



VERY LARGE LEFT VENTRICULAR HYDATIC CYST AND RELATED SURGICAL DIFFICULTIES: CASE REPORT

MESMOUDB¹, RAOUL.J¹, BENDAGHA. N¹, MOUGHIL.S², FELLAT.R¹

¹ Cardiology A Department, Ibn Sina Teaching Hospital –Mohammed V University of Rabat.

² Cardiovascular Surgery B Department, Ibn Sina Teaching Hospital - Rabat.

Abstract: The hydatidosis is due to the accidental development of *Echinococcus granulosus* larvae in humans. It is a public health problem in endemic countries such as Maghreb countries, with a preferential localization in the liver and lungs. The cardiac localization represents 0,5 to 2 % of all the hydatid localizations and remains polymorphic and potentially fatal . We present the case of a 42 year old man, living in the rural environment, who underwent two surgical operations for left than right pulmonary hydatidosis, than was reoperated for a left pulmonary relapse. The patient was admitted with a chest pain associated with an exertional dyspnea evolving for about 6 months . The complete blood count, the serological hydatid results, and the analysis of the liquid of bronchoaspiration were negative. Nevertheless, the chest X-ray showed a deformation of the cardiac silhouette and a cardiomegaly. The thoracic angioscanner revealed a left ventricular hydatid cyst sitting at its anterior wall between the anterior interventricular artery and the circumflex artery. There was a second hydatid cyst located at the thymic lodge measuring about 10 cm of major axis. The echocardiography showed the presence of a cardiac mass in the lateral wall of the left ventricle, confirmed by the MRI, which revealed: two hydatid cysts: the first one was intramyocardial, and the second one was located at the thymic lodge. The coroscanner showed a close contact of the cyst with the anterior interventricular artery.

The patient had a pericystectomy of the hydatid cyst of the thymic lodge and resection of the anterior wall of the left ventricular cyst with a reconstruction of the left ventricular anterior wall using a double patch in Bovine pericardium and Dacron under extra-corporeal circulation. The postoperative management was simple.

Through this rare case, we remind the possibility of a cardiac localization of the hydatid cyst, and the therapeutic difficulties as it relates to its intra-myocardial development.

Keywords: Hydatid cyst, *Echinococcus granulosus*, Cardiac hydatidosis , Pericystectomy

INTRODUCTION:

The hydatidosis is a parasitic disease resulting from the accidental development of larvae of *Echinococcus granulosus* in humans. It is widespread in an endemic form in different parts of the world, including North Africa. In Morocco, it affects 5.2 persons per 100 000 inhabitants, and mainly affects women, (sex ratio M/F = 0,66) and young adults (59,1% of hydatidosis were diagnosed in patients from 15 to 49 years old) ¹.

Lung and digestive damage represent the most described lesions. The cardiac localization remains rare et represents only 0,5 to 2% of all the lesions [1]. It is a medical emergency which must be diagnosed early to avoid fatal complications.

We recall through this case the possibility of a cardiac localization of the hydatid cyst, and its therapeutic difficulties encountered in its intramyocardial development.

CASE REPORT :

We report the case of a 42 years old patient who is originating and living in an endemic area, unweaned smoker. He underwent surgery in 2002 for an hydatid cyst of the lower left lobe of the lung ruptured in the pleura for which he had benefited from a decortication , and in 2003 for right pulmonary hydatidosis for which he had a cystectomy , and reoperated in 2007 for left lung recurrence. He was cholecystectomised in 2012.

The patient was admitted for a NYHA stage II dyspnea associated to a precordial chest pain without radiation , intermittently occurring for the past three months.

The clinical examination and the lab work were normal, without hypereosinophilia, with negative hydatid serology . The electrocardiogram (ECG) showed a : normal sinus rhythm with an antero-septo-apical R-wave abrasion and a low lateral QS aspect , with negative T waves in the lateral leads, and wide T waves in the lower leads . (**Figure 1**).

