Secondary multiple intracranial hydatid cysts caused by intracerebral embolism of cardiac echinococcosis: an exceptional case of hydatidosis

Case report

MEHMET TURGUT, M.D., KEMAL BENLİ, M.D., AND MUZAFFER ERYILMAZ, M.D.

Department of Neurosurgery, Adnan Menderes University Hospital, Aydın, Turkey; and Departments of Neurosurgery and Radiology, Hacettepe University Hospital, Ankara, Turkey

The authors present an extremely rare case of secondary multiple cerebral echinococcosis caused by presumed intracerebral and arterial embolism of cardiac hydatidosis in a 7-year-old girl. The first manifestations were symptoms of raised intracranial pressure. Unfortunately, before the primary ruptured echinococcosis cyst was detected in the myocardium of the left ventricle, the patient underwent nine operations over an 8-year period for hydatid embolism affecting the brain and the femoral artery and was treated with concurrent mebendazole therapy. The combined therapy would not have been successful without removal of the intracardiac hydatid cyst. This case is unusual because of the multiplicity of intracranial hydatid cysts and its embolic origin from cardiac echinococcosis. The present case is believed to be the first one in which the development of an embolism in the brain was studied by magnetic resonance imaging.

Keywords • multiple hydatid cyst • hydatidosis • embolism • cardiac echinococcosis

ECHINOCOCOSIS is an infectious disease in humans caused by the larval stage of the cestode species Echinococcus granulosus or E. multilocularis. Dogs are the definitive host and the intermediate hosts are generally sheep but occasionally humans. The human hydatid disease caused by the E. granulosus strain is endemic in Turkey, and the most commonly affected organs are the liver and the lungs; hydatid cysts of the central nervous system (CNS) are unusual. Because contractions of the heart provide a natural resistance to the presence of viable hydatid cysts, primary echinococcosis of the heart is found very rarely. Its incidence is approximately 0.5 to 2% of cases in the literature, with the left ventricle being the most common site of cyst formation.1,4,7,12,17,20,23 Vaquero, et al.,27 documented a case of hydatid embolism from cardiac cyst rupture into the left ventricle during cardiac surgery for echinococcosis. Sierra, et al.,26 described a patient with bilateral intracerebral hydatid cysts of presumed embolic origin caused by spontaneous rupture of a hydatid cyst in the cardiac cavity. Recently, a case of cardiac echinococcosis with fatal intracerebral embolism was reportedly found at autopsy.3 We report an unusual case of secondary multiple cerebral echinococcosis caused by embolized fragments from a ruptured left ventricular hydatid cyst, which we studied by computerized tomography (CT) and magnetic resonance (MR) imaging with emphasis on long-term follow-up review.

Case Report

Examination. This 7-year-old girl presented with complaints of headache, nausea, vomiting, and weakness in the left side of the body. The patient recounted a history of contact with the neighbor’s dog. Ten days before, she had consulted a local doctor about her headache and received systemic antibiotic therapy with a diagnosis of sinusitis. Subsequently, she experienced head tilting to the left side. Her left arm lost strength, and she started to experience difficulty in walking. She presented to the pediatric neurology clinic with these symptoms. Neurological examination demonstrated central facial paresis, hemiparesis, and increased reflexes on the left side. Funduscopic examination revealed papilledema. She had a tendency to fall to the left side while walking. Dysmetria, dysdakinesia, ataxia, and speech disturbances were detected in cerebellar tests. Peripheral blood sampling showed eosinophilia, and multiple cystic lesions were detected on CT scans. The patient was diagnosed with hydatid cyst and received medical treatment. A course of mebendazole was initiated at a dosage of 10 mg/kg taken three times per day with meals for the first 6 days, after which the dosage was increased to 40 mg/kg per day. She was then referred to the neurosurgery clinic.

First Admission. A course of diphenylhydantoin and dexamethasone was initiated approximately 24 hours preoperatively. On September 19, 1985, the patient under-
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went operation. A bifrontal craniotomy was performed, and four cysts, ranging from 3 to 8 cm in diameter, were removed via a cortical incision. Because two of them ruptured during this procedure, the operative field was cleaned with 3% hypertonic saline solution, as advocated previously.18 Her left-sided hemiparesis recovered partially during the postoperative period. A second surgical intervention was performed on October 9, 1985. After the left parietooccipital craniotomy flap was lifted, two cysts measuring 4 cm in diameter were encountered and were delivered intact, using the Dowling technique.9

A hypodense appearance in the areas of previous operation and a typical abscess appearance in the left parietal lobe were demonstrated on the postoperative follow-up CT scan, which was obtained on December 2, 1985. Therefore, the patient underwent reoperation on February 12, 1986. A cystic lesion filled with a yellow–gray viscous material was found at a depth of approximately 3 to 4 cm beneath the cortex, and it was cultured. There was a 3-cm thick-membraned infected hydatid cyst at the inferolateral side of this lesion. It was removed intact. Postoperatively, antibacterial therapy with crystallized penicillin-G potassium and chloramphenicol was administered for 14 days. On the postoperative CT study, a similar infected cyst was seen in the right parietal region, located parasagittally. Therefore right parietal craniotomy was performed and the lesion was punctured. Because 5 cm3 of purulent material was obtained on puncture, this cystic lesion was excised together with its wall. There was no bacterial growth in the culture. The patient’s postoperative course was uneventful, and after antibiotic treatment was completed she was discharged from the hospital.

