

## **A Case Report of Fatal Echinococcosis**

**Tayfun Hakan MD, \* Fugen V. Aker MD**

Kartal Teaching and Research Hospital Neurosurgery Department and \*Haydarpasa Numune Hospital Pathology Department, Istanbul, Turkey

### **Abstract**

**Introduction:** Alveolar hydatid disease is a lethal parasitic disease of human caused by the larval stage of echinococcosis multilocularis. Human beings are accidental intermediate hosts. Brain involvement is rare. The clinical, radiological and pathological findings in a case of multi-located alveolar hydatid disease are presented and the literature reviewed.

**Case presentation:** The patient presented with advanced disease. There were separate intracranial, hepatic and pulmonary masses. There were two lobular and three multilobular cystic lesions intracranially. Stereotactic brain biopsy provided the diagnosis. Despite diagnosis and medical treatment the patient succumbed.

**Conclusion:** Echinococcosis multilocularis remains a lethal disease even today. It should be borne in mind in the differential diagnosis of multiple intracranial lesions especially if there are other similar lesions in the liver or lungs. Stereotactic biopsy is safe, accurate and effective.

### **Key Words**

• Alveolar hydatid disease • Echinococcosis alveolaris • Echinococcosis multilocularis • hydatid cyst • stereotactic biopsy

### **INTRODUCTION**

Alveolar hydatid disease is an aggressive and a highly lethal parasitic infection caused by the larval stage of Echinococcosis multilocularis ([1-8,11-19](#)). The cestode is largely restricted to the Northern Hemisphere and is seen sporadically in rural areas ([2,4,7,8,11,14,17](#)). Approximately half of

the cases of alveolar hydatid disease reported in Turkey were found in the eastern part of the country including Erzurum, Erzincan and Kars. Foxes and small rodents represent the natural hosts ([4,5,17](#)). After ingestion of contaminated foods and/or water, embryos of Echinococcosis multi-

### **Correspondence:**

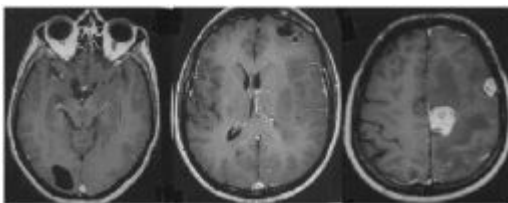
Tayfun Hakan, Acibadem Gomec sokak, Hamdibey Ap. No:45/9, Kadikoy 81020, Istanbul - Turkey ;  
Phone: +90.532 324 32 84; Fax: +90.216 363 93 33; e-mail: [tayfunhakan@yahoo.com](mailto:tayfunhakan@yahoo.com)

ocularis migrate by the portal system to the liver where vesicular masses are formed (4,5,7,8,13). The disease may extend gradually to adjacent organs or spread via the bloodstream to distant areas such as the brain (2,7,13,14).

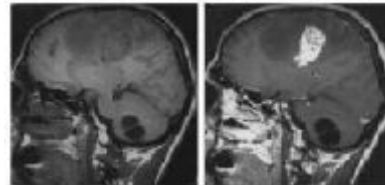
We report a case of disseminated alveolar echinococcosis. We detail the clinical, radiological and pathological features and also review the literature. We hope to alert clinicians in endemic areas about this infestation and also to stress the diagnostic value of stereotactic biopsy.

### CLINICAL PRESENTATION

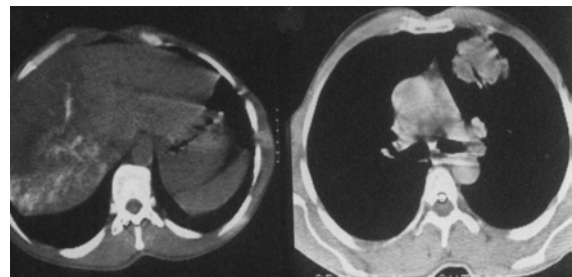
A 53-year-old man presented with headache and right-sided seizures. He was indeed from the eastern part of Turkey (province of Erzincan) but had been living in the western part of Turkey for 10 years. His general condition was very poor. Examination revealed high fever (38.5°C), hepatomegaly and right hemiparesis. Laboratory findings included an elevated erythrocyte sedimentation rate (70 mm/hr) and a white blood cell count of 20,000 mm<sup>3</sup>. Indirect hemagglutination test was positive 1/4096 titration. Multiple lesions were seen on cranial magnetic resonance (MR) imaging (Figure 1,2).



