2-ELISA) as a serodiagnostic marker (7, 12).

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Metastatic spread to the liver occurs usually through the bloodstream. Hepatic AE associated with simultaneous lung and brain metastases is a rare clinical entity and simultaneous lung and brain metastasis was only 1.1% (10). We herein report a 62-year-old patient diagnosed with AE of the liver and simultaneous lung and brain metastasis.

CASE PRESENTATION

A 62-year-old man was admitted with a history of aphasia and ataxia for one month. After neurological examination was performed, on post contrast sagittal T1 weighted MRI imaging, a ring contrast enhancement hypointense pontine lesion of 3 cm diameter with ring-shaped contrast enhancement which

was considered as a metastatic tumor, was determined (Figure 1a). To investigate the primer site of the mass, multidetector row CT (MDCT) images of abdomen and thorax were obtained. MDCT images revealed a calcified mass lesion filling the right lobe of liver and invading vena cava, right portal vein and right renal capsule (Figure 1b) and furthermore, multiple nodules in the posterobasal segments of both lungs (Figure 1c). Histopathological examination of ultrasound-guided liver biopsy confirmed the diagnosis of *Echinococcus alveolaris* (Figure 1d).

Due to presence of the findings associated with locally advanced tumors, invasion of the portal vein and inferior vena cava, the lesion in the liver was accepted as nonresectable. On the other hand, liver transplantation was not also considered because of the brain metastasis. Thus, the patient was received life-long medical treatment, namely albendazole at a dose of 10 mg/kg/day. His follow-up period has been uneventful to date except neurological problems related to brain metastases and he is currently under-observation.

DISCUSSION

Echinococcus multilocularis (*E. multilocularis*) is considered to be the most potentially lethal parasitic zoonosis in the nontropical areas in the Northern Hemisphere (6, 15). Therefore, it is not too hard to estimate that the rate of AE in the east of the Turkey is relatively high compared to the west regions. It rarely manifests symptoms. Nevertheless, diagnosis secondary to the distant metastasis, as occurred in the present case, is rather frequent. Symptoms of AE are primarily cholestatic jaundice and epigastric pain (12). Alveolar echinococcosis is detected as an incidental finding in over one-third of patients (11, 14). Zoonosis prevention and control remain important challenges in the developing world, and new tools and strategies are required. Recent improvements in diagnostic tests allow a comprehensive and integrated approach to control. The diagnosis of AE is based on clinical findings, lesion morphology as determined by imaging techniques, immunodiagnostic, and other laboratory tests (13). Lesions in the liver secondary to AE can vary from small foci of a few millimeters in size to 15-20 cm in diameter large areas (12).