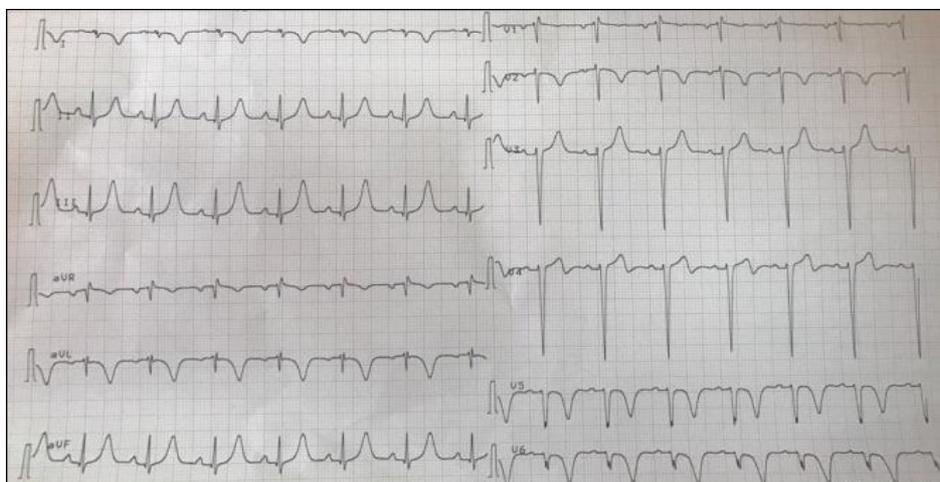


Figure 1: ECG showing a normal sinus rhythm with an antero-septo-apical R-wave abrasion and a low lateral QS aspect, with negative T waves in the lateral leads, and wide T waves in the lower leads.

The chest X-ray of the frontal chest revealed a deformation of the contours of the cardiac silhouette, with a cardiomegaly, without pulmonary parenchymal abnormalities. The echocardiography showed an undilated left ventricle, non-hypertrophied, and the presence of a cardiac mass attached to the anterolateral wall of the left ventricle, intramyocardial, probably cystic in appearance, measuring: 6.4 x 6.4 cm, with a retained left ventricular ejection fraction at 64 %. There were no other intracardiac or pericardial locations. **(Figure 2).**

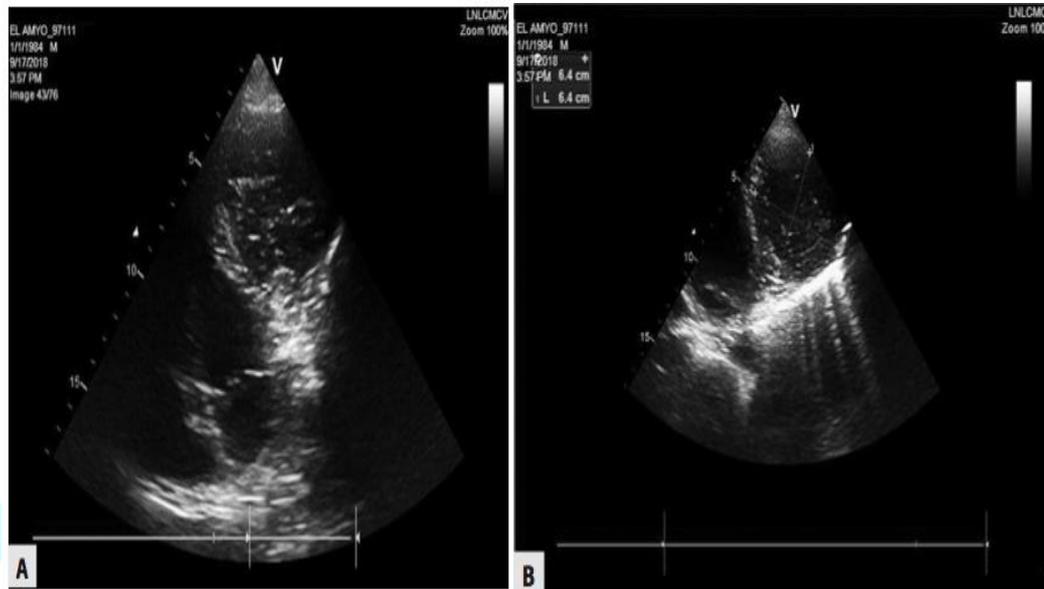


Figure 2: A-B : Echocardiography: presence of a cardiac mass attached to the anterolateral wall of the left ventricle, intramyocardial, probably cystic in appearance, measuring: 6.4 x 6.4 cm, with a retained left ventricular ejection fraction at 64 %.

