CASE REPORTS

Diagnosis of suspected cardiac echinococcosis with negative serologies: role of transthoracic, transesophageal, and contrast echocardiography

Amgad N. Makaryus\textsuperscript{a}, Craig Hametz\textsuperscript{a}, Jennifer Mieres\textsuperscript{a}, Smadar Kort\textsuperscript{a}, Justine Carneglia\textsuperscript{a}, Judy Mangion\textsuperscript{b,*}

\textsuperscript{a}Division of Cardiology, North Shore University Hospital, NYU School of Medicine, Manhasset, NY, USA
\textsuperscript{b}Echocardiography Lab, Hartford Hospital, University of Connecticut, 80 Seymour Street, Hartford, CT 06102, USA

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Abstract The early recognition and treatment of hydatid disease of the heart is important as it can result in potentially lethal complications. We present the clinical and echocardiographic features of a 71 year old Afghanistani man who presented with left-sided chest pain. Transthoracic (TTE), transesophageal (TEE), and contrast echo demonstrated a calcified cystic structure within the distal anterior septum consistent with an echinococcal cyst, despite negative serologies. Treatment strategies for this patient are discussed.

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KEYWORDS
Cardiac echinococcosis;
Hydatid cyst;
Serologies;
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Introduction

Cardiac echinococcosis is a rare clinical entity that accounts for only 1\% of all cases of echinococcosis. Most human cases originate in areas of the world where sheepherding is common such as Africa, the Middle East, India, Afghanistan, Pakistan, Southern Europe, and Latin America. The disease is exceptionally rare in the United States. Even in endemic countries, the incidence of cardiac echinococcosis is thought to be less than 2\%.\textsuperscript{1}

Clinically, the disease may present with chest pain, shortness of breath, and palpitations. However, many patients are initially asymptomatic. Early recognition and treatment of cardiac echinococcosis is important as the risk of rupture of the cyst is high, and may result in potentially lethal complications from embolization and anaphylactic reactions. Depending on the location of the cyst, there can be a variety of different cardiac manifestations including valvular dysfunction, conduction block, cardiac tamponade, and outflow tract obstruction.\textsuperscript{1–3}

Echocardiography is often the first and frequently the only required imaging modality to establish the diagnosis. Classic echocardiographic features include the presence of a cystic mass...
Figure 1  ECG findings include normal sinus rhythm, incomplete right bundle branch block, left anterior fascicular block, and non-specific T wave abnormality.

Figure 2  Dual isotope vertical long axis image during stress and rest demonstrating a discrete fixed defect in the mid-anterior septal wall.
within the myocardium or pericardium that may become calcified with multiple septae and multitectual hypoechoic contents.

Case presentation

A 71 year old male Afghanistani immigrant with a history of hypertension and asthma was admitted to our hospital with left-sided chest pain of one week’s duration. The pain was squeezing in nature and associated with diaphoresis. Symptoms were promptly relieved with sublingual nitroglycerin. On physical examination, the patient appeared overweight, and was afebrile. Cardiac auscultation revealed a grade II/VI systolic murmur heard best at the apex. ECG showed sinus rhythm with incomplete right bundle branch block, left anterior fascicular block, and non-specific T wave abnormality (Fig. 1). Cardiac enzymes were normal. Complete blood count with differential was notable for eosinophilia (7.1%).

Figure 3  (a) TTE; apical four chamber view. There is a single 2.5 × 2 cm calcified cystic mass within the distal anterior septum (arrows). (b) Following administration of Optison contrast, there is no evidence of contrast uptake by the mass.
The patient underwent dual isotope stress testing which showed abnormal myocardial perfusion with a discrete fixed defect in the mid-anterior septal wall. This was read as being more consistent with a mass than with a region of ischemia (Fig. 2).

He was referred for TTE which revealed a single 2.5×2 cm calcified cystic mass within the distal anterior septum (Fig. 3(A)). Optison contrast was given intravenously, and there was no evidence of contrast uptake within the mass (Fig. 3(B)). TEE was then performed and suggested the presence of septations within the calcified cystic mass (Fig. 4). Chest X-ray, CT, and abdominal ultrasound studies did not reveal any cystic disease of the liver or chest. ELISA testing for echinococcosis and cysticercosis was negative. The patient signed out of the hospital against medical advice and ended with further workup or therapy.

Discussion

Echinococcosis and the hydatid disease which it causes are endemic to areas of the world with tropical climates. Echinococcal eggs are ingested by humans through contaminated foods and close contact with infected animals. The organism reaches the liver through the portal circulation after ingestion into the human GI tract. In 1% of cases, the organism can invade into myocardial tissue through the coronary circulation or through pulmonary cyst rupture into the pulmonary veins. Areas of cardiac involvement include the left ventricle (60%), right ventricle (10%), pericardium (7%), pulmonary artery (6%), and left atrial appendage (6%), with rare involvement of the interventricular septum (4%).

The diagnosis of cardiac echinococcosis is difficult. Patients may experience symptoms secondary to myocardial pressure due to expansion or rupture of the cyst. Rupture of the cyst can cause embolic phenomenon, anaphylactic shock, rhythm and conduction abnormalities, hemodynamic disturbances including cardiac tamponade, and ischemia. Eosinophilia is present in up to 25% of patients, but is non-specific. Echocardiography is often the first and frequently the only required imaging modality to establish the diagnosis. CT scanning, MRI, and TEE with harmonic capabilities may help to better visualize the cyst with its septae, the surrounding tissue, and to exclude co-existing hydatid locations. Features of hydatid disease include calcification and separation of the membrane from the wall, septations, daughter cysts, and ruptured cysts. Although serologic tests are commonly used, echocardiography is more sensitive than serologies.
when isolated cardiac echinococcosis is suspected. Indirect hemagglutination and ELISA tests are sensitive for hepatic cases (85–98%), less sensitive for lung involvement (50–56%), and poorly sensitive for other organ involvement (25–56%).

The treatment of cardiac echinococcosis begins with antiparasitic medications, but frequently requires complete surgical removal of the cyst. Albendazole and mebendazole have been used in patients with small and uncomplicated cysts. However, treatment with these medications results in cyst disappearance in only 30% of cases. Most patients require total surgical excision as definitive treatment. During surgery, care must be taken to avoid rupture of the cysts and prevent metastatic disease. Therefore, surgery is undertaken after three to four days of therapy with albendazole or mebendazole. With risk of rupture felt to be high in our patient, surgery was considered the treatment of choice, however, unfortunately our patient left the hospital against medical advice, and was lost to follow-up.

Conclusion

We present a case of a 71 year old Afghanistani man with chest pain who was found to have echocardiographic findings consistent with a calcified cardiac echinococcal cyst. Serologies were negative in this patient, although CBC with differential demonstrated 7.1% eosinophilia. The lack of additional cystic organ involvement made the case particularly challenging. Since risk of rupture with cardiac echinococcal cysts is high, aggressive surgical treatment is usually recommended, with decisions frequently based solely on clinical history and cardiac imaging.

References