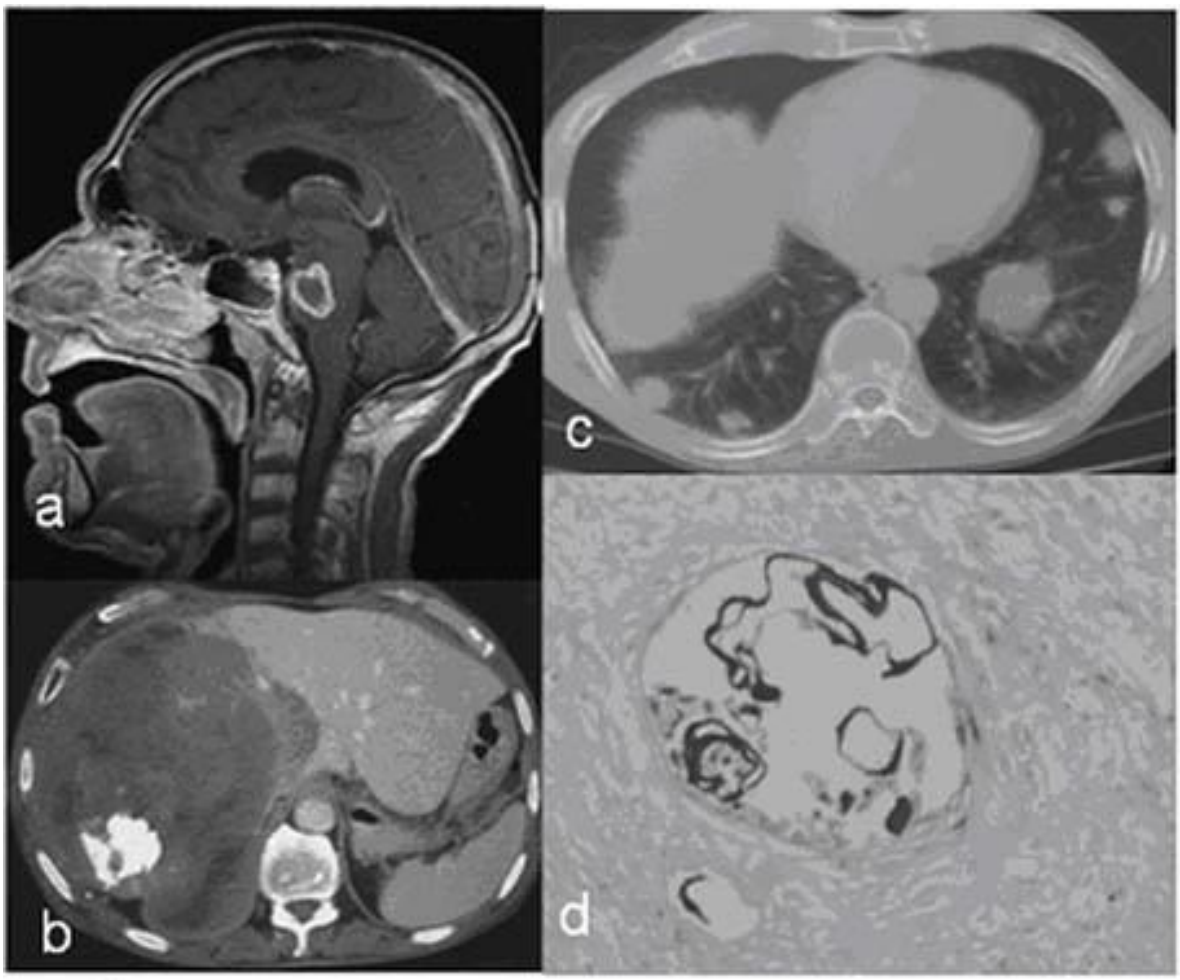

Figure 1a: T1 weighted MRI imaging, a ring contrast enhancement hypointense pontine lesion of 3 cm diameter. **1b:** Calcified and necrotic mass lesion, which is filling the right lobe of liver and invading vena cava, right portal vein and right renal capsule. **1c:** Multiple metastatic nodules in the posterobasal segments of both lungs. **1d:** Pathological examination of liver biopsy revealed *E. alveolaris*.

Treatment of AE involves a variety of options, including surgery and chemotherapy with benzimidazole derivatives (albendazole, mebendazole), and requires a specific clinical experience (8, 12, 13, 16). Modern treatment can significantly prolong the patients' survival time, but cure is only achieved if the metacestode is completely eliminated by radical surgery and complementary chemotherapy (16). In resectable cases, liver hanging maneuver is useful and safe procedure with a lower rate of recurrence (17). In non-resectable cases, chemotheraphy should be used for a long time (about ten years), because no other therapeutic option is indicated as was in our case. A fatal outcome may occur in >95% of untreated patients within a 10-year period following diagnosis (7).

The metacestode tends to spread from the liver to the other organs by infiltration and metastases. The metastatic route of liver AE is mainly via blood circulation. Bresson-Hadni et al. reported a series of 117 patients with AE (5). Pulmonary metastases occurred in 20% of the patients, while cerebral metastases were reported in only 1% and the simultaneous lung and brain metastases occurred in only 1.1% of the patients (10).

Clinical features of patients with intracerebral AE were not specific. Increased intracranial pressure, epilepsy, neurological disturbances such as dysarthria and hemiparesis, skull deformity and cranial nerve palsies have been reported (1). Our patient presented with aphasia and ataxia because of pontine localization of AE. Features of intracerebral *E.multilocularis* on CT or MRI are relatively characteristic showing a grape-like, multilocular cystic mass with definite margins. Single or multiple lesions are observed. Calcifications and surrounding edema are common. There is a contrast enhancement within the inflammatory reaction around the cysts. Differential diagnosis includes tumors and infectious lesions such as tuberculosis and bacterial abscess (4). The spectrum of disseminated AE lesions is widespread and includes metastases. CT-scan and magnetic resonance (MR) imaging are complementary imaging techniques which can be helpful when US images are not typical and/or to perform pre-therapeutic evaluation and disclose distant metastases. On post contrast sagittal T1 weighted MRI imaging, a hypoin-tense pontine lesion of 3 cm diameter with ring-shaped contrast enhancement, which was considered metastatic lesion, was determined in our cases. Therefore, geographical prevalence, clinical history of hepatic involvement and serological tests are required for diagnosis (1). In addition to Em-2, Em-18, in particular, is used for differentiation of active and inactive cases of alveolar hydatid disease (9).

Pulmonary AE is mainly caused by hematogenous dissemination from hepatic AE lesions. Physical sings and symptoms in pulmonary AE are hemoptysis, chest pain cough with expectoration and exertional dyspnea. However, the pulmonary AE caused by hematogenous spread and intrapulmonary enlargement of daughter cyst is usually asymptomatic for about 10 years. For diagnosis of pulmonary AE, circumstantial evidence, like primary lesion in the liver, an appropriate clinical history, a high prevalence of infection in the host's geographic location, and laboratory findings, is employed (3). There were no pulmonary symptoms in our case. They were found incidentally on MDCT images.

The Eastern Anatolia Region is the highest and largest geographical region in Turkey. Eighty-six percent of the patients with AE originate from eastern and central Anatolia, especially Erzurum. The patients diagnosed in the other regions of Turkey are frequently immigrants from Eastern Anatolia (17). There remains mostly single therapeutic option, namely medical treatment, in the cases who live in Eastern Anatolia due to their late admissions. Thus, they usually miss the chance of curative treatment and faces with the complications of the disease as our case. So, in this region, mass screening programs for humans may increase the rate of curative surgical resection.

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