

## Hydatidosis Presenting as Chronic Constrictive Pericarditis: A Case Report

**Hicham Wazaren<sup>1\*</sup>, Hanae Bouhdadi<sup>1</sup>, Sabrina Derqaoui<sup>2</sup>, Jaafar Rhissassi<sup>1</sup>, Rochde Sayah<sup>1</sup>, Chakib Benlafqih<sup>1</sup> and Mohammed Laaroussi<sup>1</sup>**

<sup>1</sup>Department of Cardiovascular Surgery A of Ibn Sina University Hospital Center, Mohammed V University of Rabat, Morocco

<sup>2</sup>Department of Pathology of Ibn Sina University Hospital Center, Mohammed V University of Rabat, Morocco

**\*Corresponding Author:** Hicham Wazaren, Department of Cardiovascular Surgery A of Ibn Sina University Hospital Center, Mohammed V University of Rabat, Morocco.

**Received:** January 25, 2021; **Published:** April 26, 2021

### Abstract

Pericardial hydatidosis is rare. The transformation towards constriction of hydatid pericarditis remains exceptionally a mode of revelation of the disease.

This is a 49-year-old male, suffering exercise dyspnea with ascites. The echocardiography made objectified a thickening of the pericardium with appearance of shell obstructing the filling of the left ventricle and the right ventricle without calcifications. The cardiac magnetic resonance highlighted an aspect evoking a bulky pericardial effusion encysted compressive with presence of signs of chronic constrictive pericarditis. The patient underwent a sternotomy by which a subtotal pericardiectomy was performed. The pathology study showed signs suggestive of hydatid cysts. Hydatid serology was positive.

In most chronic constrictive pericarditis, the real treatment is surgical pericardial decortication.

**Keywords:** Constrictive Pericarditis; Hydatid Cysts; Sternotomy; Decortications; Right Catheterism

### Abbreviations

NYHA: New York Heart Association; CRP: C Reactive Protein; Gamma-GT: Gamma Glutamyl Transferase; ELISA: Enzyme Linked Immunosorbent Assay

### Background

Chronic constrictive pericarditis is defined as fibrosis of the pericardial sac, causing discomfort in the diastolic filling. It is a rare condition whose diagnosis is often difficult to make. It is based on a bundle of clinical, echocardiographic, hemodynamic and radiological arguments. Among the main etiologies, we note postoperative and postradical pericarditis, tuberculosis in our context and drug causes [1].

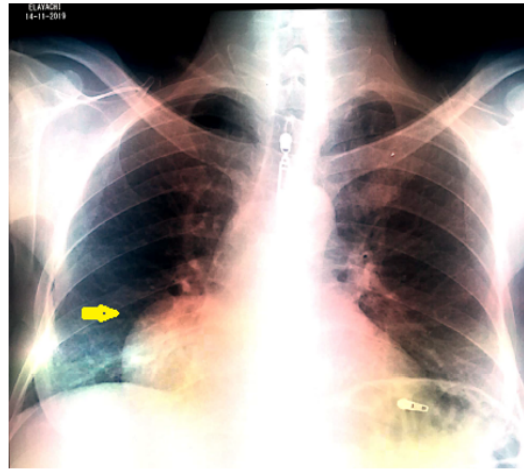
We report a case of chronic constrictive pericarditis attributable to pericardial hydatidosis etiology exceptionally described.

### Case Presentation

We report the case of a 49-year-old patient with no significant medical history. The clinical history began a year before his admission to our training with dyspnea deemed stage II of the NYHA in an acute context. The patient consulted privately where the diagnosis of

acute pericardial effusion was made. The patient underwent an evacuating pericardial puncture under the xiphoid route. The etiological investigations returned negative.

For the past few months, the patient has reported a slow progression of exercise dyspnea occurring mostly at the start of exercise. More recently, a discreet edema of the lower limbs and ascites of moderate abundance have appeared. The chest X-ray performed showed a right para-hilar opacity, suggesting a mass opposite the right atrium (Figure 1).



