

Case Report

A rare case of hydatid cyst of the interventricular septum

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ABSTRACT

A Hydatid disease or Echinococcosis is a zoonotic disease caused by the larvae (metacestode) of the cestode species of the genus *Echinococcus*. Humans are the accidental hosts of the diseases; they usually acquire it from canines; which are the definite host. It can present with systemic cyst, while cardiac manifestation of the disease is rare, due to contractile property of the heart's muscle fiber which provide resistance. In this case report, the patient is diagnosed with hydatid cyst in the inter ventricular septum; it's diagnosis and its successful treatment with surgery and albendazole. As, inter ventricular septum hydatid cyst occurs in only 0.5-2% cases, it's a unique case and its successful treatment and diagnosis can help the physicians in the future to treat a similar case as this.

Keywords: Cardiac, Cyst, Hydatid, Interventricular, Septum

INTRODUCTION

A Hydatid disease or Echinococcosis is a zoonotic disease caused by the larvae (met cestode) of the cestode species of the genus *Echinococcus* like *E. granulosus*, *E. multilocularis*, *E. vogeli* or *E. oligarthus*.¹ According to study in 2009, it is estimated that the worldwide incidence of cystic echinococcosis is about 100,000-300,000 cases annually.¹

Humans acquire primary cystic echinococcus by ingestion of *E. granulosus* eggs excreted by infected carnivores usually canines. Humans are not the definite host of the disease. There are several intermediate hosts of *Echinococcus*. They get infected from the parasite by ingesting the eggs of the parasite and in the viscera of these hosts the eggs develop into the larval stages. Humans are accidental hosts of the disease. The infection may be acquired by contact with infected definitive hosts, egg-containing feces, or egg-contaminated plants or soil followed by their ingestion.²

Human echinococcosis occurs in 4 forms- cystic echinococcosis, alveolar echinococcosis, polycystic echinococcosis and unicystic echinococcosis, but the there are two forms which are more common in humans, these are-cystic echinococcosis or hydatidosis and alveolar echinococcosis.

The disease can be prevented by programs like deworming of canines, which are the definite host of the disease.

The disease is known to occur in all continents except Antarctica and in 100 countries worldwide.³ Hydatid disease is endemic in India.⁴ For India, the highest prevalence has been reported from the erstwhile state of Andhra Pradesh, Tamil Nadu and Saurashtra region of Gujarat.⁴

This case report deals with a rare occurrence of hydatid cyst in the Interventricular Septum. Cardiac involvement in case of hydatid cyst ranges from 0.5-2%.⁵

CASE REPORT

A 36-year-old female patient presented to the OPD of the department of medicine of a tertiary care hospital of Ahmedabad with chief complaints of occasional chest pain and dyspnoea on accustomed exertion since the past 1 month.

The general physical examination was performed. She was well alert and oriented to time, place and person. Her vitals were taken which recorded a pulse rate of 96 beats per minute, in the right radial artery. She was afebrile and her Blood pressure recorded was 100/60mmHg and a saturation of 98% on room air with bilateral normal breath sounds.

Furthermore, cardiac examination was performed during which- Cardiac auscultation revealed an ejection systolic murmur in left parasternal region.

Her laboratory investigations are as follows-

- Hemoglobin (hb): 9.8mg/dl (normal range:12.0-15.5 mg/dl),
- Total leucocyte count: 15,400/mm³ (normal range-3600-11,200/mm³),
- Erythrocyte sedimentation rate (ESR): 24mm/hr (normal: <20 mm/hr),
- Platelets count- 2.4 lakh/cumm (normal range :1.7 lakh/cumm- 3.0 lakh/cumm),
- Blood Potassium - 4.5 mmol/L,
- Serum Creatinine- 0.58 mg/dl.

Radiological examination was performed -Chest X ray with Postero-anterior View (PA View) - which did not give any significant results.

Further, an Electrocardiography (ECG) investigation was performed which revealed a sinus Rhythm.

Later, 2d Echocardiography was performed for further evaluation, which revealed- Left ventricular ejection fraction - 66%, No regional wall abnormalities (RWMA) at rest, trivial Mitral Regurgitation, trivial Aortic regurgitation and trivial tricuspid regurgitation.

It also revealed an Inter-ventricular Septal Cyst with Mid Cavitory Obstruction.

Also, the Left ventricular Size appeared normal with fair Left ventricular function with reduced Left ventricular compliance.