Second Admission. One year later, the patient presented to our clinic again with an increase in loss of strength on the left side of her body. On neurological examination, papilledema, dysphasia, left central facial paralysis, left-sided hemiparesis, and increased reflexes in the left side were present. Also, a positive Babinski response and Aschill clonus were found on the left side. Computerized tomography scanning of the head showed a cystic lesion on the right parietal lobe, and the patient underwent reoperation on March 11, 1986. A 4-cm cystic lesion was removed in toto and saline irrigation was administered by the surgeon. Therefore right parietal craniotomy was performed and the lesion was punctured. Because 5 cm3 of purulent material was obtained on puncture, this cystic lesion was excised together with its wall. There was no bacterial growth in the culture. The patient’s postoperative course was uneventful, and after antibiotic treatment was completed she was discharged from the hospital.

Third Admission. Four and one-half years later, she presented to the Pediatric Emergency Service with complaints of pain, numbness, and weakness in the right leg. On physical examination, cyanosis and lowered temperature were detected below the level of right knee. Posterior tibial and dorsalis pedis arterial pulsations were absent in this leg. A CT study did not reveal a cystic lesion or shift effect. After consultation with the cardiovascular surgery department, she underwent operation on October 16, 1991 after being diagnosed with acute arterial occlusion. A No. 4 French Fogarty catheter was advanced to 20 cm proximal and 35 cm distal to the right femoral artery. Pearly white gelatinous cysts were encountered in both the proximal and distal regions, and an embolectomy was performed to remove these materials. A treatment protocol composed of 80 mg per day of acetylsalicylic acid, 150 mg per day of dipyridamole, and 1600 mg per day of mebendazole was prescribed. Three months later, bilateral peripheral pulses were present on follow-up examination.

Fourth Admission. Ten months after she was discharged from the hospital, the patient presented to the neurosurgery department complaining of loss of strength in the left arm, nausea and vomiting for 8 to 10 days, and double vision for the last 1 to 2 days. Neurological examination revealed anisocoria (4 mm right and 2 mm left), bilaterally positive pupillary reflexes to light, papilledema, left homonymous hemianopsia, left central facial paralysis, left-sided hemiparesis (4/5 strength), increased reflexes, positive Babinski sign on the left, and disturbances in cerebellar tests. Psychometric evaluation performed using the revision of the Wechsler Intelligence Scale for Children showed that her intelligence quotient score was 40 points; therefore, she was diagnosed as severely mentally retarded. Multiple cystic lesions were seen on follow-up head CT scanning, two on the left frontal, one on the right frontal, and one on the right occipital lobes (Fig. 1), and the patient was hospitalized. Abdominal and pelvic ultrasonography examinations were normal, but ultrasonography of the neck showed stenosis of the middle portion of the left carotid artery and also hyperechogenic and irregular regions in the lumen of the vessel, which is normally anechoic. Multiple cystic lesions corresponding to hydatid cysts were present on cranial MR images (Fig. 2). Studies were subsequently conducted to explore the primary origin of the emboli. Echocardiography revealed a 12 × 20–mm mass lesion on the left ventricular surface of the posterior mitral valve. Repeated echocardiographic studies showed that this mass was contacting the valve and the valve was immobile. Cardiac MR imaging revealed that the source of infection was on the left ventricular surface and was linked with the mitral valve (Fig. 3).
A surgical intervention was planned for this cardiac hydatid cyst lesion after correction of increased intracranial pressure. The patient underwent operation on November 12, 1992. After bifrontal craniotomy, a cyst measuring 4 cm in diameter was encountered in the right frontal tip. There were also three cysts measuring 1.5 to 5 cm in diameter within the area adjoining the right motor cortex. All of the cysts were delivered intact via the “hydatid birth” technique, via an episiotomy of the cortex. Five cysts ranging from 2 to 4 cm in diameter were removed without rupturing in a second operative session conducted 1 month after the first procedure. On the early follow-up CT head scan, there were multiple cystic lesions in the left frontal and parietal regions as well as the right parietooccipital regions and it was necessary to reoperate. Two weeks later, the third operative session was conducted and a total of seven cysts were delivered intact via “hydatid birth.” In the postoperative period, a combined treatment consisting of anticonvulsant and antiparasitic drugs was administered. Before the patient was discharged from the hospital, a CT scan disclosed the presence of only residual changes in previously treated areas of the brain (Fig. 4).