**Figure 1:** Chest x-ray showing the presence of suspicious right para-hilar opacity.

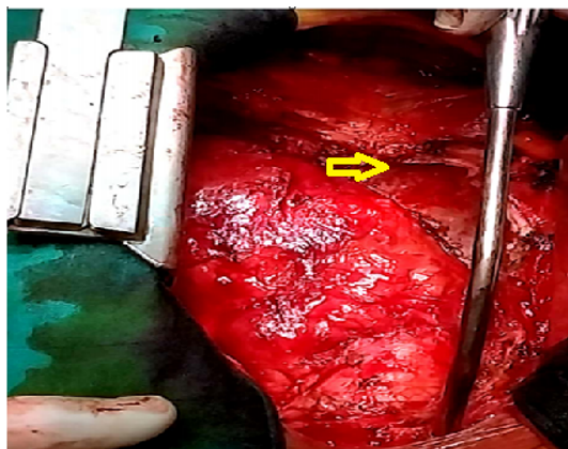
The echocardiography made objectified a thickening of the pericardium with appearance of shell obstructing the filling of the left ventricle and the right ventricle without calcifications. The left ventricular function remains satisfactory with an Ejection Fraction at 58%. The right ventricular function is faulty with an S wave at 0.08 m/s.

The complement of the imagery by a cardiac magnetic resonance highlighted an aspect evoking a bulky pericardial effusion encysted, measuring 137 \* 65 mm lodged between the right cavities and the right costal edge, compressive with presence of signs of chronic constrictive pericarditis (infero and left latero-ventricular pericardial thickening, pathological ventricular coupling, septal bounce, very dilated inferior vena cava).

The biological assessment noted a CRP at 15 mg/l, gamma-GT at 150 IU/l, normal transaminases. The search for Koch's bacillus in sputum was negative.

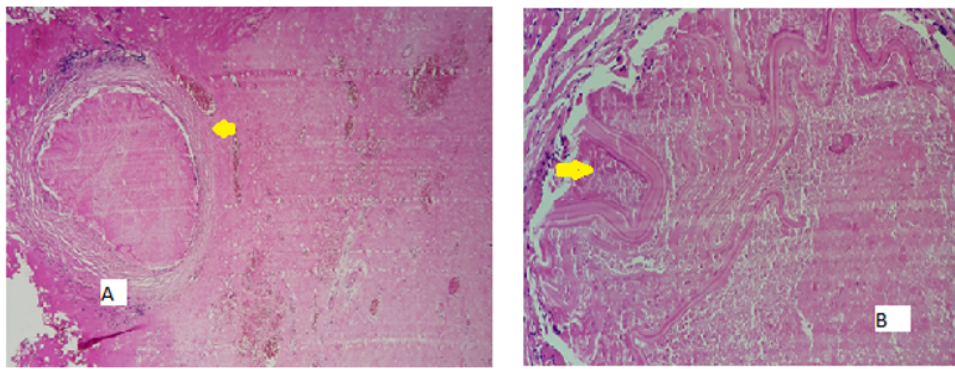
The patient underwent a sternotomy by which a subtotal pericardiectomy was performed, involving the entire anterior surface of the heart going from the phrenic nerve to the other, with extensive decortication above the base vessels and below towards the diaphragmatic side and the inferior vena cava. The cystic mass opposite the right cavities was opened with aspiration of yellowish liquid which was sent for a bacteriological study. A pericardial biopsy was also performed (Figure 2). The use of extracorporeal circulation was not necessary.

The pathology study showed that on gross, removed cardiac fragment was ranged from 1 cm to 3 cm, presenting as grey-white and homogenous masses. Hematoxylin-eosin stained sections revealed a totally fibrotic, thickened pericardic wall with rare calcifications.



**Figure 2:** Operative view showing the pericardiectomy, the presence of a cystic mass next to the right atrium (yellow arrow).

These fibrotic changes were associated to a sparse inflammatory infiltrate, consisting of lymphocytes, plasmocytes and a granulomatous foreign body-type giant cell reaction (Figure 3A and 3B).

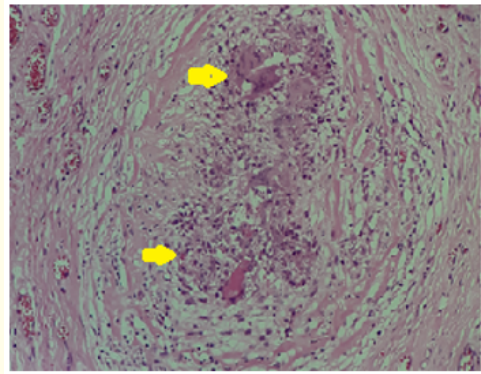


**Figure 3A and 3B:** Hydatid cysts: lamellated layer of cyst wall (A: HE x10; B: HE x 20).

The presence of avascular, eosinophilic, laminated membranes within the pericardic wall was highly suggestive of hydatid cysts (Figure 4). However, scolices were absent.