**Fig. 1:** Axial CT-Head with contrast; The multiloculated cystic lesions in right occipital, in left frontal lobe, and solitary lesions, one in left frontal and the other one in left parietal lobe were detected.



**Fig. 2:** Sagittal T1-weighted MR images demonstrating a supra-tentorial, subcortical, solitary lesion with good contrast-enhancement and an infra-tentorial, multilocular, cystic lesion.

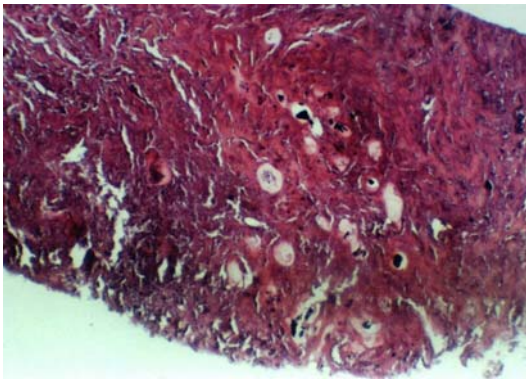


**Fig. 3:** CT (with contrast) images of liver and lung demonstrating: **A.** mass formation having diffuse amorphous calcifications especially in periphery of right lobe of the liver **B.** A lobular contoured mass lesion in 5 cm diameter having soft tissue density in the lingular segment of the left lung; another 1.5 cm diameter mass is also seen in supra-hilar area.

There were 3 multiloculated cystic lesions, in the left cerebellum, in the right occipital and left frontal lobes; and 2 solitary lesions, in the left frontal and left parietal lobes. Computerized Tomography (CT) showed multiple masses in the liver and lung (Figure 3).

A percutaneous CT-guided lung biopsy was non-diagnostic. A liver biopsy was

speculated to be a case of Echinococcosis. Results of the Serological test were not immediately available and in view of the serious neurological condition of the patient, stereotactic biopsy was carried out. We undertook a stereotactic biopsy of the subcortically located solitary lesion in the left parietal lobe under local anesthesia. Echinococcosis multilocularis was found (Figure 4).



**Fig. 4:** Photography of a stereotactically obtained biopsy specimen showing multiple calcifications and microcystic structures, having eosinophilic lamellas in their lumen, in necrotic/necrobiotic fibrous stroma. No scoleces are seen (HE x 20).

Albendazole (2x400mg/day) was administered. Assuming an opportunistic infection, cephotaxim (2x2gr/day) and ornidazole (4x500mg/day) were also added. Unfortunately, the patient died due septicemia and hepatic failure.

## DISCUSSION

Virchow described the infection of echinococcosis multilocularis for the first time in 1855. In our country,

Turkey, Kiatibian first described the infection in 1872 (4). It has been reported that Echinococcosis alveolaris and Echinococcosis granulosus are not seen together in the same areas. Both are endemic in Turkey (1,2,4,11,13,16,17).

While the infection of *E. multilocularis* is endemic in high altitude zones and areas of cold climate, *E. granulosus* infection is mostly seen in areas of moderate climate (17). Both may however be seen outside endemic areas due to migration from endemic areas or due to tourism.

Echinococcosis is caused by the larval (metacestode) stage of the cestode species *Echinococcus granulosus* or *Echinococcus multilocularis*. The most prevalent species is the *Echinococcus granulosus* causing unilocular hydatid disease. *Echinococcus multilocularis* causes multilocular or alveolar hydatid disease. Hydatid cysts are commonly round or oval, but they may assume the shape of any organ or space in which they grow. Several membranes, germinal membrane, laminated membrane and fibrous capsule form the cyst. In *Echinococcus multilocularis* the germinal layer allows asexual budding to form "daughter cysts" within the primary cyst (4,7,8).

Dogs and foxes are the definitive hosts and sheep, cattle, goats, horses, camels or swine are the intermediate hosts for the larval stage of *E. granulosus* (7,8). The definitive hosts of *E. multilocularis* are foxes and wolves while domestic

dogs and cats may also be. Small rodents such as rats, squirrels, voles and field mice are the intermediate hosts in Turkey (17).