The next investigation performed was a Multislice Computed Tomography Angiogram (MSCT) which provided the following results- Profound evidence of large, rounded, well-defined, thin-walled, minimally enhancing, cystic lesion involving inter ventricular septum- projecting towards both right and left ventricles

at mid- cavity level with external compression over ventricular cavities (left ventricle more than right ventricle)- This suggested the probable diagnosis of hydatid cyst of approximate size 48.3 x 36.5 mm in axial plane with craniocaudal extent of 44.2 mm (Figure 1).

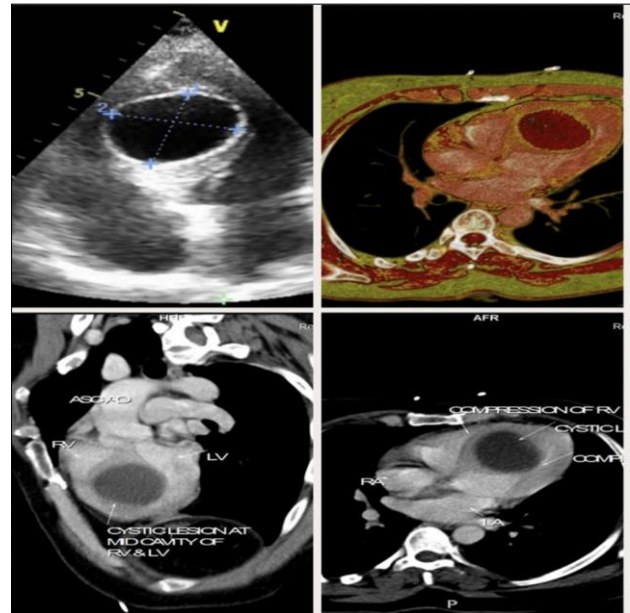


Figure 1: Cardiac multislice computed tomography (MSCT) - pre-operatively.

The patient was started on oral Albendazole and was referred to the CTVS department for surgical excision of the cyst. The cyst was excised successfully leaving behind a pseudo aneurysm.

The diagnosis of hydatid cyst disease was confirmed by histopathological examination following the operation. The patient was discharged on oral Albendazole following an uneventful postoperative stay.

After the successful surgery - 2d Echocardiography was performed which had the following results- Left Ventricular Ejection fraction - 60%, No regional wall abnormalities at rest. There was still presence of Mild Mitral Regurgitation, Trivial Aortic Regurgitation and Mild Tricuspid Regurgitation.

There was presence of a freely mobile structure attached to mid septal capsule. There was absence of any effusion.

Post-operatively, Cardiac Multislice Computed Tomography was performed which gave the following results- Positive Evidence of thickened inter ventricular septum with contrast filled out pouching within the inter ventricular septum communicating with the Left Ventricular Cavity, measuring approximately 28 x 10mm in axial plane and 30 mm in crania-caudal extension with neck measuring 8-9 mm - suggesting pseudoaneurysm formation (Figure 2).

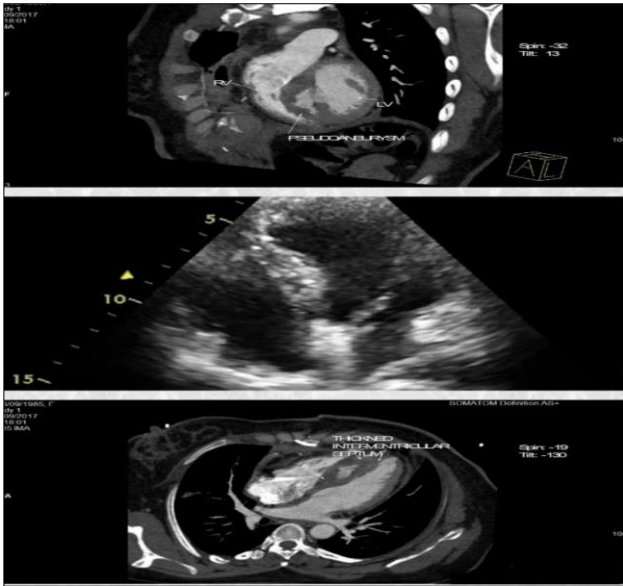


Figure 2: Multislice computed tomography (MSCT) done post-operatively.

