Letter to the Editor

# Rare recurrent brain alveolar echinococcosis complicated by systemic multiorgan infection—a case description

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Submitted Dec 01, 2022. Accepted for publication Mar 28, 2023. Published online Apr 07, 2023.
doi: 10.21037/qims-22-1336
**View this article at:** https://dx.doi.org/10.21037/qims-22-1336

## Introduction

Echinococcosis, formerly known as Hydatid disease (HD), but now "hydatid" refers to a cystic lesion formed by cystic echinococcosis (CE) (1). Echinococcosis is a zoonotic disease that is endemic in many parts of the world and is more prevalent in rural areas, especially in resource-poor countries, but it has recently increasingly moved to urban areas (2). Humans function as accidental intermediate hosts and become infected by ingesting food or water sources contaminated with eggs. The eggs penetrate the intestinal wall and circulate in the blood (portal venous system) to the liver, so the most common site of infection is the liver (3). Echinococcosis in other parts is rare, but the disease can occur in almost any organ of the body (4). Because the lesions grow slowly, they may be asymptomatic for many years. In general, the clinical presentation is nonspecific, with resulting symptoms depending on lesion location, size, host immune response, and complications such as superimposed bacterial infection and cyst rupture. Therefore, diagnosing echinococcosis can be a serious clinical challenge for inexperienced clinicians.

There are three main types of echinococcosis: CE, alveolar echinococcosis (AE) and neotropical echinococcosis (NE) (1). of which echinococcosis caused by AE is very rare. The annual CE incidence ranges from <1 to 200 per 100,000, whereas that of AE ranges from 0.03 to 1.2 per 100,000 (5). The case fatality rate for untreated human alveolar echinococcosis (human AE) at 10–15 years is approximately 90%. There are 18,235 new cases of human AE every year worldwide (6). New human AE cases in China account for 91% of the global human AE burden every year (7).

In this case, we reported a man from a pastoral area of Qinghai, China, who was infected with echinococcosis throughout his body, underwent two brain AE resections and one hepatic AE resection and was hospitalized again for seizures and vision loss.

## Case presentation

All procedures performed in this study were in accordance with the ethical standards of the Ethics Committee of Beijing Friendship Hospital Affiliated to Capital Medical University and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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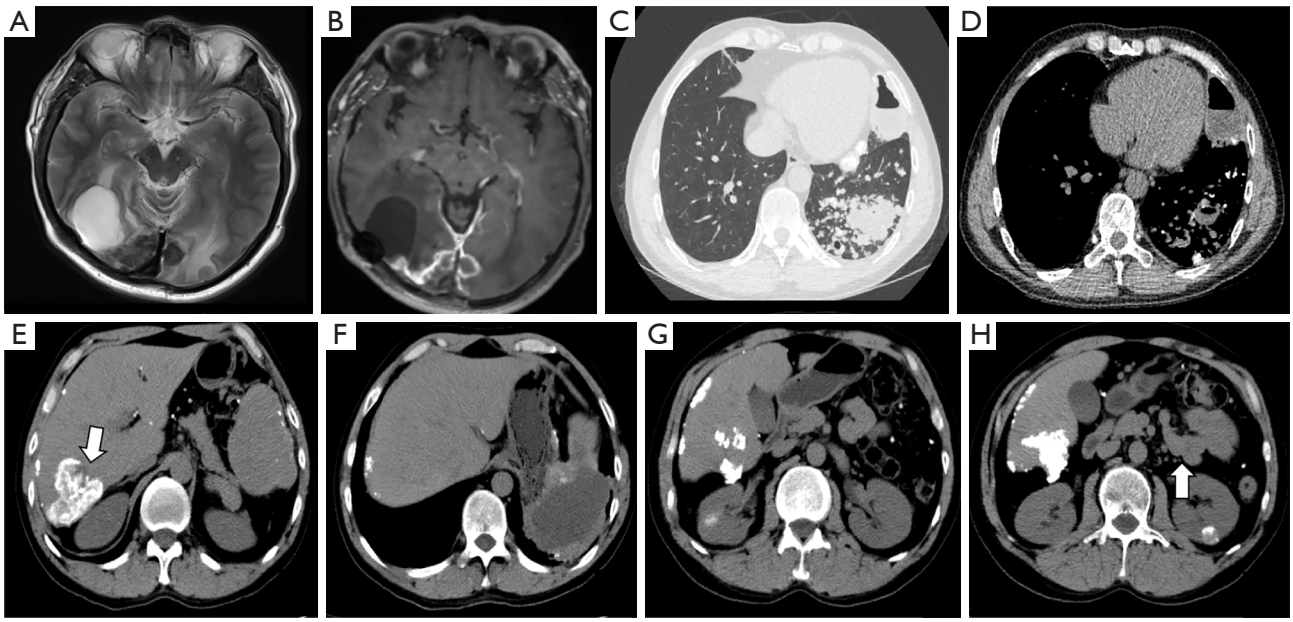


