

EXTENSIVE CARDIAC HYDATIDOSIS: IMAGING FINDINGS AND REVIEW OF LITERATURE

Ummara Siddique Umer, Sadia Gul, Shahjehan Alam, Seema Gul, Syed Ghulam Ghaus, Aman Nawaz Khan, Shafqa Fayyaz, Shazia Naweed

Department of Radiology, Rehman Medical Institute, Peshawar, Pakistan.

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ABSTRACT

The purpose of this report is to review the CT findings of cardiac hydatid disease and to emphasize the role of multidetector CT scan in diagnosing cardiac hydatidosis. The most common locations of cardiac hydatid cysts are left ventricle (LV), interventricular septum and right ventricle respectively. We reported three cases of cardiac hydatid disease, in which one patient had extensive lesions in pericardium and cardiac walls. Second patient had single lesion in the interventricular septum while third patient had lesion in wall of LV. The diagnosis was not definite on echocardiography, but MDCT was utilized to assess cysts and extent of disease. We present the CT imaging details and review of literature in cardiac hydatidosis.

Keywords: Hydatid cyst, cardiac hydatid, hydatidosis, MDCT.

Introduction

Cardiac hydatidosis is an uncommon but potentially fatal disease¹ accounting for 0.5% to 2.0% of all hydatid infestations² and can occur as isolated cardiac involvement or as part of a widespread systemic infection. It remains endemic in developed and developing countries. The most common locations of cardiac hydatid cysts are left ventricle (LV), interventricular septum and right ventricle respectively.³ It may remain asymptomatic for a remarkably long period of time. It is usually detected accidentally because there is no specific clinical picture of disease. Initial diagnosis is made on echocardiography but both computed tomography (CT) or magnetic resonance imaging (MRI) should be used for further identification.

Extent of involvement: Cardiac involvement occurs by invasion of the myocardium, first through the coronary artery circulation. The second route of infestation

is the pulmonary vein from rupture of pulmonary echinococcal cysts into the vein. The left ventricle is more often involved than the right ventricle, possibly because of the dominance of the left coronary artery, which brings blood to the left ventricle; the greater myocardial mass in the left ventricle, which provides optimal conditions for development of the parasite; and the varying pressure conditions.⁴ Right-sided cardiac hydatid cysts have characteristics different from those of left-sided cysts. Right-sided cysts have a tendency to expand intracavitary and subendocardially and they rupture more frequently, so they lead to pulmonary embolus (as seen in our patient), anaphylaxis or sudden death. Rupture into the pericardial cavity can lead to pericarditis, effusion, and cardiac tamponade, whereas left-sided cysts tend to grow subepicardially.⁵ The heart also can be secondarily affected by direct contact with hydatid cysts originating from the liver or the lung. Arterial

Correspondence : Dr. Ummara Siddique
Department of Radiology,
Rehman Medical Institute,
Peshawar, Pakistan.
Email: ummara_81@hotmail.com

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involvement with a hydatid cyst usually develops after a cardiac hydatid cyst ruptures and embolism of the germinative membrane causes acute symptoms. Hydatid cyst inside the pulmonary arteries is an exceptional localization and most frequently a consequence of embolism from primary cardiac locations.⁶ Other hypothesis are that the parasite can cross the arterial wall through previous small breaks in the intima or aneurysms or by entering the vas nutritia. Pericardial involvement in hydatid disease is rare and frequently occurs as a result of rupture of a primitive hydatid lesion, the location of which is almost exclusively cardiac. Intrapericardial rupture of a cyst can lead to pericarditis, effusion, and cardiac tamponade, which can be fatal.⁷

We reported three cases of cardiac hydatid disease, in which one patient had extensive lesions in pericardium and cardiac walls.

Case Report

We present a retrospective analysis of rare cases of cardiac hydatidosis. 3 patients were diagnosed with cardiac hydatidosis in Radiology department of Rehman Medical Institute Peshawar between 2013 and 2016. Contrast enhanced CT was performed on 128-MDCT Toshiba scanner. One patient had single lesion in the interventricular septum, second patient had lesion in wall of LV, whereas third patient had extensive lesions in pericardium and myocardium. The diagnosis was not definite on echocardiography, but MDCT was utilized to assess cyst activity and extent of disease. Patients with lesions in ventricular wall were referred for suspicion of fluid collection, whereas the patient with extensive hydatid cysts had echocardiographic suspicion of pericardial effusion with septae. We present details of this case with multiple hydatid cysts in the heart and pericardium and provide a literature review of this rare condition: A 27 year-old man presented to Radiology department for cardiac CT. He had burning sensation in chest for 2 days. His ECG showed changes of inferior wall myocardial ischemia. Echocardiography revealed multiple septations in pericardium. He was referred to Radiology department for CT scan. CT chest was done with cardiac protocol and showed multiple cystic

lesions in the pericardium and cardiac walls. One unilocular cyst was bulging into the left atrial chamber. Another cyst along left pericardium was invading into the main pulmonary artery consistent with pulmonary embolism (Fig. 1 & 2). Echinococcus granulosus titer was positive for cystic hydatidosis. Cardiac hydatidosis was diagnosed because of typical imaging findings and positive serology.

