

Surgical Treatment of Isolated Cardiac Echinococcosis: Report of Five Cases

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Objective: Cardiac echinococcosis is a rare pathology and in about half of cases it is not accompanied by other organs involvement. We report on our experience of surgical treatment of isolated cardiac echinococcosis (ICE).

Methods: Among 5 patients with ICE operated in our clinic 1 had pericardial and epicardial involvement, 1 patient had the single cyst in the apicolateral wall of the left ventricle and 3 patients had singular cysts of the interventricular septum. All patients underwent chest X-Ray, transthoracic echocardiography, ultrasound examinations of the internal organs, hemagglutination tests and microbiologic evaluation of operative specimens.

Results: Two operations were performed on a beating heart and three - using cardiopulmonary bypass. In cases of intramyocardial localizations the cyst enucleation and closure of residual cavities by plication were performed and in a case of peri- and epicardial involvement the extirpation of cysts and partial pericardiectomy were carried out. All patients postoperatively received therapy with albendazole. No intra- and postoperative complications were observed. Control echocardiography did not reveal any disturbances of myocardial performance. Two years follow-up showed absence of cyst recurrence.

Conclusion: The prevailed localization of echinococci cysts in ICE is the myocardium of the LV and IVST, places with the highest myocardial mass and good perfusion. Ideal echinococcectomy is an effective surgical treatment of isolated cardiac echinococcosis. (*Anadolu Kardiyol Derg, 2003; 3: 137-143*)

Introduction

Hydatid disease with formation of hydatid cyst in parenchymatous organs and caused by tapeworm *Echinococcus granulosus* is usually seen in the farmer regions where contact with sheeps and dogs is high. This parasite usually settles and forms cysts in lung with frequency about 20-30% of cases and in 60% of cases in liver, with higher prevalence of lung involvement in children (1). The involvement of other organs may occur usually in case of rupture of cysts and dissemination of scoleces by blood stream.

However isolated cardiac echinococcosis (ICE) is

a rare pathology and its frequency ranges between 0.01 and 2% (2). Because of localization in myocardium and pericardium it may lead to different clinical manifestations and life-threatening complications (3-8) necessitating aggressive treatment. Though antiparasital treatment with albendazole has been shown to be effective in the treatment of lung echinococcosis (9-10) cardiac occurrence requires more active treatment, as operation. As we have mentioned, hydatid cysts may form anywhere in heart including intracavitary, intramyocardial, intrapericardial localizations, causing mechanical obstruction at localization sites and specific manifestations (3-8).

Despite the cardiac echinococcosis is a very rare localization; it has gained a growing interest during last years because of evidence of early diagnosis and surgical treatment (3-8). Rarity of intracardiac localization of echinococci cysts, variety of clinical manifestations, diagnosis and success of surgical treatment forced us to present our experience of surgical management of cardiac echinococcosis.

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Materials and Methods

For the period from 1999 to 2001 years totally 5 patients with ICE were examined and operated at the department of cardiac surgery of our clinic. Patient's age ranged between 9 and 40 years, among them 3 girls of 9, 12 and 19 years, consequently, 10 years old boy and adult male of 40 years. One patient had presented with signs of jugular vein distention, hepatomegaly, hypotension and fever, he had also been observed in district hospital with diagnosis of effusive pericarditis; 2 - with signs of oedema and hepatomegaly; 1- with complaints of dyspnea, weakness and one was suffered from palpitations.

All patients underwent clinical examinations, electrocardiography, 2D-transsthoracic echocardiography (ECHO CG), as well as ultrasound investigation of liver, kidneys, spleen and pancreas, and chest X-Ray were performed in order to exclude the internal organs involvement. Computerized tomography (CT)

for the diagnosis and establishment of cyst localization was done in one patient with multiple cysts of pericardium and epicardium. Hemagglutination test was performed in all patients and operative specimens (membranes and cyst content) were analyzed histologically and microbiologically.