**Fifth Admission.** Echocardiographic studies performed approximately 1 year later showed a cystic mass measuring $15 \times 27$ mm located between the posterior leaflet of the mitral valve and the posterior endocardium. This mass was limiting the movements of the posterior mitral valve, but the movements of the posterior septal wall were normal. There was a I/VI systolic murmur in all cardiac areas. The patient was hospitalized and underwent operation in the cardiovascular surgery department on January 15, 1993. A diagnosis was made of a cystic lesion inside the papillary muscle fibers of the mitral valve. A left atriotomy was performed, and a small cystic mass was found in the anterior mitral leaflet, with a large cystic mass situated inside the papillary muscle of the posterior leaflet, dis...
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turbing its structure. The mitral valve was resected, a 27-mm cardiac valve prosthesis (Monostrut; Shiley, Inc., Irvine, CA) was used to replace it, then the atriotomy was repaired (mitral valve replacement procedure). Postoperatively the patient received prophylactic anticoagulant, antithrombotic, and antiplatelet therapy with aspirin. The patient was started on a course of albendazole at a dosage of 10 mg/kg per day for 12 weeks. No serious side effects or cyst recurrence were observed during a 2-year follow-up period.

Discussion

Hydatid cyst disease is rare in the CNS and comprises only 1.6 to 5.2% of all hydatid cysts reported.\textsuperscript{6,9,10,13,15,18,21,26} The diagnostic and therapeutic characteristics of CNS hydatid disease were reviewed previously in two excellent articles from our clinic.\textsuperscript{13,14} First, Özgen, et al.,\textsuperscript{13} reported that cerebral involvement was seen in approximately 2.9% of patients with enlarging mass lesions. Second, Pamir, et al.,\textsuperscript{14} reported that spinal cord involvement was documented in 3.8% of patients diagnosed with spinal compression. Primary cysts are the most common types, and are always solitary; secondary cysts are usually multiple and follow embolization of cardiac cysts that rupture into the left ventricle, or are caused by spontaneous, traumatic, or surgical rupture of a primary CNS cyst.\textsuperscript{2} Typically, hydatid cysts observed on CT scans are spherical well-defined cystic lesions containing fluid with a density similar to that of cerebrospinal fluid and without ring enhancement.\textsuperscript{18,21} As a rule, secondary multiple hydatid cysts of the CNS caused by cardiac embolization are acephalocele and infertile. They are very rare and we have encountered only one such case among 130 patients treated for CNS hydatid cyst in our clinic in the past 30 years. Recently, however, Erongun, et al.,\textsuperscript{16} reported a similar case of infected hydatid cyst. The patient presented here had three suppurating cysts at the first admission to our clinic, whereas the other cysts were uninfected. In our literature review we found that the current study is the second case published in Turkey and that there have been only three similar cases reported in the English-language literature.\textsuperscript{3,11,16} On review of the world literature, we were able to find only 67 cases of secondary multiple hydatid cysts of the brain reported to date.\textsuperscript{1,4,5,8-10,13,15,22-27} Of the 67 cases reported in the literature, only five (7%) were caused by embolization from the rupture of intracranial hydatidosis; our case raises the number to six.

In our case, another interesting feature of intracranial hydatid cysts is their presumable embolization followed by spontaneous rupture of a cyst in the heart. To our knowledge, this is the first report in which development of cerebral hydatid cysts after hydatid embolism to the brain was studied using MR imaging. In our case, MR imaging allowed us to demonstrate some features of hydatid disease clearly, including the relationship between the hydatid cysts, the falx, and the pericystic edematous area. It is well known that one of the causes of multiple intracranial hydatid cysts is rupture of an embolism from cardiac echinococcosis into the left ventricle during cardiac surgery for hydatid cysts.\textsuperscript{1,4,16,27} In our patient, however, the source of the multiple cerebral hydatid cysts was hydatid embolism from the heart by spontaneous rupture of a cardiac cyst. Furthermore, there was an embolic dissemination of infection into the right femoral and the left internal carotid arteries, resulting in acute occlusion of these vessels.

Surgical treatment is usually required when hydatid disease attacks the brain. The recommended surgical procedure is cyst removal without rupture via the hydatid birth technique, known as the Dowling technique.\textsuperscript{3} However, there is no risk of rupture in secondary embolic cysts because of the typical infertility of scolices, in contrast to fertile primary cysts. It is important to remember that, in geographic areas in which hydatidosis is endemic, the possibility of parasitic emboli should be considered, particularly in young patients with otherwise unexplained cerebral embolism. Furthermore, in patients with secondary multiple intracranial hydatidosis caused by intracerebral embolism of cardiac echinococcosis, the surgical removal of the primary source is important to prevent multiple recurrences due to embolism, as were seen in our case.

References


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Address reprint requests to: Mehmet Turgut, M.D., Cumhuriyet Mahallesi, Cumhuriyet Caddesi, 2 Sokak Darcan Apartmani No: 1/6, TR-09020 Aydin, Turkey.