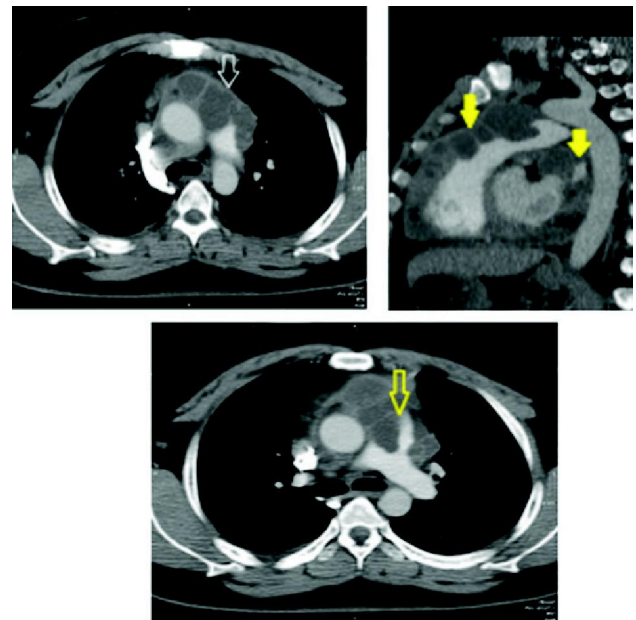


Figure 1: Axial and sagittal cardiac CT images showing multiple hydatid cysts in pericardium. There is infiltration into the main pulmonary artery (open arrow in lower image)

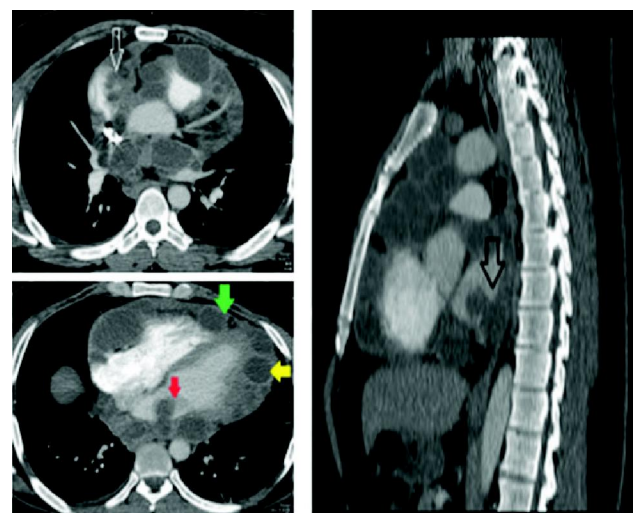


Figure 2: Multiplanar CT images showing multiple pericardial cysts, few seen invading into the cardiac chambers (open black arrow).

Imaging Features

Echocardiography, CT, and MRI can show the cystic nature of the mass and its relation to the cardiac chambers. CT best shows wall calcification, depicts the exact anatomic location and nature of the internal and external structures. MRI is the technique mostly used for post treatment follow-up⁸. The appearance of hydatid cysts on CT is usually characteristic, oval or rounded cystic lesions of fluid density with no post contrast enhancement of walls. If contrast enhancement is seen, then possibility of superadded infection should be considered. In cardiac involvement, single or multiple clumped cysts are seen in the myocardium, which can extend subendocardially. The cysts located within the myocardium and septum tend to bulge into the chamber cavity, which sometimes show direct infiltration into the atria and ventricles, which is considered as direct embolization (Fig. 2). The pericardial cysts on the other hand appear as cystic lesions in the pericardial space causing mediastinal widening. Presence of internal membranes is variable.