Two-dimensional echocardiography in the 1st patient showed signs of pericardial effusion surrounding the heart: 2.5 cm behind left ventricular (LV) posterior wall and 1.5 cm behind LV lateral wall. There were also multiple echolucent cysts of epicardium and pericardium with smooth contours, among them one large egg-like at the apex with signs of right ventricle (RV) compression, and similar large cyst behind lateral wall of the LV (Fig 1). In another four cases echocardiography revealed singular echolucent cysts in the apicolateral wall of the left ventricle (LV) in 1 case and in the interventricular septum (IVST) (Fig 2.) in 3 patients (membranous and muscular parts with bulging into the right ventricle (RV) in 2 ca-

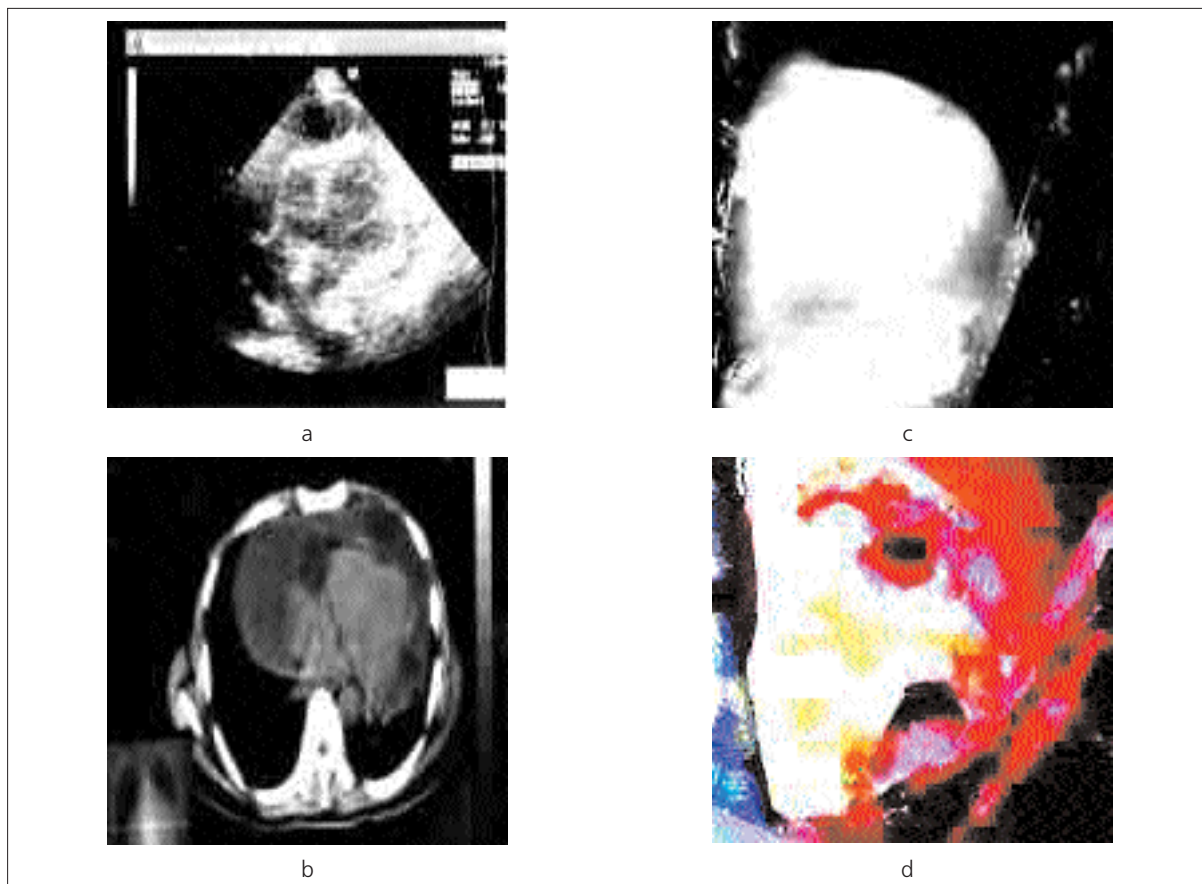


Figure 1: Four chamber echocardiographic view of large pericardial fluid and big egg-like cyst at the apex of the heart (a); computer scan of the gross pericardial fluid and oval form cysts of epi- and pericardium (b); intraoperative view of the hydatid cyst before (c) and after cyst removal and below the orifice of the opened chitin membrane of another cyst could be seen (d) in the same patient.

ses, and with bulging into both ventricles in 1 case). Computer tomography performed in 1 case confirmed presence of pericardial effusion and multiple unilocular cysts with smooth contours (Fig 1b).

