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Case Report

Cardiac Echinococcosis, a Multidisciplinary Approach to the Diagnosis and Treatment of This Rare Entity: Two Case Reports and Literature Review

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Abstract

We present two case reports of cardiac echinococcosis. Case 1 was a 33-year-old woman with hepatic and cardiac echinococcosis. The parasitic cyst was located intramyocardially in the free wall of the left ventricle leading to cranial dislocation of the left circumflex coronary artery (LCx). The patient was successfully operated. Case 2 was a 28-year-old woman with hepatic and cardiac echinococcosis. The parasitic cyst was located in the left ventricular myocardium in the area of the apex and manifested clinically as paroxysms of ventricular tachycardia. The ultrasound study showed a 3.2×2.8 cm cyst dislocating the papillary muscles and causing moderate mitral regurgitation.

Bulgaria ranks first in the European Union in terms of the number of echinococcosis patients. Although cardiac involvement is uncommon, occurring in only 0.5%–2% of cases, it can cause a wide range of clinical symptoms. Multimodal imaging is a key step in the management of patients with cardiac involvement.

Keywords

cardiac echinococcosis, computed tomography, echocardiography, magnetic resonance imaging, surgical treatment

INTRODUCTION

Echinococcosis is a parasitic disease caused by *Echinococcus granulosus*, *Echinococcus multilocularis,* or *Echinococcus vogeli*. Dogs and cats are the main carriers of this parasite. Humans are intermediate hosts in the life cycle of echinococci and become infected by ingesting eggs from tapeworms that have developed in the intestinal tract of the final host.[1] Hydatid cysts can be found in a variety of tissues and organs, but the liver and lungs are the most usually affected – 50%-70% and 20%-30% of cases, respectively.[2] Cardiac involvement in hydatid disease is an unusual and rare presentation of parasitosis, occurring in only 0.5-2% of all cases.[3]

CASE REPORTS

Case report 1

We present a clinical case of a 33-year-old woman employed in animal husbandry. She was admitted to a cardiology clinic because of scapular and precordial pain with positional dependence. Radiography revealed an oval, homogeneous opacity measuring 51 mm in diameter over the left cardiac contour. Ultrasound revealed cysts in the liver and a large cyst lateral to the left ventricle in the pericardium ***(Fig. 1)****.*

Computed tomography was performed to clarify the cardiac findings: it found a large, 56×63×65 mm intramyocardial cyst located anterolaterally to the left ventricle and presence of peripheral calcifications in the wall. The cystic formation caused cranial dislocation of the left circumflex artery (LCx), with the ramus intermedius (RIM) passing over it. A well-circumscribed calcified lesion was found in the 6th hepatic segment with the presence of thin folded membranes in depth with a characteristic water-lily sign ***(Fig. 2)****. In order to better define the myocardial involvement and the risk of pending rupture, we performed cardiac magnetic resonance imaging (CMRI). CMRI showed a large complex formation on the upper-lateral wall of the left ventricle from the level of the mitral valve to the midmyocardial level and a length of 5 cm. In depth, the formation was liquid-equivalent, with a peripherally located rim of uneven thickness, with intermediate to low signal intensity in T1 and T2 weighted images and with the presence of areas of signal loss. In myocardial perfusion, the formation remained unperfused, and in the late postcontrast series, intense peripheral enhancement of the formation was seen with good delineation of the formed capsule. In the dynamic CINE images, impaired contractility was found in the area of the involved myocardium, while in the other segments, the contractility was relatively preserved, with an extremely reduced left ventricular volume. The posterior mitral leaflet was displaced by the formation but remained free* ***(Fig. 3)****.*

Laboratory tests revealed mild anemia with hemoglobin of 112 g/l. The transaminases were within the reference range. Serological studies found elevated IgG for echinococcosis using the ELISA method (1.5, normal value <1.1).

On the basis of all imaging and laboratory tests performed and the anamnesis for professional employment in animal husbandry, we established the diagnosis of liver and cardiac echinococcosis.