**Figure 1** Image of head, chest and abdomen. (A) Brain MRI T2WI sequence. The right occipital lobe cystic signal shadow; adjacent brain tissue edema; bilateral occipital lobe adjacent to the dura mater thickening showed a slightly low signal shadow, and the low signal in the skull next to the lesion was an artifact after the previous operation. (B) Brain MRI T1WI enhanced, no enhancement of the right occipital cyst, bilateral paraoccipital dura enhancement. (C,D) Axial CT of the chest shows multiple cystic lesions in the lower lobe of the left lung with multiple calcifications, left subpleural cystic lesions with gas accumulation, and multiple calcified nodules on the left side of the pericardium. (E) Axial CT image of abdomen. Multiple calcifications in the right lobe of the liver; patchy low-density lesions are seen next to the calcifications, and uncalcified hydatid lesions are considered (the white arrow). (F) Axial CT image of abdomen. Patchy low-density shadows in the spleen, multiple punctate calcifications on the edge of the lesion, and multiple punctate calcifications near the stomach wall. (G) Axial CT image of abdomen. Right renal hydatid calcified nodule with scattered calcified lymph nodes in the abdominal cavity. (H) Axial CT image of abdomen. Left renal hydatid calcified nodule, left adrenal hydatid invasion (white arrow). MRI, magnetic resonance imaging; T2WI, T2-weighted image; T1WI, T1-weighted image; CT, computed tomography.

A 46-year-old male patient from a pastoral area had a history of hepatic AE excision in 2014. In 2016, the patient had no obvious cause for vision loss, and he went to a local hospital and was diagnosed with brain echinococcosis metastasis. A craniotomy was performed at a local hospital, and the postoperative recovery was good. In December 2021, the patient had sudden syncope without any incentive, accompanied by epileptic seizures, was diagnosed with recurrence of intracranial echinococcosis in the local hospital, and underwent right frontoparietal echinococcosis excision again. In June 2022, the patient had persistent headache with no incentive and blurred vision. For further treatment, he was admitted to the Neurosurgery Department of Beijing Friendship Hospital Affiliated to Capital Medical University. Ask the patient's family to learn that the patient did not insist on taking albendazole regularly.

The patient was admitted to the hospital for relevant imaging studies (*Figure 1*). Magnetic resonance imaging (MRI) of the brain showed a cystic lesion in the right occipital lobe with edema adjacent to the brain parenchyma, no enhancement in the lesion on enhanced scan, and linear and circular enhancement in the meninges around the lesion. It was postoperatively confirmed to be calcified hyperplasia tissue, which was considered to be meningeal irritant hyperplasia caused by repeated craniocerebral surgery. Chest computed tomography (CT) showed multiple small nodules and masses in the lower lobe of the left lung, with calcification at the edges of some lesions. The largest lesion was approximately 4 cm in diameter, and an air-fluid level was seen inside. There were also multiple cystic low-density lesions in the left subpleural region, and an air-fluid level could be seen in some of the lesions. Considering that the lung tissue was damaged by pulmonary

 