Humans serve as a accidental intermediate hosts for both species, the disease being acquired by oral ingestion of the eggs of the parasite (5,7,8,14,15).

Multilocular alveolar cyst disease is very aggressive. Lesions develop rapidly in the liver where an invasive pseudomalignant mass may be produced (7,11,15). This may spread locally to adjacent organs such as the lung or metastasize to distant structures such as thyroid, bone, vertebra, scrotum, spleen, ovarium, heart (4), kidney, peritoneum, lymph nodes, rectus muscle, knee, lacrimal sac (17), palpebra (3) and occasionally the nervous system (1,2,13,14) where severe headaches and seizures are generally the presenting features (11). The liver and lung are mainly involved in *E. granulosus* infection (7).

Immuno-fluorescence, passive hemagglutination tests, ELISA and immunoelectrophoresis with antigenic extracts of *E. multilocularis* may provide a diagnosis. Serologic testing can be positive but this does not always help us to make a definite differentiation between the infections of *E. granulosus* or *E. multilocularis*. We were able to do only indirect hemagglutination test among them; and it was found to be positive in 1/4096 titration.

Imaging techniques may be more sensitive than serodiagnosis (7,11). Contrast enhancement in *Echinococcus alveolaris* presents as multiloculated cysts, which are described as grape-like (3) or as more solid heterogenous lesions (13,14). In the present case three of the cerebral lesions were multiloculated and two were more solid. Fungal infections, tuberculoma and metastatic disease form the main differential diagnosis (12,14).

Cysticercosis is also should to be kept in mind. It is caused by infestation of the larval stage of the pork tapeworm *Taenia solium*. It is common in Eastern Europe, Mexico, Central and South America and Africa. *Cysticercus cellulosae* and *Cysticercus racemose* may give rise to brain cysts. In *C. cellulosae* infection cysts are regular, round and thin walled. In *C. racemose* infection cysts are larger and characterized by multiple grape-like small vesicles. While, *C. cellulosae* tend to localize to parenchyma and contains a scolex in it, *C. racemose* tend to be found in basal subarachnoidal spaces and contains no larvae. Eosinophilia in the peripheral blood is typical in cysticercosis (9,10). Echinococcosis is unusual in Turkey as pork is rarely eaten.

In our case previous biopsies of other organs were non-diagnostic. The lung biopsy consisted of necrotic material only. The liver biopsy was suspicious of hydatid cyst but not diagnostic. The lesions were multiple and widespread

in the brain and we performed stereotactic biopsy of the lesion located in the left parietal lobe under local anesthesia. To our knowledge, this report is only the second such biopsy (14).

Surgical excision of solitary lesions of *E. multilocularis* has been described (1,2,3,11,13,14,16). However cyst rupture during removal may lead to dissemination and/or chemical meningitis. Fatal anaphylactic shock has also been described (8).

Pathologists must rely on the characteristic appearances of the membranes. As in our case scoleces are rarely demonstrated (11).

Long-term adjuvant chemotherapy may provide significantly improved outcomes even for severely ill patients (7,18,19). Chemotherapy is advised in disseminated disease, recurrent cases, if the cyst rupture occurs during surgery, in cases having very high serologic levels and in poor grade patients

(3,6,11,18,19). Mebendazole, albendazole or flumendazol are used (3,7,11).

Hepatic failure, local extension or metastases to the vital organs lead to mortality. In untreated patients the mortality may reach 90% (15,18,19). In the present case there were multifocal brain lesions, hepatic failure and superimposed secondary infection.

## CONCLUSION

In patients with multiple cerebral lesions who live in or have visited an endemic area and especially if there has been contact with foxes, dogs or rodents; Echinococcosis multilocularis should be remembered in the differential diagnosis. Echinococcosis multilocularis remains today a very lethal disease. The cerebral lesions may be multiloculated, solid, solitary or multiple and may involve the infra- or supra- tentorial areas. Stereotactic biopsy of a cerebral lesion is a safe, accurate and effective diagnostic procedure where the need arises.