No evidence of other organs involvement were observed by ultrasound examinations and chest X-Ray.

Operation of echinococectomy (enucleation of chitin membranes) with preliminary puncture and evacuation of cyst content, sterilization of cavity with scolicedal agents (iodine solution in 4 four case and both iodine and hypertonic solutions in 1 case), removal of chitin membrane, and further closure and plication of fibrous capsule were performed in 4 patients with intramyocardial localizations of hydatid cysts, and in patient with multiple cysts of pericardium and epicardium the extirpation and partial pericardiectomy were done. In three cases of cysts localization in the interventricular septum operations were performed using cardiopulmonary bypass by conventional technique and in two cases (multiple cysts

of epicardium and pericardium, singular cyst of apico-lateral wall of LV) operations were accomplished on a beating heart.

In case of multiple hydatid cysts of epicardium and pericardium operation of echinococectomy and cystectomy was carried out through median sternotomy on a beating heart. The heart was covered with caseous masses and contained multiple cysts of different size. Moreover there was about 1700 ml of green liquid containing chitin membrane residuals (Fig 1c). Intraoperative control revealed 12 cysts of 1 to 8 cm in diameter, located subepicardially (Table 1) and in thickened (up to 8 mm) pericardium. Totally 5 cysts from epicardium and 7 cysts from pericardial surface were removed by enucleation and extirpation of residual fibrous caps, and partial pericardiectomy at the areas with deep location of small cysts in pericardial surface (Fig 1d). The approach to the cyst behind lateral wall of the LV was difficult and its removal was not performed.