The thoracic angioscanner showed a hydatid cyst of the left ventricle sitting on its anterior wall between the anterior interventricular artery and the circumflex artery with a localized thinning at the middle segment of this wall. There was a second hydatid cyst located at the level of the thymic lodge with a major axis of about 10 cm.

It was completed by a cardiac MRI which confirmed the presence of two hydatid cysts. The first located at the level of the thymic lodge of multivesicular appearance classified stage III of the Gharbi classification. The second intramyocardial measuring 9x 7 cm, developed at the expense of the anterolateral wall of the left ventricle, with a fine interface, less than 6 mm, between the endocardium and the cystic membrane at the level of the anterolateral medial segment **(Figure 3).**

The coroscanner showed: a close contact of the cyst with the anterior interventricular artery.

The radiological assessment was completed by a cerebral CT scan and an abdominal ultrasound scan, which did not find any other hydatid localizations.

The per operative diagnosis confirmed the presence of: a first multilobed hydatid cyst sitting in the thymic lodge at the left pulmonary starting point, and a second hydatid cyst of the anterior wall of the left ventricle.

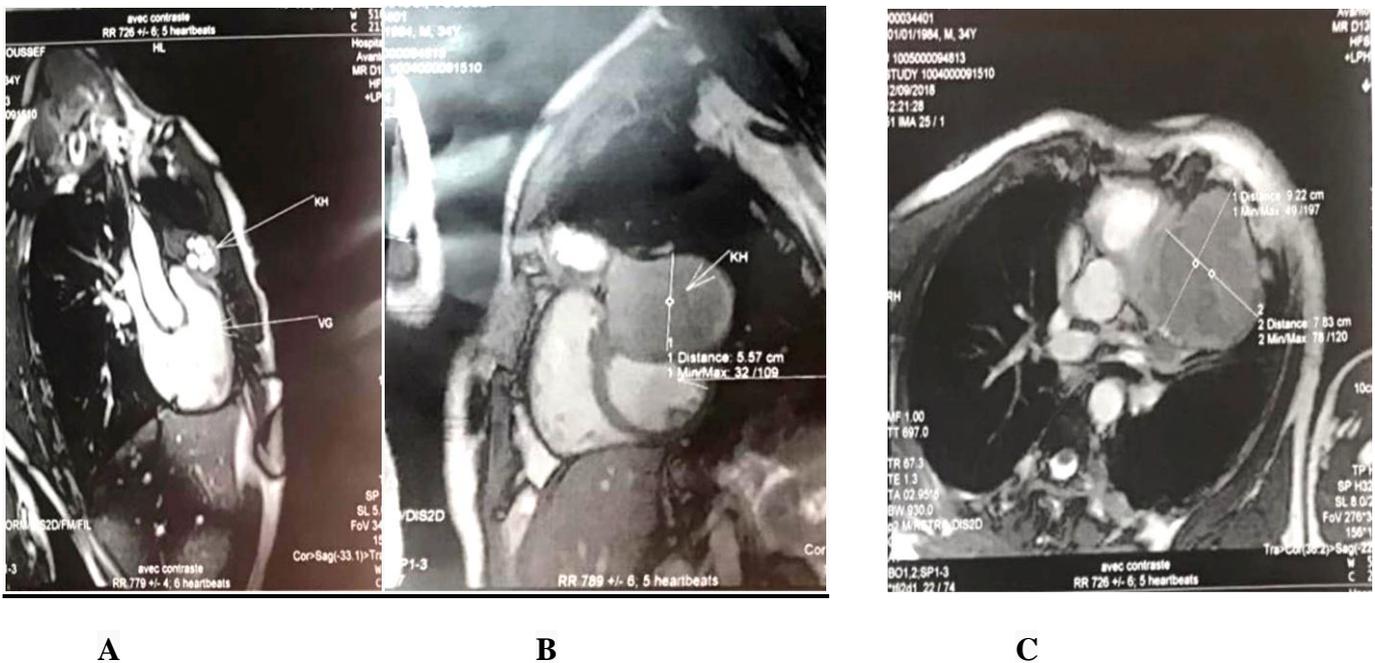


Figure 3 : A-B-C : *Intra-myocardial hydatid cyst measuring 9x 7 cm, developed at the expense of the anterolateral wall of the left ventricle.*