## REFERENCES

1. Altinors N, Kars Z, Cepoglu C, Gurses L, Sagbil S, Ariyurek M: CT findings and surgical treatment of double intracranial Echinococcal Cysts. [Infection19:110-114, 1991.](#)
2. Aydin Y, Barlas O, Yolas C, Aydin IH, Ceviz A, Aladag A, Oren D, Akdemir D: Alveolar hydatid disease of the brain. Report of four cases. [J Neurosurg 65:115-119, 1986.](#)
3. Bensaïd AH, Dietemann JL, Flippi De La Palavesa MM, Klinkert A, Gangi A, Jacquet G, Cattin F: Intracranial alveolar echinococcosis CT and MRI. [Neuroradiology 36:289-291, 1994.](#)
4. Canda MS, Canda T: The presentation of 47 cases and the echinococcus problem of Turkey. (in Turkish) *T Parazitol Derg (Acta Parasitologica Turcica)* 19:64-82, 1995.
5. Ciftcioglu MA, Yildirgan MI, Akcay MN, Reis A, Safali M, Aktas E: Fine needle aspiration biopsy in hepatic echinococcus multilocularis. [Acta Cytologica 41:649-652, 1997.](#)

6. Goldman IS, Brandbong LL: Parasitic disease of the liver, in Zakim D, Boyer TD (eds): Hepatology, A Text Book of Liver Disease. Ed 2, vol 2, Philadelphia, W.B Saunders, 1994, pp 1074-1076.
7. King CH: Cestodes (tapeworms), in Mandell GL, Bennett JE, Dolin R (eds): Principles and Practice of Infectious Disease. Ed 4, vol 3, New York, Churchill Livingstone, 1995, pp 2544-2553.
8. Koneman EW, Allen SD, Janda WM, Schreckenberger PC, Winn Jr. WC: Parasitology. Color Atlas and Textbook of Diagnostic Microbiology. Ed 5, vol 2, Lippincott, Philadelphia, 1997, pp 1071-1162.
9. Leblanc R, Knowles KF, Melanson D, MacLean JD, Rouleau G, Farmer JP: Neurocysticercosis: Surgical and medical management with Praziquantel. [Neurosurgery 18:419-427, 1986.](#)
10. Martz RD, Hoff JT: Parasitic and fungal disease of the central nervous system, in Youmans JR (ed): Neurological surgery, Vol.6. Philadelphia, W.B. Saunders Company, 1996, pp3742-3751.
11. Oktar N, Karabiyikoglu M, Demirtas E, Altintas N, Korkmaz M, Ozdamar N: Cerebral alveolar echinococcosis. Review of the literature and report of a case. *Norol Bil D (Turkish Journal of Neurological Sciences)* 16:1, 1999.
12. Osborn AG: Diagnostic Neuroradiology. Mosby, St. Louis, 1994, pp 187-190, 673-712.
13. Pamir MN, Ozer AF, Keles GE, Tozun N, Gurmen N, Kullu S: Cerebral echinococcosis multilocularis. Case report. [J Neurosurg Sci 35:161-164, 1991.](#)
14. Piotin M, Cattin F, Kantelip B, Miralbes S, Godard J, Bonneville JF: Disseminated intracerebral alveolar echinococcosis: CT and MRI. [Neuroradiology 39:431-433, 1997.](#)
15. Taratuto AL, Venturiello SM: Echinococcosis. [Brain Pathol 7:129-131, 1997.](#)
16. Topsakal C, Aydin Y, Aydin F, Sahin Y, Bilge T, Kasaroglu D, Delibalta N: Cerebral alveolar hydatidosis: case report. [Surg Neurol 45: 575-581, 1996.](#)
17. Uysal V, Paksoy N: Echinococcosis multilocularis in Turkey. [J Trop Med Hyg 89:249-255, 1986.](#)
18. WHO informal working group on echinococcosis: Guidelines for treatment of cystic and alveolar echinococcosis in humans. *Bull World Health Organ* 74:231-242, 1996 (abstr).
19. Wilson JF, Rausch RL, Wilson FR: Alveolar hydatid disease. Review of the surgical experience in 42 cases of active disease among Alaskan Eskimos. [Ann Surg 221:315-323, 1995.](#)

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 Professor and Chairman,  
 Department of Neurosurgery  
 LSU Medical Sciences Center  
 Post Office Box 33932  
 Shreveport, LA 71130-3932  
 318-675-6404  
 318-675-4615(fax)