Cardiology in the Young

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Echinococcosis in both heart and lungs, the first case reported in Albania

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Abstract

An 11-year-old male was admitted with cough and fever for the last 4 days and also complained of pain in the right lung for some weeks. The boy did not show any other symptoms and his past medical history was unremarkable as well. The radiologist findings showed an aspect that suggested for echinococcosis.

At first, it was realised the heart intervention. About a 2-month period later, the child underwent another cyst removal in lung. He had begun taking albendazole 5 days before the heart intervention. The therapy was continued until the lung intervention and for 12 weeks post-operatively. The patient had an uneventful recovery and after about 4 years.

Original Article

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Echinococcosis was firstly described by Hippocrates and Galen.

Cystic echinococcosis is the larval cystic stage of a small taeniid-type tapeworm that may cause illness in intermediate hosts, generally herbivorous animals and people who are infected accidentally. It was Redi who realised in the 17th century that echinococcus is transmitted from animals to humans.[1,3]

In their normal life cycle, echinococcus species inhabit the small intestine of carnivorous definitive hosts, such as dogs, coyotes, or wolves, and echinococcal cyst stages occur in herbivorous intermediate hosts, such as sheep, cattle, and goats.[1,2]

Because humans play the same role of intermediate, they can also become infected by ingesting tapeworm eggs passed from an infected carnivore. This occurs most frequently when individuals handle or contact infected dogs or other infected carnivores or inadvertently ingest food or drink contaminated with faecal material containing tapeworm eggs.[1–3]

In primary echinococcosis, larval cysts may develop in every organ. Most patients (as many as 80%) have single-organ involvement and harbour a solitary cyst.[2]

Approximately 65% of patients experience liver echinococcosis, the second most common organ involved is the lung (25%).Cardiac hydatid cyst is seen rarely, occurring in about 0.5–2% of all cases of hydatid disease.[4] The first in-life diagnosis of heart echinococcosis was made by Kashin in 1862.

In each anatomic site, cysts are surrounded by the periparasitic host tissue (pericyst), which encompasses the endocyst of larval origin. Inside the laminated layer, the cyst is covered by a multipotential germinal layer. The central cavities of cysts are filled with clear fluid, numerous brood capsules, and protoscolices.[1,2]

Case presentation

An 11-year-old male was admitted to our hospital due to cough and fever for the last 4 days. He also complained of pain in the right lung for some weeks. The boy did not show any other symptoms and his past medical history was unremarkable as well.

In the physical examination, we did not notice specific findings or signs of respiratory or cardiovascular dysfunction.

Chest X-ray showed up an oval mass which was noticed in the right lung with well-defined borders, with regular contours, clear, homogeneous, aspect that suggested for echinococcosis. (Fig 1)

CT and MRI confirmed the presence of cyst echinococcus in the right lung but simultaneously noticed a big heart formation (Figs 2 and 3).

In the echocardiography, a cystic formation of 356 × 295 cm was depicted with a clear outline, encapsulated that occupied over the half of the cavity located mainly in the apex, but that did not prevent the flow in the mitral valve, so it was also confirmed the presence of echinococcus cyst in the left ventricle except the one in the lung (Fig 4).

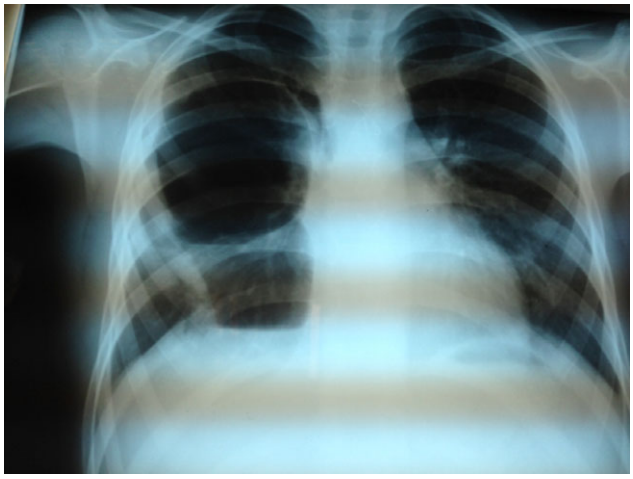

Figure 1. Chest X-ray showed up an oval mass, aspect for echinococcosis.