DISCUSSION

Hydatid cyst can affect various systemic organs and therefore, it can present a variety of signs and symptoms. This was a particularly rare manifestation of hydatid cardiac disease which in itself is an uncommon entity accounting for 0.5-2% of cases of cystic Echinococcus disease.⁵ The most affected areas of cardiac involvement are the left ventricle (60-70%) and right ventricle (10%), while inter-ventricular septum, pericardium, and atria are the least affected.⁵

The clinical symptoms of cardiac hydatid cyst disease depend on various factors which include- the localization, size, age, the number of the cysts and the extent of calcification. In multiple cases of the disease, signs and symptoms may develop resulting from compression of the nearby cardiac structures such as - coronary artery, inter cardiac chamber or the conductive system of the heart. Electrocardiographic changes have been reported also, such as myocardial infarction, arrhythmias, bundle branch conduction disturbances, and sudden cardiac death.^{6,7}

Cardiac hydatid disease, though rare, can be fatal with complications like cardiac tamponade and disseminated disease. While, it's also given that the patients can remain asymptomatic during throughout; It is estimated that up to 10% of the patients remain asymptomatic for many years, although they are under the continuous threat of rupture. Mostly, the patients asymptomatic during the early states, diagnosis is most important during this stage of the disease to prevent dissemination, a careful clinical examination is crucial in early detection of the disease, Since It has been reported that up to 20% of fatal cases present with sudden death having previously shown no signs or symptoms of cardiac echinococcosis.⁷

In this case, the hydatid cyst was formed in the inter-ventricular system. According to a research study, involvement of the inter-ventricular septum occurs only in 4% of the cardiac cases.⁸ Alternatively, sometimes a cardiac hydatid cyst may simulate mitral, pulmonary, and tricuspid valvular diseases.⁹

There are various clinical manifestations that can present to the hospital, they vary according to the cyst site, size, number and are due to related complications. Symptoms are nonspecific and include atypical chest pain, breath shortness, asthenia, and palpitations.¹⁰ Chest pain may be due to compression of the small coronaries mimicking coronary artery disease and sometimes causing ischemic changes in the electrocardiogram.

Patients with a cardiac hydatid cyst usually have symptoms after its rupture. Sudden death caused by anaphylactic shock may occur after the rupture of a cyst.¹¹ The rupture may be silent, and metastatic echinococcosis of various organs may be a late and often solely due to the presence of an underlying cardiac involvement.¹² Rupture into the pericardium may result in pericardial effusion, cardiac tamponade, and, in rare cases, constrictive pericarditis.^{13,14} Rupture in the right-side chamber may lead to pulmonary emboli and rupture in the left side chamber may lead to systemic emboli; these can be grave situations for the patient.

Pulmonary embolism resulting from a hydatid cyst may cause pulmonary hypertension that is chronic, subacute, or acutely fatal.¹⁵ Several of these patients appear to follow a path of continued pulmonary hypertension with acute attacks of acute pulmonary embolism. Pulmonary emboli are caused by vesicles or daughter cysts and by scolices.¹⁶

The rupture of a left-sided hydatid cyst may lead to form a cerebral embolus. As a contrast to primary cysts of central nervous system; these cysts are found in multiple numbers. Also, secondary multiple hydatid cysts of the central nervous system caused by cardiac embolization are infertile.¹⁷

The marked incidence of catastrophic complications of cardiac hydatid cyst emphasizes the need for early diagnosis.¹⁸ currently, the following tests have lost their value due to their false positive results- Casoni's intradermal test, the Weinberg reaction, and a peripheral blood eosinophil count. A definite diagnosis based on electrocardiography and chest radiography is not possible and adequate. Since the rupture of cyst can happen during Cardiac catheterization which can lead to an anaphylactic shock, its dangers outweigh its benefits. Furthermore, angiography cannot visualize a small intramural cyst and ruptured or degenerated cysts. Two-dimensional echocardiography allows the differentiation of a cyst from a solid mass.¹⁸ In addition, computed tomography and nuclear magnetic resonance imaging may show a cyst in the heart and in other sites of the body.¹⁹

Two-dimensional echocardiography allows the differentiation of a cyst from a solid mass.²⁰ Furthermore, computed tomography can also be used, as it was used in this case.

A trans-thoracic echocardiogram can prove invaluable in the diagnosis of cystic lesions and structural anomalies after which a Trans-oesophageal echocardiography and further imaging can be done if required.

Surgical management with oral albendazole therapy remains the mainstay of treatment. The main principle of surgical treatment is to empty the cyst, remove daughter cysts and the germinative membrane, excise the pericyst, and then obliterate the residual cavity with sutures (capitonnage). The use of local scolicedal solution such as hypertonic saline solution is obligatory after cystopericystectomy in order to minimize the risk of dispersion of cystic content.²¹

CONCLUSION

As, Hydatid cyst occurring in the inter-ventricular septum is an extremely rare phenomenon, it can be easily misdiagnosed and wrongly treated, therefore; this case report is of extreme value as it has provided the method of diagnosis as well as successful treatment of the hydatid cyst in this rare case.

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