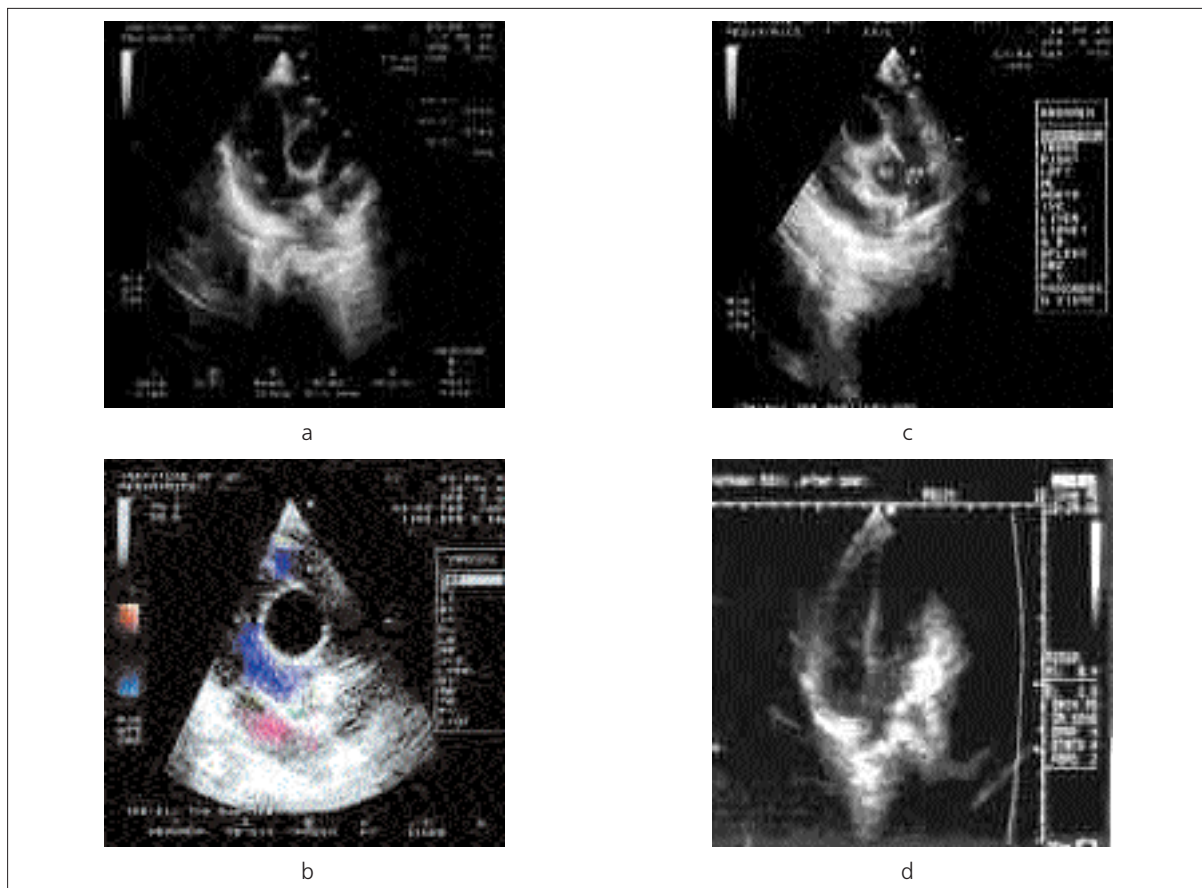


Figure 2: Modified apical long axis view of the heart displaying the oval large cyst of the interventricular septum with smooth contours and echonegative content (a); color Doppler imaging of the same view (b); short axis view at the aorta level demonstrating the cyst propagating to the right ventricular outflow tract (c) and the 4-chamber apical view of the heart after cyst enucleation (d) in 10 years old boy with hydatid cyst of interventricular septum.

In the second case there was an intramyocardial location of the cyst of 4X4.5 cm in size at the apico-lateral wall of the left ventricle. In this case operation approach was through the left antero-lateral thoracotomy at the 4th intercostal space and operation of cyst enucleation (11 ml of translucent fluid was evacuated) was performed on a beating heart.

In three other cases of hydatid cysts localization within the IVST cysts were removed through median sternotomy on the "open" heart and using cardiopulmonary bypass and the elongated incision of the right atrium.

Intraoperatively in case of involvement of muscular part of IVST cyst was 3 cm in diameter, containing 12 ml of fluid and bulging into the RV. In another case large cyst was seen in the membranous part of the interventricular septum (3.5 cm in diameter) with its dissection. The puncture of the cyst was performed under the septal cusp of the tricuspid valve just below the crista supraventricularis and about 14 ml of translucent fluid was evacuated. Tricuspid valve chordae were not injured during operation. In the third case of IVST localization of hydatid cyst int-

raoperative revision showed cyst (5 cm in diameter) originated from IVST just beneath septal cusp of the tricuspid valve, bulging into both ventricles, and with fibrous cavity adherent to the chordae and papillary muscles. The cyst was approached through the incision of the septal cusp of the tricuspid valve and about 30 ml of cyst content was removed.

In all cases the special attention was done to avoid contact of antiscolicidal iodine solution with adjacent myocardial surface while irrigating the fibrous capsule cavity.

The analysis of the cyst's content and membranes were performed in all patients and in all cases diagnosis of echinococcosis was confirmed histologically and microbiologically.

Results

Nobody of patients had signs of complications in early and late postoperative periods. On control echocardiography performed after operations the signs of pericardial effusion were relieved in 1st case, and only small residual echo-signals were detected.

Table 1: Clinical and intraoperative findings of patients with isolated cardiac echinococcosis