After multidisciplinary discussion, which included a parasitologist, it was determined that the risk of rupture during treatment with albendazole is unacceptably high due to the large size of the cyst and the substantial myocardial involvement. The patient underwent successful surgical treatment.

Case report 2

A 28-year-old woman working in animal husbandry was admitted to the cardiac surgery clinic for surgical treatment of an echinococcal cyst with intramyocardial location in the area of the apex of the heart. The clinical picture included non-sustained ventricular tachycardia diagnosed with a 24-hour Holter monitoring, easy fatigue, and loss of appetite. Echocardiographic examination revealed a cystic oval forma-

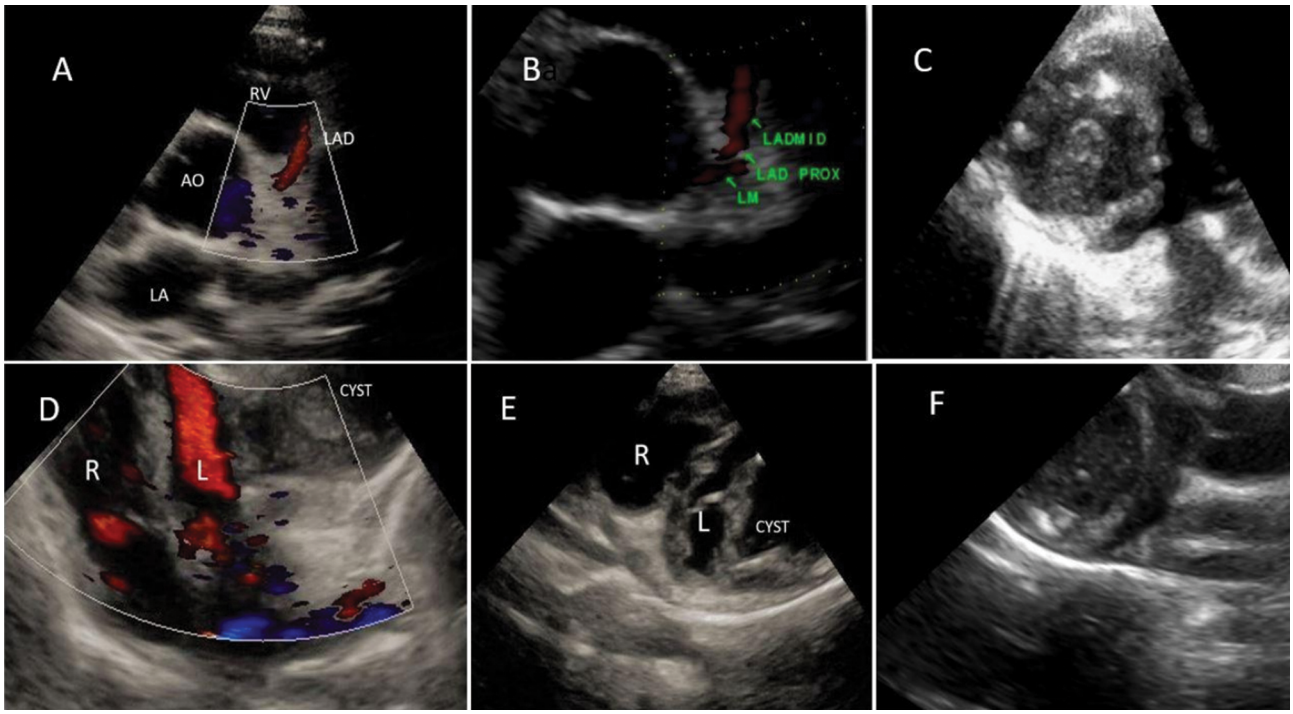

Figure 1. **A**, **B**. Left main and LAD coronary arteries; **C**. The cyst with calcinosis; **D**. A4C view; **E**. Short axis view; **D**. Parasternal view focused on the relation to the aortic valve.