A pericystectomy of the hydatid cyst of the thymic lodge was performed. After installation of extracorporeal circulation, the left ventricular hydatid cyst was isolated with compresses soaked in hypertonic saline solution and then punctured. The cyst wall was incised with aspiration of its contents, which were whitish and then thick greenish in appearance, and containing membranes and vesicles.

The extraction of this material was done at the same time as a sprinkling of hypertonic saline. After emptying and sterilizing the residual cavity, which was about 20 cm long, its walls were completely resected. The residual anterior wall of the left ventricle was indeed thinning with a zone of fragility in its middle segment as demonstrated on the thoracic angioscanner. This zone was reinforced by a double bovine and Dacron pericardial patch, sutured with a series of 3/0 prolene sutures passed in a U-shape on the external wall of the left ventricle, respecting the anterior interventricular artery which is very close. The hemostasis was completed by the biological glue around the edge of the patch. The extracorporeal circulation was stopped and the walls were closed plan by plan. The postoperative management was simple.

DISCUSSION :

Mediastinal hydatidosis accounts for 0.1-0.5% of all hydatid locations.² It is associated with cardiac damage in 33% of cases.³ Left ventricular involvement is elevated relative to other cardiac structures. It accounts for 55-60% of cardiac involvement because of its blood flow and large mass, whereas the right ventricle accounts for only 15% of cases.

The interventricular septum is involved in 5-9% of cases, and the right auricle in 3-4% of cases⁴. The distribution in the pulmonary artery, left atrium, and pericardium is about 7-8%⁵. This distribution is parallel to the extent of coronary blood flow. Cardiac involvement is associated in 20-40% of cases with other visceral locations.

Human contamination can occur by ingesting food contaminated with the eggs of the parasite or by direct contact with an infested animal. Ingested eggs pass through the intestines and reach the liver through the portal circulation, which is why the liver is the most affected organ by hydatidosis. The passage of the larvae through the liver can reach the right auricle and the lung through the venous circulation. Thus, the lung is the second most common location.⁶

There is no consensus regarding the dissemination of hydatid cyst in the heart. Indeed, it is assumed that hydatid cysts reach the left heart via the coronary circulation. However, direct contamination from adjacent tissues is also possible, especially from pulmonary veins, lymphatics, thoracic duct, vena cava or portal circulation⁷.

According to the results of the monocentric study of Alptekin Yasim which includes 25 cases, hydatid cysts that pass through the liver reach the right heart through the venous system and thus affect the right auricle, the right ventricle, and the inter-atrial septum. Cysts that pass through these areas may also affect the lungs.

On the other hand, cysts that pass into the lungs can affect the left heart through the pulmonary veins and the systemic circulation which can move the larvae into the coronary circulation and into the left ventricular subepicardium and interventricular septum. This mode of dissemination could explain why extra-cavitary development of hydatidosis is more frequently found in the left ventricle.

In the literature, the most affected site remains the left ventricle because it has the largest coronary blood flow and mass. It has been reported in various studies that hydatid cysts of the right heart tend to develop intracavitary and subendocardial, while hydatid cysts of the left heart develop extracavitary and subepicardial⁸⁻⁹. This is explained by the cardiac pathway of the hydatid cyst. Indeed, cysts that pass through the liver invade the low-pressure areas of the right heart and have intraluminal development, and often cause signs of obstruction.

They can also invade the cardiac conduction system and cause arrhythmias. Cysts that pass through the left heart pass through the lungs and reach the myocardium through the coronary circulation. Thus, the high pressures of the left ventricle explain the growth of the cyst through the pericardium, which explains the chest pain and signs of ischemia caused by the compression of the coronary vessels. Rupture of subepicardial cysts is rare and often silent. Sometimes, it can cause tamponade, pericardial hydatidosis or constrictive pericarditis.