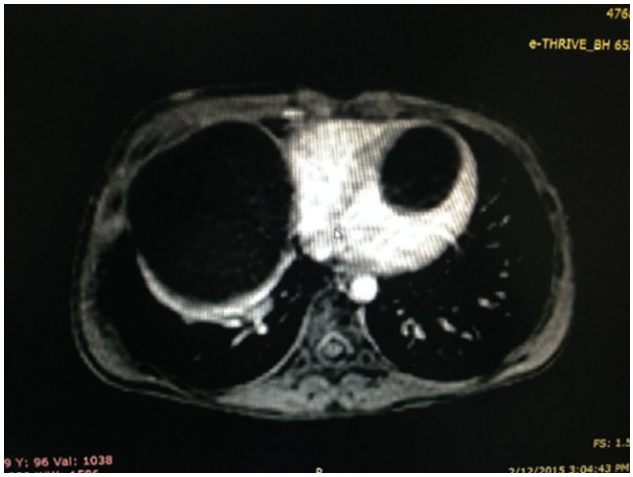

Figure 2. Presence of cyst echinococcus in torachal CT.

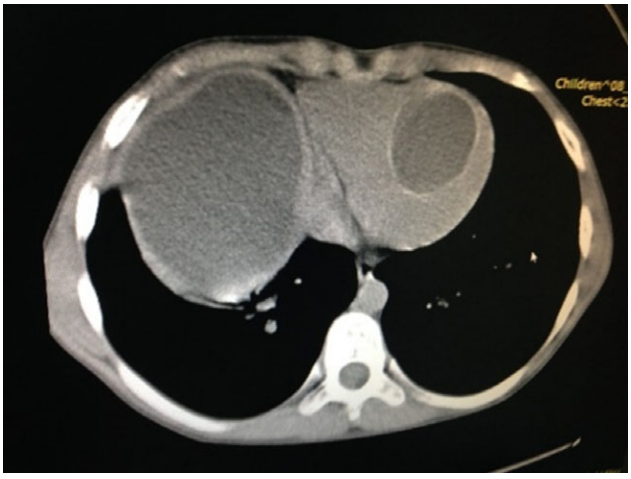

Figure 3. Presence of cyst echinococcus in torachal MRI.

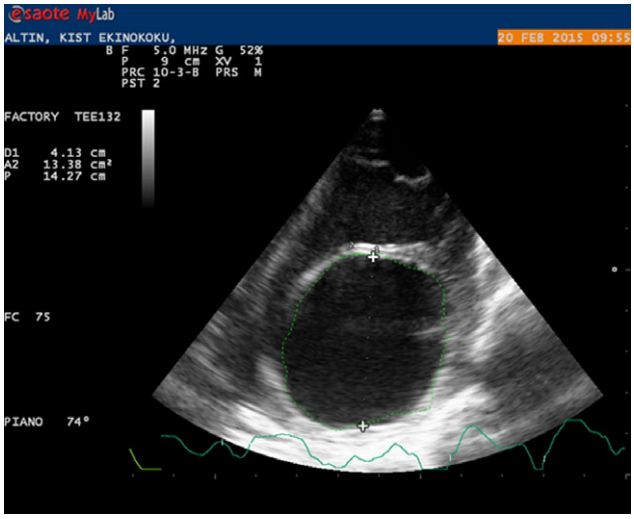

Figure 4. A cystic formation in echocardiography.

Fortunately, in our case, the diagnosis was clear, even the data connected to the surgery, so we do not need the transesophageal echocardiography for managing this case.

At first, it was realised the heart intervention. We used a transmitral approach to expose the cyst. A median sternotomy was performed and the pericardium was suspended. We placed gauzes impregnated with 20% of NaCl all around the heart. We proceeded on cardio pulmonary bypass through an aorto-bicaval cannulation. After cross-clamp, we administered tepid-mixed blood cardioplegia (one part was crystalloid cardioplegia and three parts were blood). We initiated the vertical left atriotomy anterior to the right superior pulmonary vein and posterior to the interatrial sulcus (Sondergaard's groove) and extend it superiorly behind the superior vena cava, avoiding injury to the right pulmonary artery, and inferiorly into the oblique fissure behind the inferior vena cava. The mitral valve was kindly retracted to expose the cyst. The cyst occupied the apex of the left ventricle without compromising the papillary muscles and tendine cords of the mitral valve. The cyst was first injected with 10 ml of 20 % NaCl liquid and after aspirated, being careful not to spread and not to suck the liquid. Then, we continued to delaminate the endocardial layer separating the cyst from the myocardium until we removed it intact from the left ventricle. The myocardial wall was thinner in the whole area where the cyst laid before. A hydrodinamic test for the mitral valve proved competent. Then the left atrium was closed with two running sutures. After de-airing the heart, we removed the cross-clamp (40 minutes) and we weaned the patient from the cardiopulmonary bypass machine.