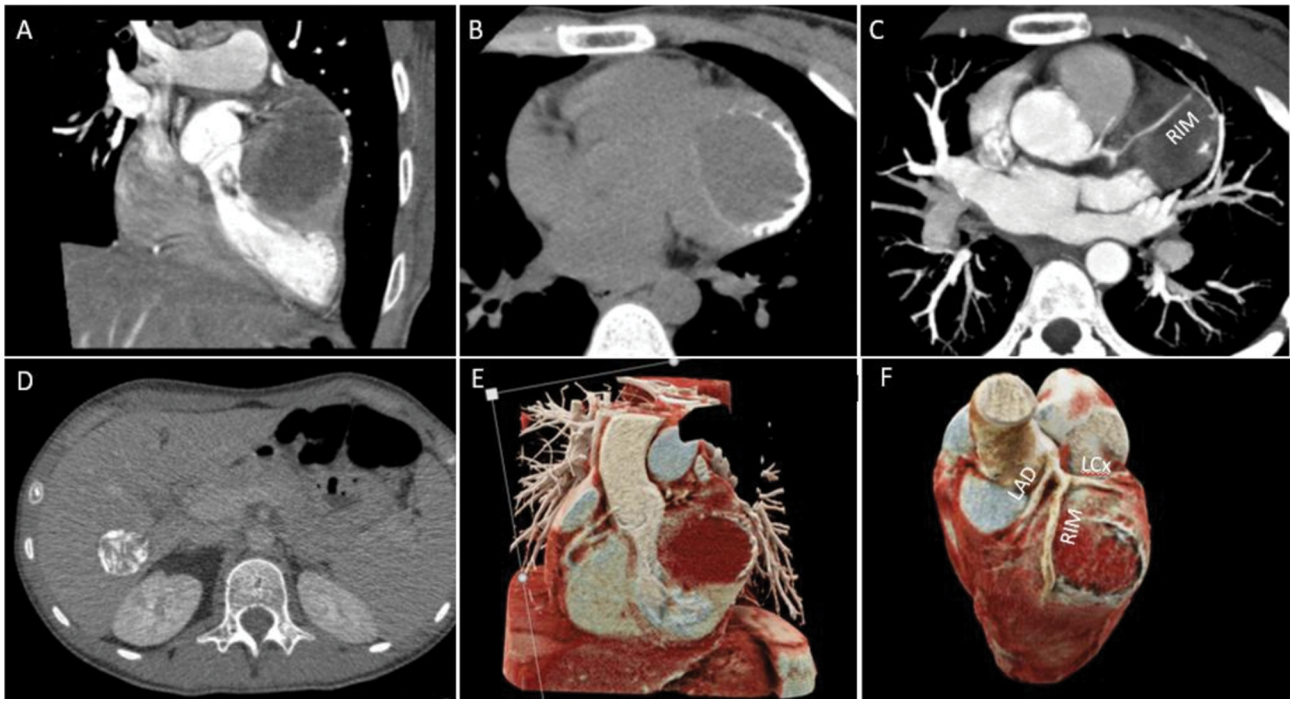

Figure 2. Computed tomography demonstrating the cystic lesion in the heart, located in the left ventricle lateral wall with additional 3D cinematic VRT. The typical peripheral calcifications are well demonstrated (**B**). RIM passing over the cyst (**C**). Liver lesion with typical water-lily sign is also shown (**D**). A coronal view of the cyst (**E**) and volume rendering of coronary arteries (**F**). LAD: left anterior descending artery; LCx: left circumflex artery; RIM: ramus intermedius.

