Disseminated Alveolar Echinococcosis Mimicking a Metastatic Malignancy

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Abstract

Alveolar echinococcosis, which is caused by *Echinococcus multilocularis*, is a very aggressive and potentially fatal infestation which always affects the liver primarily and metastasizes to any part of the body. Imaging studies are usually highly suspicious of carcinoma or sarcoma, and biopsy may provide the first indication of infection. We report a case of disseminated alveolar echinococcosis with liver, lung, and bone involvement mimicking a metastatic malignancy.

Key words: *Echinococcus multilocularis*, lung, liver, bone, echinococcosis

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Introduction

Alveolar echinococcosis (AE) is a rare but highly malignant form of echinococcosis and it is caused by *Echinococcus multilocularis*. Wild canine appear to be the definitive hosts, and rodents, deer, moose, reindeer, and bison as the intermediate hosts. Domestic dogs and cats may also cause the disease. Although *E. granulosus* is widely distributed throughout the world, *E. multilocularis* is restricted to the northern hemisphere (1). The liver is the primary site of cyst development in almost all patients, and lung involvement is rare. Herein, we report a case with disseminated AE with liver, lung and bone involvement.

Case Report

A 22-year-old housewife was referred to our clinic with pathologic chest X-ray showing bilateral multiple nodular lesions (Fig. 1). One year ago, she was admitted to a hospital because of night sweats, weight loss, fever and a painful palpable mass in her gluteal region. At that time, lower abdominal CT revealed a cystic lesion in the right iliac bone, extending posteriorly to the soft tissue (Fig. 2a, b). The cystic lesion was drained under ultrasonography and gram staining of the fluid showed numerous polymorphonuclear cells. The aerobic and anaerobic cultures were sterile and acid-fast bacilli was negative. Biopsy of the lesion revealed chronic inflammation with necrotic regions.

On admission to our clinic, nearly one year after the onset of symptoms, she was in good condition without any respiratory complaints. Physical examination was completely normal except for the palpable lesion in the right pelvic area. In room air, oxygen saturation was 98%. Thorax CT revealed bilateral innumerable nodular lesions with internal calcifications and abdominal CT showed a large mass with internal calcifications in the liver (Fig. 3a, b). Pathologic examination of the biopsy from this mass showed areas of necrotizing granulomatous inflammation containing germinative membranous structures of *E. multilocularis* (Fig. 4).

To evaluate the possible infection routes, we detailed the past history. The patient has been living in a city located in Eastern Anatolia and had no history of living in other parts of Turkey or other countries. She had contact with dogs only during childhood. No other contact with the animals could be elucidated.

Echinococcal IgG by an enzyme-linked immunosorbent assay (ELISA) was weakly positive (0.433 with a cut-off...
Peripheral eosinophilia was not present, erythrocyte sedimentation rate was 62 mm/h (0-20) and high sensitive C-reactive protein was 101.1 mg/L (0-5). Clinically and radiologically, there was no sign of liver dysfunction, portal hypertension or cholestasis. Some important biochemical markers were as follows; aspartate aminotransferase (AST), 17 U/L (5-42); alanine aminotransferase (ALT), 12 U/L (5-45); alkaline phosphatase, 117 U/L (90-260); gamma glutamyl transferase, 34 U/L (5-85); lactate dehydrogenase, 364 U/L (240-480); prothrombin time, 13.2 sec (10-15). Pulmonary and pelvic lesions were considered to be the sites of involvement of *E. multilocularis* based on clinical and radiological findings. Cranial CT was normal. There was no sign of other organ involvement such as heart or eye.

Albendazole was started at a dose of 15 mg/kg/day and planned to be used lifelong. The patient was referred for the surgical resection of the lesions in the liver and bone. In the follow-up, the patient did not agree to the surgery. Currently, the patient is on the 6th month of the albendazole therapy without any complaint. She has no limitation in her daily activities and regained her normal weight.

**Discussion**

Echinococcosis is classified under the category of invasive cestode (tapeworm) infections. Mainly two forms of echinococcosis are defined: 1) cystic echinococcosis, which is...
caused by *E. granulosus* and has a worldwide distribution with a predominance in sheep-raising areas; 2) alveolar echinococcosis which is rare and potentially fatal. *E. multilocularis* is restricted to the northern hemisphere (1). Both CE and AE are known to be endemic in Turkey but little relevant data has been collected. CE has been encountered throughout Turkey but AE has only been observed in the cold, mountainous areas of Anatolia, in the eastern part of the country. For the period 1980-1999, a total of 201 cases of AE were reported from Turkey (2). The present patient has been living in a city located in the eastern part of Turkey and had no history of travelling to other parts of Turkey or other countries.

Patients with cystic echinococcosis mostly have single organ involvement (85-90%), and the liver is the most common site of involvement, followed by the lung. On the other hand, the liver is consistently involved in patients with alveolar echinococcosis (3).

The cysts of *E. multilocularis* grow slowly with an incubation period of 5-15 years. Exogenous budding and proliferation of the *E. multilocularis* cyst causes infiltration into adjacent tissues and results in pressure necrosis of the surrounding tissue (1). It extends beyond organ borders and metastasizes to distant sites, such as the lungs, brain, bone, and others.

Lung involvement always occurs after hepatic involvement. Transdiaphragmatic contiguous migration of hepatic lesions is common and intrathoracic rupture of hepatic cysts into the bronchial tree, pleural cavity or mediastinum may occur (3). Lung lesions of alveolar echinococcosis on CT scan generally have an irregular contour with intra-lesional and wall calcifications. Imaging studies are usually highly suspicious of carcinoma or sarcoma, and biopsy may provide the first indication of infection. The patients’ overall condition is generally better than would be expected with a malignancy (4, 5).

A fatal outcome may occur in 95% of untreated patients within a 10-year period following diagnosis. Radical surgical resection of the localised lesions is typically the only curative measure for alveolar echinococcosis. A minimum of 2 years of chemotherapy is recommended after surgery. If the resection is not possible or not complete; long-term, even life-time chemotherapy, is recommended (5). Albendazole (10-15 mg/kg/day; orally into two divided doses) can significantly extend the length of survival, but this therapy is normally parasitostatic rather than parasitocidal (6).

In conclusion, we have presented a case of disseminated AE of liver, lungs and bone. Since AE is a very aggressive infestation and may involve any part of the body, other organs must be screened to detect lesions in the early resectable stage because resection is probably the only curative treatment.

References