Cytology of the aspirated cyst fluid was consistent with the diagnosis of hydatid cyst (Fig 5).

After the intervention, the patient suffered from atrial fibrillation, so he was treated for several weeks with anticoagulant (warfarine and clexane) and antiarrhythmic in order to get sinus rhythm.

After about a 2-month period, the child underwent another cyst removal in lung.

He had begun taking albendazole 5 days before the heart intervention. The therapy was continued until the lung intervention and for 12 weeks post-operatively.

The patient had an uneventful recovery and after about 4 years his condition becomes stable (Figs 6 and Fig 7).

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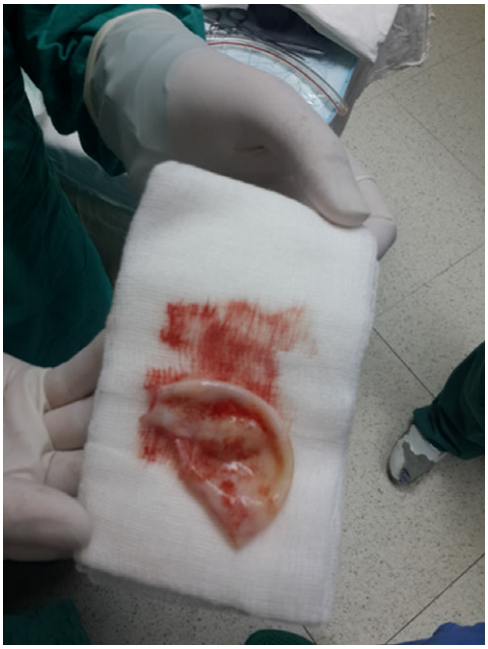

Figure 5. Surgically removed hydatid cyst.

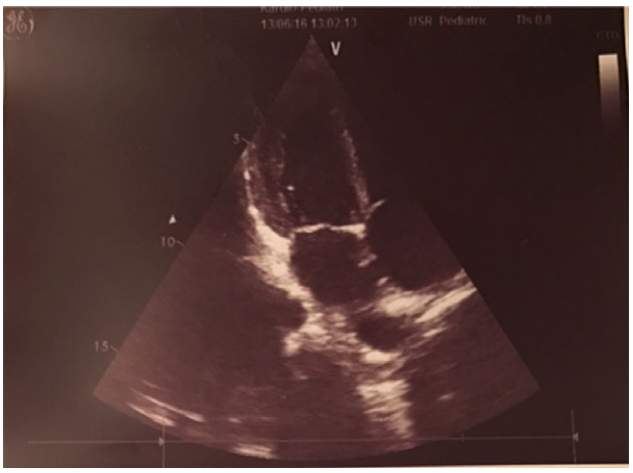

Figure 6. Echocardiography 4 years after surgery.

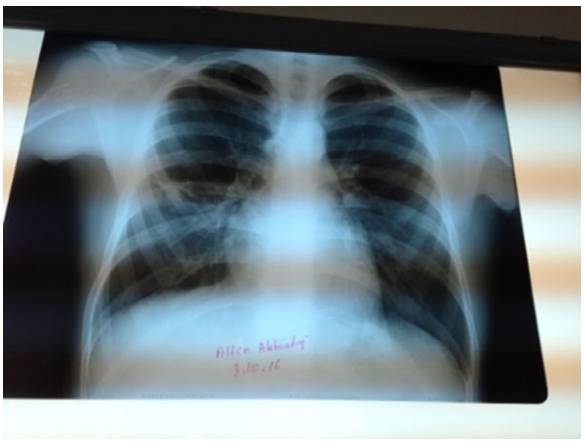

Figure 7. Chest X-ray 4 years after surgery.

Discussion

Because the clinical signs and symptoms of cardiac hydatid cyst are non-specific and highly variable, this disease may be difficult to diagnose.[5]

The clinical features or the spectrum of symptoms of cystic echinococcosis are highly variable and depend on the involved organs, size of cysts, and their sites within the affected organ or organs, the interaction between the expanding cysts and adjacent organ structures, the complications caused by rupture of cysts, bacterial infection of cysts and spread of protoscolices and larval material into bile ducts or blood vessels, immunologic reactions or membranous nephropathy secondary to release of antigenic material.[2,3]

Cystic echinococcosis is rarely fatal. Occasionally, deaths occur because of anaphylactic shock because of the release of cystic fluid due to cyst rupture or cardiac tamponade in heart echinococcosis.[6,11] Individuals of all ages are affected. In some endemic countries, children have higher infection rates because they are most likely to play with dogs.[3]

Months or years may pass before an individual exhibits any signs or symptoms of infection with the cystic larval stages. During the natural course of infection, the fate of E.granulosus cysts is variable. Some cysts may grow to a certain size and then persist without noticeable change for many years. Other cysts may rupture spontaneously or collapse and completely disappear.