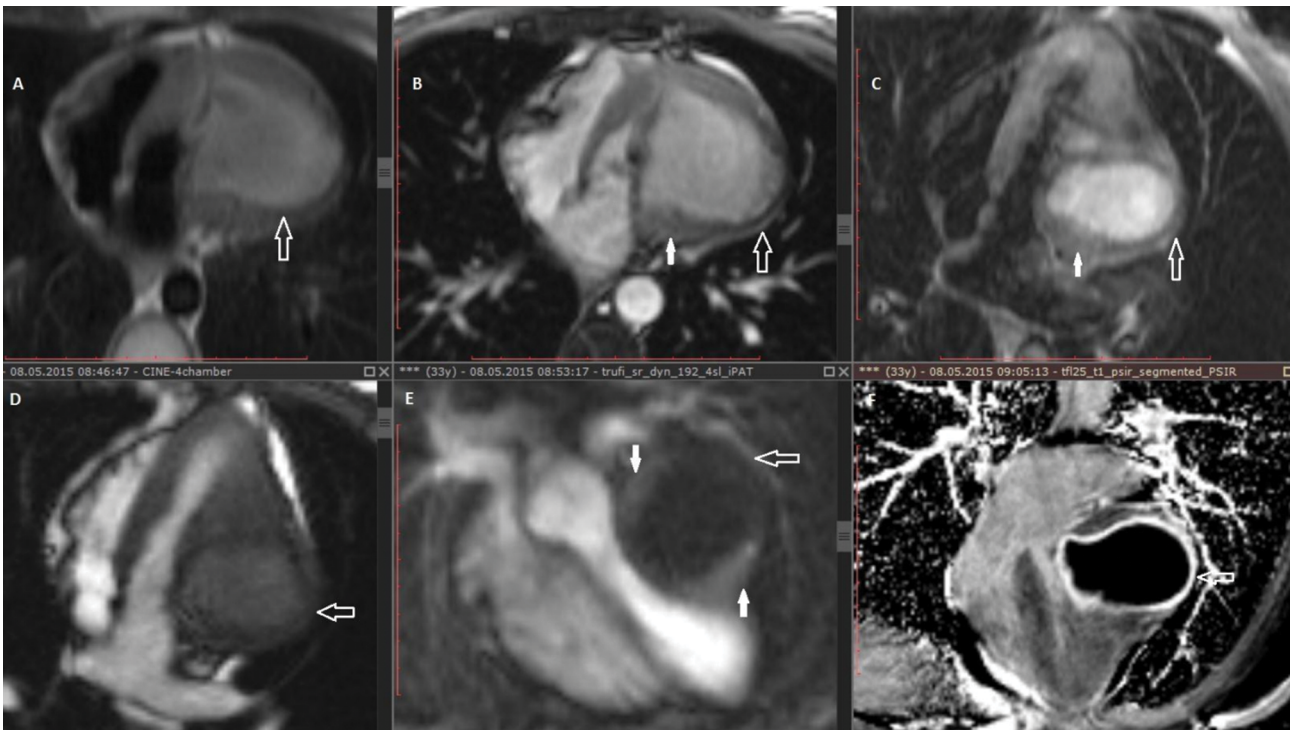

Figure 3. CMR images demonstrating cystic lesion in the lateral free wall of the left ventricle. The cyst shows high signal on HASTE (**A**), SSFP (**B**), TIRM (**C**) sequences and is surrounded by a thin hypointense rim (**C**). Dynamic first pass myocardial perfusion (**D**, **E**) shows no enhancement of the intramyocardial mass and in the same time well depicts the intramyocardial location of the lesion, showing the normally perfused myocardium surrounding the lesion (**E** - white arrows). Late gadolinium enhancement shows intense enhancement in the fibrous capsule (**F**).

tion measuring 3.2×2.8 cm in the area of the heart's apex and slightly laterally, causing dislocation of the papillary muscles and moderate mitral regurgitation. A second cystic formation was found in the liver measuring 2×1.5 cm. The patient underwent surgical treatment to remove the echinococcal cyst from the heart and subsequent drug therapy of the liver cyst with albendazole.