Patients	Presentation	Transthoracic echocardiography	Intraoperative findings	Operation
1- male, 40 years old	Neck vein distention, hepatomegaly, hypotension, fever, pericardial effusion	Pericardial fluid, multiple cysts of pericardium and epicardium (large cysts at the apex of the heart and behind lateral wall of LV)	5 large cysts of epicardium: - an apical anterior wall of RV (7-8 cm in size), area between VCS and RAA, RVOT beneath PA, RV posterior wall, LVOT, and 7 cysts adjacent to pericardial surface	Cyst extirpation, partial pericardiectomy, beating heart
2- female, 19 years old	Palpitations	Singular echolucent cyst at the apico-lateral wall of LV -	Apico-lateral wall of LV- 4-4.5 cm in size	Cyst enucleation, plication of fibrous cavity, beating heart
3- female, 12 years old	Oedema, hepatomegaly	Singular echolucent cyst at the muscular part of IVST	Muscular part of IVST - 3 cm in diameter, bulging in RV	Cyst enucleation, plication of fibrous cavity, CPB
4- male, 10 years old	Oedema, Hepatomegaly, weakness	Singular echolucent cyst at the membranous part of IVST	Membranous part of IVST - 3.5 cm in diameter, bulging in RV	Cyst enucleation, plication of fibrous cavity, CPB
5- female, 9 years old	Dyspnea, Weakness	Singular echolucent cyst at the IVST	IVST below septal leaflet of tricuspid valve 5 cm in diameter, bulging in both ventricles	Cyst enucleation, plication of fibrous cavity, CPB

CPB- cardiopulmonary bypass, IVST- interventricular septum, LV- left ventricle, LVOT- LV outflow tract, PA - pulmonary artery, RAA - right atrial appendage, RV-right ventricle, RVOT- RV outflow tract, TTE-trans thoracic echocardiography, VCS - vena cavae superior

ted at the myocardium of IVST and LV lateral wall in 4 cases of myocardial localization, may be due to residual connective tissue after removal of chitin membrane. No signs of contractility disturbances or diastolic dysfunction were noted after operation.

The treatment with albendazole in doze of 15mg/kg/daily was started after operation in all patients and continued for 6 months. No adverse reactions were observed.

All patients were discharged from the hospital without complications and were taken albendazole. Control examinations at the end of the 2nd year after operation did not reveal recurrence of hydatid cyst in heart and other organs.

Discussion

Echinococcosis is a parasitic disease caused by *E. granulosus*, which more commonly affects parenchymatous organs. Cardiac involvement in the setting of echinococcosis is rare and about 50% of cases are accompanied by other organs involvement, while another half manifests only by heart involvement (2, 11). According with our and other literature findings (2, 4, 5) the prevailed localization of hydatid cysts is myocardium of the LV and interventricular septum. It may be explained by good vascularization and higher possibility of parasite invasion from systemic circulation to the well perfused myocardium with further formation of cysts (2, 11, 12, 13). Cardiac localizations are usually intramyocardial affecting more often ventricles, then atria and rare are intracavitary (3-5, 7, 8, 13-16). The pericardium involvement is thought to be due to rupture of subepicardial cysts and dissemination of its content in pericardium (6, 13).

In our cases all have intramyocardial localization of cysts affecting IVST and LV, except pericardium and epicardium cysts. In the latter case we thought that the involvement of pericardium developed mainly due rupture of subepicardially located myocardial cysts with dissemination of their content and formation of multiple daughter cysts.

Previous investigations have demonstrated that about 22% of cardiac echinococcosis cases are incidental findings, because of absence of specific symptoms and variety of clinical manifestations, as well due to diagnosis was difficult up to recent time (2).

The singular cysts located on the surface of myocardium or intramurally may not clinically manifest for a long period of time. Only with enlargement of the cyst size they could be found out on chest X-Ray (accidental findings). In these cases they usually sho-

uld be differentiated from the cysts of pericardium and lungs. With further increase of the cysts size the clinical manifestations are due to syndrome of compression of the cardiac chambers (2-8). Clinically these lesions manifest by arrhythmias, heart block and signs of left or right ventricular outflow obstruction in cases of location of hydatid cysts in the IVST and signs of cardiac compression in the setting of pericardial involvement (2-8), anaphylactic shock, even emboli in case of rupture of intracavitary cysts (15).

Usually, pericardial and intramyocardial cysts are presented as singular cysts with smooth contours, detected by echocardiography, computer tomography and magnetic resonance imaging (4, 14, 17).

The diagnosis of cyst may be well established by transthoracic echocardiography by showing the cyst with its echonegative content and smooth contours. And usually in case of singular nonruptured cysts the intraoperative correlation is high both for transthoracic echocardiography (TTE) and computed tomography (18, 19). However in the cases of multiple cysts specially after rupture and dissemination with formation of daughter cysts the transesophageal echocardiography (TEE) and intraoperative echocardiography have the advantage to precisely locate small cysts, as well as to display those cysts that could not be shown by TTE (13, 20). Though TTE is quite informative for establishment of diagnosis (16, 19), the use of transesophageal approach is superior specially in the planning of operative approach (13, 20).