Subendocardial rupture, on the other hand, can cause many symptoms such as anaphylactic shock, sudden death, peripheral systemic embolism, or pulmonary embolism⁷. Thus, the clinical manifestations of hydatid cysts are variable, often latent and misleading, depending on the number, location, or size of the cysts.

Our patient had a hydatid cyst of the left ventricle with a myocardial extension, causing a precordial chest pain and an exertional dyspnea. According to the literature, chest pain and dyspnea are the most frequent manifestations of cardiac hydatidosis. The notion of exposure to dogs and sheep, endemic background or personal history of other hydatid locations should suggest the diagnosis.

The diagnosis of cardiac hydatid cyst can be evoked on a chest X-ray, which may be normal in the case of a small or intracavitary hydatid cyst, or show either a localized arch or an extensive deformation of a cardiac edge, or a bulge in the left middle arch, or a calcified mass or fine calcifications projecting from the cardiac shadow, or a cardiomegaly.

There is no specific biological test for cardiac hydatid cysts. Hypereosinophilia is not an absolute sign, it may increase or remain normal as it is in our patient. Hypergammaglobulinemia is found in 30% of hydatidose cases. Highly sensitive immunological tests: IgG-ELISA- the latex agglutination test, indirect immunofluorescence, can identify circulating antibodies but there are some false negatives. Immune electrophoresis is the most specific test⁵. Hydatid serology is positive in only half of the cases of cardiac hydatid cysts. Our patient had a negative hydatid serology.

Imaging has an important role in the diagnosis and injury assessment of cardiac hydatid cysts. Indeed, the echocardiography is the reference examination and represents a simple means for the diagnosis of cardiac hydatid cyst which presents as a transonic mass frequently containing daughter vesicles or trabeculations. More rarely, a ruptured cyst presents as a solid mass, which raises the problem of differential diagnosis with other cardiac tumours.

Cardiac ultrasound can also be used to determine the number, size and location of the cysts. Transoesophageal ultrasound has a significant contribution in intra-auricular localizations.

Multi-slice volume CT allows a very precise locoregional morphological assessment during the same session and in a very short time, in particular a very precise mediastinal morphological assessment. A study of the relationship of the cyst with the cardiac cavities and with the coronary vessels in the different planes of space and an assessment of the spread of the disease by thoraco-abdomino-pelvic acquisition. It allows better counting in case of multiple hydatidosis.

Cardiac-synchronized MRI is more accurate in topographic analysis and in studying the relationship of cysts to the heart muscle and surrounding tissues, without irradiation or contrast injection.¹⁰

Because of the severity of the spontaneous evolution, including the risk of compression, rupture and sudden death¹¹ surgical treatment of cardiac hydatid cysts is required whenever possible, even when the patient is asymptomatic.

In fact, surgical excision remains the only treatment that allows a complete healing. Intraoperative surface echocardiography provides an important support to the surgeon because it determines the surgical approach area of the cyst. Indeed, the thoracic approach depends mainly on the location of the cyst. Sternotomy is preferable as it allows all locations to be approached, but an anterolateral submammary thoracotomy may be an aesthetic alternative. Extracorporeal circulation (ECC) is very useful for a complete myocardial assessment, which is the only way to guarantee a definitive procedure¹¹. The first step consists in protecting the operating field with compresses soaked in hypertonic saline solution using Povidone-iodine sometimes. The cyst is punctured after being sterilised by injection of the same solution to avoid any risk of dissemination. Pericystectomy and cystectomy with treatment of the residual cavity are the two surgical techniques described.¹²⁻¹³

Pericystectomy is a dilapidating procedure with risks of damage of the surrounding structures and bleeding of the residual cavity, which is more suitable for subendocardial cysts with intracardial development. For subepicardial cysts with extracardial development, cystectomy is preferable by resection of the protruding dome and removal of the germinal membrane and daughter vesicles.