Laboratory tests revealed elevated levels of IgG for echinococcosis. Median sternotomy and extracorporeal circulation with ascending aortic and right atrium cannulation were used for surgical access. Myocardial protection was performed with blood cardioplegic solution infused antegradely in the aortic root. Examination of the left ventricular myocardium revealed a specific deformity in the area of the heart's apex of the underlying cyst. An incision was made in the myocardium to reach the cuticular membrane, which opened. The germinative membrane located inside was removed and the cuticular cavity was treated with 80% glycerol solution and hypertonic NaCl solution for 20 minutes. The site of the myocardial incision was closed with felt strips with a wrapped suture ***(Fig. 4)****. The early postoperative period passed without complications for the patient, and the degree of mitral regurgitation decreased to a minimum. She was referred to an infectious disease specialist for medical treatment of the liver echinococcal cyst with albendazole.*

DISCUSSION

Humans are intermediate hosts in the life cycle of echinococci and become infected by ingesting eggs from tapeworms that have developed in the intestinal tract of the final host.

Echinococcosis is an anthropozoonosis, a cosmopolitan parasitic infection caused in our latitudes by *Echinococcus granulosus*.[1] Man is an intermediate host and becomes infected by ingesting eggs of the tapeworm that has developed in the intestinal tract of the final host - carnivorous predators (dog, wolf, jackal).[1] Hydatid cysts in humans gradually increase in size, reaching up to 20 cm in diameter and a volume of several liters.[1] Historically, cardiac echinococcosis was first described by Williams in 1836, and the first successful surgery was performed in 1932 by Long. The incidence of echinococcosis in humans varies from 1 to more than 50 cases per 100,000 (in endemic areas).[4] According to the annual report of the European Center for Disease Prevention and Control (ECDC) for 2017, Bulgaria reported the highest number of cases of echinococcosis in the European Union (218 cases out of a total of 832 confirmed cases in the EU).[5] The incidence of cardiac involvement in the affected individuals is low – only 0.5% to 2% of cases of echinococcosis are located in the heart.[4] The liver and lungs are the most commonly affected organs.[2] Cardiac involvement includes various symptoms such as tachycardia, dyspnea, chest pain (including anginal pain due to coronary artery compression), tamponade, abdominal pain with hepatosplenomegaly, and acute stroke.[6] Rupture of intracardiac cysts is the most serious life-threatening complication that can cause acute pericarditis or tamponade, systemic or pulmonary embolisation, and severe anaphylactic shock.[7]

The preferred site for hydatid cyst growth in the heart is the free wall of the left ventricle (60% of cases). The higher incidence of left ventricular involvement is probably due to its greater myocardial mass and pressure variations, which provide better conditions for the development and growth