Taking in account the danger of cyst rupture with formation of daughter cysts and risk of emboli, presence of alive cyst is an indication for surgical treatment (11, 12, 13, 21, 22, 23). It is important to consider the localization, number and size of the cysts while choosing the operative approach and use of cardiopulmonary bypass or performing operation on a beating heart (21, 23). Echinococcectomy (enucleation of the cyst) is the method of choice in case of alive parasite (12, 13, 21). The generally accepted method is puncture and needle aspiration of the cyst content before extirpation of cyst (12, 13), because of potential danger of dissemination of cyst content over myocardium and development of anaphylactic reactions with cyst rupture. This method is also useful for the establishment of diagnosis by analyzing of the cyst content if the imaging techniques are inconclusive (13).

However in cases of difficulties of cystectomy/extirpation the chitinectomy may be done after aspiration of the cyst content with further washing out the fibrous cavity with scolicidal agents like hypertonic solution, iodine, and remained cavity may be closed with obliteration or plication (12, 24).

There are several concerns how to deal with residual cavities after removal of chitin membrane. Several authors used to close the residual cavities with patches or glue (25), while others argue on the risk of abscess formation (13, 26). Birincioglu and colleagues considered all residual cavities should be left open for the self closure by secondary healing, because in case of closure by suture there may be regional contractility and relaxation abnormalities, even tears in myocardium (13, 23, 26). We performed suture closure and plication of residual pericyst cavity in all 4 cases of intramyocardial location of the cyst including IVST localization and no above mentioned complications including acute tear, pump or relaxation abnormalities were observed during echocardiographic follow-up. Previous studies on IVST localizations applied similar techniques without complications (12), however reported (13, 26) complications should be kept in mind.

The another point is selection and use of scolical agents for prevention of scoleces dissemination and relapses. Different agents have been used with this aim as hypertonic solution, iodine solutions and alcohol with reported different success in their scolical efficacy (21, 27). The major concern is regarding the use of alcohol and high concentration of alcohol based iodine solutions, which while contact with adjacent myocardium may cause coagulation of tissues and creating the source for emboli. However, we use iodine solution for cleaning the remnant fibrous cavity with special attention to prevent contact with adjacent tissues, because of its higher scolical efficacy than of hypertonic solutions (27, 28).

The postoperative complications include myocardial tear (13, 26), development of atrioventricular blocks requiring pacemaker implantation (12), ventricular arrhythmias and sudden death due to the scar (12).

Along with the surgical treatment the use of benzimidazoles for prevention of cyst recurrences is widely used nowadays. Albendazole is a benzimidazole that has been shown to be effective in the treatment of lung hydatid disease without operation and to prevent recurrence of cysts after operation on lungs (9). Moreover it has been shown that albendazole given before operations may limit dissemination of parasite during enucleation. The main mechanism is reduction of parasite growth. The typical dosages are 400 bid with cycle treatment. Though, treatment by albendazole in cases of lung echinococcosis was accompanied by such complications as thinning of the cyst membrane and rupture when advocated before surgery on lung hydatidosis (29), no such complications have been reported when albendazole was administered after operations on heart. Other adverse effects of the treat-

ment with albendazole include gastrointestinal disorders, alopecia, neutropenia, proteinuria, increase in transaminases levels and neurological disorders (30). Albeit, nobody of our patients had such adverse effects and we also did not observe cyst recurrences after surgery during 2 years of follow-up.

The main limitation of our study is that we did not perform transesophageal or intraoperative echocardiography which could be very useful in case of multiple cysts of epicardium and pericardium. Secondly due to technical difficulties in access we could not remove the cyst behind posterolateral wall of the LV in the same patient.

In conclusion: the prevailed localization of echinococci cysts in ICE is the myocardium of the LV and IVST, places with the highest myocardial mass and good perfusion. Ideal echinococectomy is an effective surgical treatment of isolated cardiac echinococcosis.

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