After a variable incubation period, infections may become symptomatic if cysts are growing and exerting pressure on adjacent tissue and inducing other pathologic findings.

Sudden symptomatology is usually due to spontaneous or traumatic cyst rupture.

Usually, cysts do not induce clinical symptoms before they have reached a size sufficient to exert pressure on adjacent organs. The presentation of human echinococcosis is protean. Patients come to the clinician's attention for different reasons, such as when a large cyst has some mechanical effect on organ function or rupture of a cyst causes acute hypersensitivity reactions. The cyst may also be discovered accidentally during radiographic examination, body scanning, surgery, or for other clinical reasons.[16] Common chief symptoms are upper abdominal discomfort and pain, poor appetite, and a self-diagnosed mass in the abdomen. Physical findings are hepatomegaly, a palpable mass on the surface of the liver or other organs, and abdominal distention. If cysts in the lung rupture into the bronchi, intense cough may develop, followed by vomiting of hydatid material and cystic membranes.[7]

No standard, highly sensitive, and specific serologic test exists for cystic echinococcosis antibody detection, but serologic studies can be useful in confirming a diagnosis of echinococcosis, even the false-negative rate may be as high as 50%. Children aged 3–15 years may produce minimal serologic reactions.[2,8]

Ultrasonography is the procedure of choice when making the diagnosis of asymptomatic cystic echinococcosis because it is safe, non-invasive, and relatively inexpensive. Ultrasonography is an imaging technique that uses the reflection of ultrasound waves emitted by a probe on the bodily organs to build images of the organs explored.[17]

In 2003, the World Health Organization Informal Working Group on Echinococcosis proposed a standardised ultrasound classification based on the active–transitional–inactive status of the cyst as suggested by its sonographic appearance.[9,12]

Although diagnostic value of transthoracic echocardiography in cardiac echinococcus is well established, the role of transesophageal

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echocardiography in both the diagnosis and the management of this entity is not well known.[13]

CT scanning is indicated when ultrasonography is unsatisfactory, particularly it has the advantage of inspecting any organ (lungs cannot be explored with ultrasonography), detecting smaller cysts when located outside the liver, locating cysts precisely, and sometimes differentiating parasitic from non-parasitic cysts.[14]

Today MRI has been accepted as superior in many circumstances. MRI may have some advantages over CT scanning in the evaluation of post-surgical residual lesions, recurrences, and selected extrahepatic infections, such as cardiac infections.[15]

For simple, accessible cysts, the preferred therapy involves ultrasound- or CT-guided percutaneous aspiration, instillation of hypertonic saline or another scolicidal agent, and re-aspiration (PAIR). Compared with surgical treatment alone, PAIR plus albendazole results in similar cyst disappearance with fewer adverse events and fewer days in the hospital. Compared to albendazole alone, PAIR with or without albendazole provides significantly better cyst reduction and symptomatic relief. Spillage with PAIR is surprisingly uncommon, but prophylactic albendazole therapy is routinely administered 4 hours or even 1 week prior to PAIR or surgery and continued for 1 month thereafter.[1]

Mebendazole and albendazole are the only anthelmintics effective against cystic echinococcosis. Albendazole and mebendazole are well tolerated but show different efficacy.

Albendazole is significantly more effective than mebendazole in the treatment of liver cysts. Benzimidazole treatment alone requires prolonged administration over many weeks, with an unpredictable outcome in terms of response rates in individuals.[10]

Duration of therapy and doses are also important. Albendazole efficacy increases with courses of up to 3 months in the more common cyst sites.[8,18]

The puncture of echinococcal cysts has long been discouraged because of risks of anaphylactic shock and spillage of the fluid.[11]

These patients must be evaluated for ultrasonographic appearance modifications and changes in serology titres after 3 months of treatment and then for about 4 years.[9,10]

Conclusion

In cases of an interventricular cardiac hydatid cyst, the combination of surgical resection and albendazole therapy typically yields excellent results.

Surgical excision under cardiopulmonary bypass is the treatment of choice.

Chemotherapy in the post-operative period can decrease recurrence in many instances.

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Conflicts of interest. None.

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