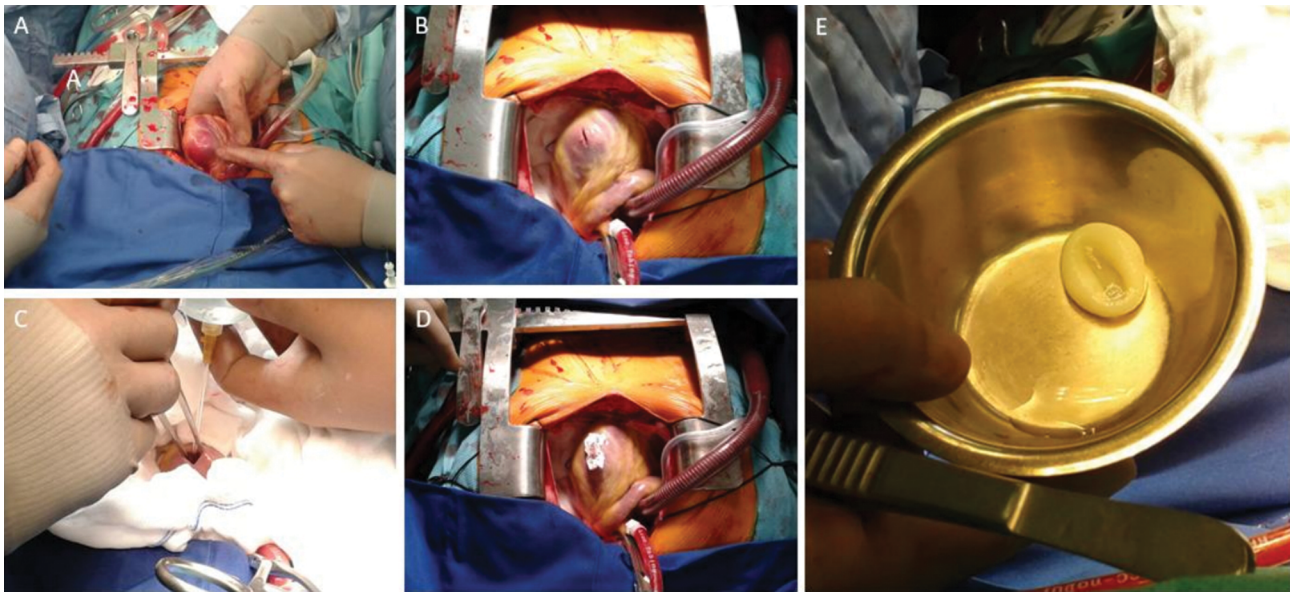

Figure 4. Intraoperative view of the heart and the extracted germinative membrane. **A**. Apex ot the left ventricle with the prominent cyst. **B**. Incision of the left ventricle. **C**. Tretament of the cuticular cavity with 80% glycerol solution and hypertonic NaCl solution. **D**. The sutured left ventricle after the procedure. **E**. The extracted germinative membrane.

of the parasite.[8] The next most commonly involved cardiac structure is the right ventricle (10%), followed by pericardium (7%), pulmonary artery (6%), left atrial appendage, and the interventricular septum (4% of cases). One of the possible mechanisms of myocardial invasion is through the coronary circulation. The second route of infection is by rupture of a primary pulmonary or hepatic echinococcosis cyst, through the pulmonary veins or inferior vena cava.[9] In a review including six case reports of cardiac echinococcosis, the left ventricle free wall was reported to be affected in five of the patients.[6] Primary pericardial cysts are rare and more often pericardial involvement is due to rupture of a myocardial cyst. Liver cysts are also found in most of the described cases.[6] In all cases, varying degrees of altered cardiac contours were observed on chest radiograph, and in 4 patients ECG changes resembling ischemia were described.[6] Physical findings in patients with cardiac echinococcosis may be normal or heart murmurs may be present (in case of valve obstruction or regurgitation), as well as pericardial friction (in case of pericardial involvement) can be heard. Eosinophilia is often present in the blood count. In cases of liver or lung disease, positive serology is observed in 80% and 65% of cases, respectively. When other organs are affected, the incidence of seropositive patients decreases, and when the heart alone is affected, it is less than 50%.[10]

Diagnostic imaging

The most common first-line method in imaging is echocardiography with 90% sensitivity in cases of cardiac involvement.[11] The advantages of this modality are that it is non-invasive, easily accessible, relatively inexpensive, and sensitive method. A contrast agent may be used if necessary. The differential diagnosis of cardiac masses includes cysts, tumors, vegetations, blood clots, calcifications and some others. In cases of difficulties in the diagnostic process or the need for a more detailed anatomical localization of the echinococcal cyst, computed tomography and magnetic resonance tomography of the heart are capable of supplying the necessary additional information. CT and CMRI can show the cystic structure of the lesion and its exact location in the heart cavities. CT is the best method to visualize the echinococcal cyst-specific calcifications in the wall. CMRI, due to its good soft tissue resolution and the possibility of tissue characteristics, has an advantage in depicting the typical morphological characteristics of echinococcosis. CMRI finding of a hydatid cyst usually includes an oval lesion that is hypointense on T1 sequences and hyperintense on T2. Typically, there is the presence of the so-called pericyst - hypointense peripheral ring of T2 images which is a dense fibrosis capsule consisting of modified host cells: fibroblasts, giant cells, and eosinophils.[9,12] Over time, the cyst may degenerate and present as a solid intracardiac mass, making it difficult to be differentiated from some cardiac tumors such as myxoma. Individual membranes from the cyst wall can be detached and 'floating' linear structures can be imaged inside the lesion. First pass myocardial perfusion allows the differentiation of the hydatid cyst from some cardiac tumors. Echinococcal cysts do not show contrast enhancement.[8]