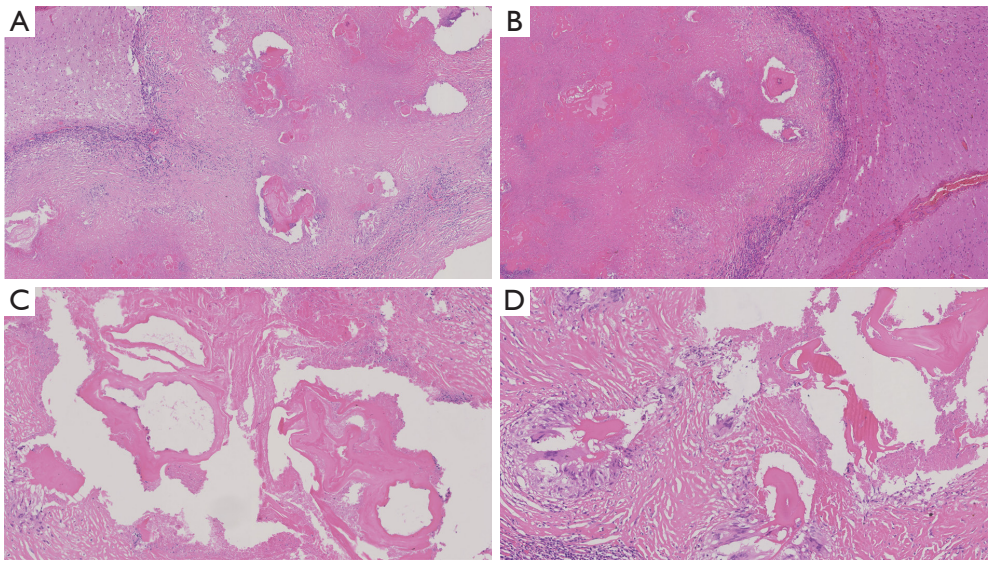


**Figure 2** Pathological picture. Histological features. (A) Several cysts of different sizes were seen in the brain parenchyma (HE staining, ×40). (B) Necrotic tissue was seen in the nodules of the cyst wall (HE staining, ×40). (C) Most of the cyst wall was composed of a keratin membrane, which was a red-stained plate lamellar structure (HE staining, ×100). (D) Granulomatous nodules of histiocytes and multinucleated giant cells were seen around the keratinocytes (HE staining, ×100). HE, hematoxylin-eosin.

hydatid, the lesions were thought to be connected with the bronchus. Abdominal CT examination showed multiple calcified nodules in the right lobe of the liver, and some calcified borders showed patchy, slightly low-density shadows. Uncalcified hydatid lesions were considered, and there was no dilation of intrahepatic bile ducts. The spleen was enlarged with blurred margins, and large lamellae with low density were seen. The lesion invaded the diaphragm and was connected with the lesion in the left lower lobe of the lung. The demarcation between the anterior edge of the spleen and the greater curvature of the stomach was unclear, and multiple calcifications were seen in the adjacent gastric wall, so we considered the invasion of the gastric wall. The left adrenal glands were enlarged and abnormal in shape, and echinococcosis transfer was considered. Multiple nodular calcifications were seen in the parenchyma of both kidneys, and the transfer of echinococcosis in both kidneys was considered. Diffuse calcified lymph nodes in the abdominal cavity were also observed. Serum IgG antibody of echinococcosis was positive by enzyme-linked immunosorbent assay (ELISA).

This patient's presentation was mainly due to craniocerebral symptoms causing severe physical discomfort, and chest and abdominal lesions did not cause obvious clinical symptoms. The patient had a definite history of two cerebral echinococcosis resections, and the diagnosis of recurrence of cerebral echinococcosis was clear. Resection of recurrent echinococcosis cysts was performed in our hospital. The original incision was reopened, and after the craniotomy, the dural tissue was found to be hardened, and the tension was high. After the dura was incised, the calcified scar tissue appeared to be diffusely distributed, and a gray-white cyst, approximately 4 cm × 3 cm in size, with a thin and damaged wall was seen. Afterward, yellow liquid was seen flowing out. It was managed with gauze to avoid contamination of surrounding tissues. The tumor was almost completely removed, and the yellow calcified hyperplasia tissue on the surface of the brain was peeled off at the same time.

The cystic lesion was excised and sent for pathology. Gross features were as follows: a ruptured cyst, 5 cm × 4 cm × 2.5 cm in size, 0.3–0.5 cm thick cyst wall, rough cyst wall, a nodule attached to the inner wall of the cyst, and a diameter of 1.5 cm. Histological features included several cysts of different sizes in the brain parenchyma. The walls of the cysts were keratinocytes. Necrotic tissue was seen in nodules of the cyst wall, and numerous lymphocytes infiltrated around the nodules. Most of the cyst walls were composed of keratin membranes, which were red-stained lamellar plate structures. Granulomatous nodules of histiocytes and multinucleated giant cells were seen around the keratinocytes (*Figure 2*).

 

The final pathological diagnosis was brain echinococcosis disease. Since this section belonged to the third operation and had multiple calcifications, the worm body had not been cut, so the type of echinococcosis could not be distinguished. Real-time fluorescence quantitative polymerase chain reaction (PCR) technique for formalin-fixed paraffin-embedded (FFPE) tissue samples can improve the accuracy of AE diagnosis (8). However, based on the comprehensive consideration of the patient's past history and the pathological results of the first surgery outside the hospital, it was confirmed that it was a brain AE.

## Discussion

Echinococcosis is also known as "neglected tropical diseases" (1). However, the adverse consequences it brings are very serious, the AE type has a very high mortality rate. This is a typical case of a multiorgan AE, the primary site of which is the liver. AE is more prone to systemic metastasis than CE, usually through blood and lymph to distant metastases, brain metastases occur in about 2–3% of cases (9). This case is a typical case of multiple organ echinococcosis. Therefore, AE has a high degree of malignancy, and it is necessary to focus on early diagnosis and active treatment.