The cysts may be single or multiple, uniloculated or multiloculated, and thin or thick walled. More specific signs include calcification of the cyst wall, presence of daughter cysts, and membrane detachment. They can become solid in cases of chronic dead cysts and can sometimes be difficult to differentiate from heart tumors.⁹ A variety of tumors in the heart and a congenital pericardial cyst must be considered in the differential diagnosis, especially cystic metastasis; however, the multivesicular nature of the cystic mass and membranes indicate the true diagnosis. Cardiac hydatidosis can be an isolated finding or associated with other visceral hydatid cysts e.g in liver and lungs. On MRI, cysts follow the fluid signals appearing hypointense on T1-weighted images and hyperintense on T2-weighted images. A typical finding on T2-weighted images is a hypointense peripheral ring, which represents the pericyst.

Discussion

Human beings are exposed to echinococcus through direct contact with hosts or by eating contaminated, insufficiently cooked food. Embryos enter the portal

or lymphatic circulation from the human intestine. Hydatid disease results from the expansion of parasitic cysts in the visceral organs. Cardiac involvement is uncommon. Larvae reach the myocardium through the coronary circulation. Cardiac involvement through the pulmonary veins has also been reported.¹⁰ Hydatid cysts can involve any part of heart and pericardium. Left ventricle (LV) is most frequently involved (in 55% - 60% of reported cases),¹⁰ followed by the right ventricle (15%), the interventricular septum (9%), the left atrium (8%), the right atrium (4%), and the interatrial septum (2%).¹¹ The frequency of pulmonary artery involvement has been reported to be 7% to 8%, and of pericardial location, 5%.¹² In a case series,¹³ the most common sites of occurrence were the LV and IVS (and not the RV), apparently due to the high myocardial mass and good perfusion in both regions. Once the larvae reach the myocardium via the coronary arteries, cysts form in 1 to 5 years.¹⁴ Due to the slow growth of the cysts, cardiac involvement usually occurs and is typically detected after the patient has reached adulthood. However, there are some reports of cardiac hydatid disease in children.¹⁵ Clinical signs and symptoms vary according to the number, size and site of the cysts. As cysts grow, they are pushed toward a weaker side of the cardiac wall, either the epicardium or the endocardium. Hydatid cysts of the LV are usually located subepicardially and rarely rupture into the pericardial space. However, when they do, rupture may be silent, or it may cause acute pericardial tamponade. The rupture of subendocardial cysts, which can also be silent, may cause anaphylactic reaction and sudden death, peripheral systemic embolism, or pulmonary embolism.^{16,17} In the RV, local intracavitary rupture is more frequent than in the LV and can cause pulmonary embolization, pulmonary hypertension and death.¹⁸ Cysts growing towards the epicardium can compress the small coronary arteries, disturbing blood flow. These signs may lead to the misdiagnosis of coronary artery disease. Our patients had nonspecific symptoms, such as dyspnea, chest pain, fever, fatigue, and palpitations. On the basis of such clinical variety, diagnosis is difficult. It is essential to consider cardiac hydatid cysts in a differential diagnosis, especially when a patient has spent time in a geographic area where hydatidosis is endemic. Electrocardiograms

and chest radiographs can show alterations that are not specific for the disease. Serologic tests are more sensitive and specific for liver hydatid disease than for cardiac hydatid disease. On the other hand, the immunoglobulin enzyme-linked immunoadsorbent assay (ELISA) was reported to be the most sensitive (94%) and specific (99%) test for most cyst locations.¹⁹ In the absence of a medical history of a visceral hydatid cyst, the diagnostic investigation of patients should involve a two-dimensional echocardiography, which if inconclusive should be followed by a cross sectional study (MDCT scan or MRI). Echocardiography is the initial investigative procedure of choice for studying cardio pericardial hydatidosis but it rarely enables direct visualization of the pulmonary embolus or the exact nature of cysts, which was the case of our patient. In our case report, pulmonary embolism of hydatid cysts was confirmed by CT scan. MDCT scan and MRI have been used successfully in the diagnosis of hydatid cysts of the lungs and the heart and with images in multiple phases, a more complete anatomical picture is achieved. However, MRI has an advantage over MDCT in examination of the heart and the great vessels, because of no radiation and even non-contrast MRI can diagnose the location and extent of hydatids.

Although mebendazole and albendazole have shown promise in the treatment of hydatid disease, cardiac hydatid disease should be treated surgically. The operative risks are minimal and the operation allows exploration of the cardiac cavities. The results of surgical treatment are excellent with complete recovery in the majority of cases.


Conclusion

Cardiac hydatidosis can be an isolated finding in absence of hydatid cysts in abdominal viscera and lungs. Cardiac CT plays instrumental role in diagnosing pericardial and cardiac masses.

Conflict of Interest: The authors declare that they have no conflict of interest.

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