Due to its volume, the residual cavity may be left open in some patients, as several authors believe that the cushioning of the residual cavity may interfere with contractility and segmental myocardial relaxation, causing tears with tension sutures. The post-operative results of the surgery are encouraging. ¹ Post-operative treatment with Albendazole for 3 to 6 months is recommended to avoid the risk of local recurrence.⁷

The option of isolated medical treatment has been proposed as an alternative to surgery in some cases such as calcified or small cysts or in the elderly or in cases where surgery is declined.

A hydatid serology every two months for two years is justified to detect a possible recurrence.

CONCLUSION:

Cardiac localization of hydatid cyst remains rare, even in endemic countries. Early diagnosis must be made by echocardiography, CT scan or cardiac MRI to avoid fatal complications. First-line treatment is surgery, which, despite its difficulties, remains the first-line treatment. Treatment with Albendazole is prescribed after surgery to prevent recurrences.

REFERENCES :

- [1] Derfoufi O, Akwa EN, Elmaataoui A, Miss E, Esselmani H, Lyagoubi M, et al. Profil épidémiologique de l'hydatidose au Maroc de 1980 à 2008. *Ann Biol Clin* 2012;70(4):457-61.
- [2] N. Zaghba, H. Benjelloun, N. Yassine, Z. Rochaidi, A. Bakhtar, A. Bahlaoui. Hydatidose médiastinale : A propos de 5 cas. *Revue des Maladies Respiratoires*, Vol 26, N° HS1 - janvier 2009 p. 77
- [3] Charon P, Hamwi A, Beroud P, Dauplain J, Lefort JF, Cazal J. KH médiastinal, intérêt des méthodes actuelles d'imagerie médicale. *Sem Hôp Paris* 1993;69:100-3.
- [4] Maroto LC, Carrascal Y, López MJ, Forteza A, Pérez A & Zavanella C 1998 Hydatid cyst of the interventricular septum in a 3.5-year-old child. *Annals of Thoracic Surgery* 66 2110-2111. (doi:10.1016/S0003-4975(98)01079-0)
- [5] Shevchenko YL, Travin NO, Musaev GH & Morozov AV 2006 Heart echinococcosis: current problems and surgical treatment. *Multimedia Manual of Cardiothoracic Surgery* 2006 810. (doi:10.1510/mmcts.2005.001115)
- [6] WHO Informal Working Group on Echinococcosis. Guidelines for treatment of cystic and alveolar echinococcosis in humans. *Bull World Health Organ* 1996;74:231-42.
- [7] Alptekin Yasim, MD*, Hasim Ustunsoy, MD, Gokhan Gokaslan, MD, Erhan Hafiz, MD, Yavuz Arslanoglu, MD. Cardiac Echinococcosis: A Single-Centre Study with 25 Patients. *Heart, Lung and Circulation* (2016) xx, 1-7
- [8] Akar R, Eryilmaz S, Yazicioglu L, Eren NT, Durdu S, Uysale A, et al. Surgery for cardiac hydatid disease: an Anatolian experience. *Anadolu Kardiyol Derg* 2003;3:238-44.
- [9] Parvizi R, Joudati AR, Montazeri V, Salmasi SH, Varshouchi M. Cardiac echinococcosis: surgical treatment and results in 10 cases. *Iranian Heart Journal* 2006;7:67-71.
- [10] Kyste hydatique du cœur : A propos d'une observation : cardiologie-pratique, 2010-2018.
- [11] A. Bakkalia, I. Jaabaria, H. Bouhdadia, R. Razineb, N. Bennani, Mechtab J, El arraga A, Belkhadira C, Benlafqiha M, Laaroussia. Les kystes hydatiques cardiaques : A propos de 17 cas opérés. *Annales de Cardiologie et d'Angéiologie* Volume 67, Issue 2, April 2018, Pages 67-73
- [12] Kaplan M, Demitras M, Cimen S, et al. Cardiac hydatid cysts with intracavity expansion. *Ann Thorac Surg* 2001;71:1587-90.
- [13] Shevchenko YL, Travin NO, Musaev GH, Morozov AV. Heart echinococcosis: current problems and surgical treatment. *Multimedia Man Cardiothorac Surg* 2005;1115.