When cardiac echinococcosis is detected, four different therapeutic approaches could be applied as appropriate: surveillance, drug therapy (albendazole), percutaneous therapy, or surgery, which is the definitive treatment for cardiac echinococcosis.[13]

Cardiac surgery

The surgical excision of echinococcal cysts aims to prevent the life-threatening complications the cysts can cause. Depending on the location, number, and size of the cysts, surgical access can be sternotomy, thoracotomy or performed in several stages. In practice, the median sternotomy is most often used. Yan et al., regarding surgical treatment of echinococcosis, used sternotomy in 20 of the cases and left thoracotomy in the remaining six.[15] In multifocal echinococcosis, an abdominal surgical approach can also be used. The principle in surgical treatment is to remove those cyst that cause the highest risk of complication. In addition to one-step surgical interventions, in multifocal echinococcosis, it can be staged. In the research, the authors give priority to pulmonary cysts, followed by those in the heart and liver. This can be accounted for by the increased risk of rupture of pulmonary cysts during anesthesia and mechanical ventilation. In cardiac echinococcosis, if the cyst is located in the pericardial space, the operation is performed on a beating heart, while intramyocardial cysts require extracorporeal circulation and cardioplegic arrest of the heart.[14] The purpose of the surgical intervention is to remove the germinal membrane and prevent the dissemination of helminths in the circulation. There are three main methods of surgical treatment of echinococcosis: aspiration cystectomy, enucleation of the germinal membrane, and total excision of the cyst. Cyst excision is an appropriate method for pericardially positioned cysts, while aspiration cystectomy and enucleation of the germinal membrane are considered for intramyocardial localization. After removal of the germinal membrane, the underlying cavity is treated with 70-95% alcohol or 15-20% hypertonic NaCl solution for at least 15 minutes. After surgery, patients remain on drug therapy with albendazole 10-15 mg/kg for 28-30 days followed by a 14-day break, with 1-6 such courses.

CONCLUSIONS

Diagnosis of cardiac echinococcosis, especially in the early stages of the disease, can be difficult due to the various and non-specific nature of the symptoms. The diagnosis should be considered in the presence of a cystic intracardiac lesion, especially in endemic areas, even in negative serological tests. Echocardiography, CT, and CMRI are useful for elucidating the structure, size, exact location, and

relationship to surrounding tissues, which is also crucial in selecting the right therapeutic approach. Surgical removal of echinococcal cysts remains the mainstay of treatment for this disease. In the cases of intramyocardial cysts, surgical intervention is performed with extracorporeal circulation and cardioplegic arrest of the heart to eliminate the risk of hematogenous dissemination. Subsequent therapy with anthelmintic drugs is a prerequisite for successful treatment.

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Эхинококкоз сердца, междисциплинарный подход к диагностике и лечению этого редкого заболевания: два клинических случая и обзор литературы

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Резюме

Мы представляем два случая эхинококкоза сердца. Случай 1: женщина 33 лет с эхинококкозом печени и сердца. Паразитарная киста располагалась интрамиокардиально в свободной стенке левого желудочка, что привело к краниальной дислокации левой огибающей коронарной артерии (LCx). Больная была успешно прооперирована. Случай 2: женщина 28 лет с эхинококкозом печени и сердца. Паразитарная киста располагалась в миокарде левого желудочка в области верхушки и клинически проявлялась пароксизмами желудочковой тахикардии. При УЗИ выявлена киста размером 3.2 × 2.8 см, вывихивающая папиллярные мышцы и вызывающая умеренную митральную регургитацию.

Болгария занимает первое место в Европейском Союзе по количеству больных эхинококкозом. Хотя поражение сердца встречается редко, всего в 0.5 – 2 % случаев, оно может вызывать широкий спектр клинических симптомов. Мультимодальная визуализация является ключевым шагом в лечении пациентов с поражением сердца.

Ключевые слова

эхинококкоз сердца, компьютерная томография, эхокардиография, магнитно-резонансная томография, оперативное лечение