The diagnosis of echinococcosis needs to be informed by a variety of examinations, including clinical manifestations, medical history, serology and imaging methods, among which imaging examination is very important, providing a basis for the staging of lesions and surgical methods. Ultrasound is good for abdominal organ observation, but CT and MRI are more valuable for lung, brain, bone and other lesions. CT is particularly valuable for the presence and shape of calcification in cysts and can also show lesion size, number, local complications, and adjacent tissue involvement. MRI multisequence imaging can help in the identification of echinococcosis and tumors (10). Heavy T2-weighted image (T2WI) sequences can better show biliary involvement and evaluate the nerve involvement of intracranial lesions. Positron emission tomography (PET)/CT has a very high value in liver metabolic activity and is an irreplaceable imaging method for evaluating or indirectly assessing parasite activity (11,12). However, a lack of detectable metabolic activity does not imply parasite death but rather indicates suppression of inflammatory activity around the parasite (13). In this case, a variety of examination methods, such as MRI and CT, were used comprehensively to achieve good diagnostic performance in each affected organ from multiple sequences, multiple parameters, and multiple perspectives.

The CT of this patient showed multiple lesions in the liver, most of which were calcified, and patchy low-density shadows were also seen at the edges of some calcified lesions, which were considered to be uncalcified AE lesions, except that the lesions were active. According to the imaging WHO-Informal Working Group on Echinococcosis (WHO-IWGE) PNM (parasite lesion, neighboring organs invasion, metastases) classification (13), the patient was classified as P3N1M1, that is, intrahepatic lesions involving multiple liver lobes, accompanied by adjacent multiorgan invasion and multiorgan metastases. This classification is used to indicate the severity of the lesion and helps clinicians develop treatment plans for patients. The patient had undergone radical resection of liver AE disease, but the patient still developed brain and pulmonary AE disease 2 years later. The patient had undergone two intracranial echinococcosis operations in the past, and now the condition recurred, showing the dilemma of AE treatment. At the same time, it is related to patients' irregular medication. There are literature reports that the core of AE treatment is still continuous albendazole drug therapy, and individualized intervention measures should be taken when necessary. This approach can improve outcomes for most patients with AE (13). Studies have shown that AE is associated with immune deficiency, but the patient had no history of immunodeficiency (14,15). In this patient, multiple abdominal organs were involved, the spleen parenchyma was destroyed, and the adjacent tissues were invaded, indicating that the patient's disease was advanced and the prognosis was poor. Today, with the rapid development of living standards and diagnosis and treatment, advanced AE involving multiple organs is rare.

The lung imaging manifestations of AE in this case were misdiagnosed as pulmonary tuberculosis, lung abscess, liver cancer lung metastasis and other diseases. The imaging manifestations overlap with pulmonary echinococcosis metastasis. Therefore, it takes a more experienced physician who combines history, laboratory and imaging to make the correct diagnosis. Correct diagnosis is the precondition of treatment and determines the therapeutic approach for the disease. Where the cysts are accessible, surgical removal offers the best mode of treatment, but recurrence after surgery is common (16). In this patient, although the primary site of the lesion was resected, multiple metastases still occurred throughout the body, so more treatment methods needed to be explored. Any treatment method is inseparable from imaging technology support, which enables assessment

 

of preoperative metastasis, the positioning of intraoperative lesions, and the detection of postoperative recurrence of lesions. This is also the charm of imaging.

## Conclusions

The liver is usually the site of primary disease in AE, followed by possible spread of the tapeworm tissue to other organs through the lymph or blood. It is recommended that a comprehensive imaging examination be conducted to rule out metastatic lesions in the lungs and brain of patients with hepatic AE and to provide accurate information for surgery and achieve early detection, early diagnosis, and early treatment, improving the survival rate of patients.

## Acknowledgments

We especially thank Caili Su from Department of Pathology, Beijing Friendship Hospital, Capital Medical University, for the support with our research.
*Funding*: None.

## Footnote

*Conflicts of Interest*: All authors have completed the ICMJE uniform disclosure form (available at https://qims.amegroups.com/article/view/10.21037/qims-22-1336/coif). The authors have no conflicts of interest to declare.

*Ethical Statement*: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the Ethics Committee of Beijing Friendship Hospital Affiliated to Capital Medical University and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.



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**Cite this article as:** Wang Y, Yan Y, Wang Z, Wei X, Wang Z. Rare recurrent brain alveolar echinococcosis complicated by systemic multiorgan infection—a case description. Quant Imaging Med Surg 2023;13(7):4731-4736. doi: 10.21037/qims-22-1336

 