Hydatid serology was positive at 34 (IgG ELISA) with an Indirect Hemagglutination test at 1/640 in favor of active hydatidosis.

The diagnosis was in favor of chronic constrictive pericarditis linked to pericardial hydatidosis on clinical, echocardiographic and magnetic resonance data. The etiology objectified by the anatomopathological data. The patient was started on albendazole 15 mg/kg/day. The evolution was favorable.



**Figure 4:** Cyst wall showing foreign body type giant cell reaction (HE x 20).

## Discussion

The term pericarditis includes all of the terms of inflammatory conditions of the serous envelope of the heart that constitutes the pericardium.

Pericarditis is an inflammation of the two layers (parietal and visceral) of the pericardial serosa. This inflammatory reaction is mediated by cytokines (such as interleukins or tumor necrosis factor) [2].

Harmful agents of a very diverse nature can, when they affect the pericardium, be at the origin of an inflammatory reaction of this serosa. This aggression of the pericardium can result from infectious agents (viruses, bacteria, parasites, yeasts) which can irritate the pericardium, either directly, or through the immune reaction which they trigger (for example rheumatic fever) [3].

Inflammation of the pericardium progresses to a dense, inextensible fibrosis, with adhesions, and secondary calcifications which begin in the atrioventricular, interventricular furrows and on the lower and anterior surface of the heart leading to the formation of an inextensible sac, annoying the diastolic filling [4].

Clinical examination shows permanent dilation of the jugular veins increasing on inspiration (Kussmaul's sign), diastolic vibrance (rare) and hepatomegaly possibly associated with signs of liver dysfunction (such as jaundice and ascites).

Patients seem less symptomatic than before. In the Ling., *et al.* series evaluated between 1936 and 1982, 69% of patients were in stage III and IV [5]. One explanation probably lies in an earlier diagnosis as well as a decrease in the tuberculous etiology deemed to be more symptomatic due to the more intense pericardial fibrosis. However, chronic pericarditis is not a silent pathology because 82% of patients are symptomatic (at least NYHA class II) [6].

The electrocardiogram usually shows sinus tachycardia or atrial fibrillation related to prolonged elevation of atrial pressures and enlargement of the atrium. T waves are generally flattened or even inverted in a diffuse manner, atrioventricular and intraventricular conduction disorders and pseudo-signs of infarction (deep Q waves) [7,8].

In the chest x-ray, conventionally, the heart volume was considered small and the heart failure with small heart made evoke, of prejudice, a pericardial constriction. The widening of the cardiac silhouette can result from the thickening of the pericardium and especially

from an associated effusion. The chest x-ray can immediately guide the diagnosis if there are pericardial calcifications [8].

It is the cardiac ultrasound which often allows the diagnosis by showing a thickened, hyperechoic and immobile pericardium, an unexpanded left ventricle, a dilation of the left atrium, a dilation of the inferior vena cava without decrease or inspiratory collapse and abnormal posterior displacement of the septum in protodiastole [9,10].

Magnetic resonance imaging is a morphofunctional examination which, in summary, shows, in the case of chronic constrictive pericarditis, thickening of the pericardium, and dynamic anomalies related to diastolic dysfunction. Performing the differential diagnosis between constriction and restriction is a priority objective of the examination [11].

The reference hemodynamic examination remains right catheterism [12] which highlights a “dip and plateau” type curve.

The “and” is essential because it signs the manometric chronology of the two events.

It's about:

- A protodiastolic “dip” (decrease in pressure, literally a pressure drop);
- monitoring (“and”) of a sudden increase in pressure (from the start of filling) and of too rapid reaching (in a “plateau”) of the maximum right pressures.

From the beginning of the century until the 1950s, tuberculosis was the main cause of pericardial constriction in Western countries and remained current in developing countries. Recent series from Saudi Arabia, Mexico, Turkey and India find a percentage of constrictive tuberculous pericarditis from 38 to 83% [13].

Currently, in developed countries, idiopathic constrictive pericarditis is the most common. Etiologies which were rare formerly are seen more and more; these are chronic post-surgical pericarditis, post-radiotherapy, in the context of cancer and post-viral pathology. Postoperative constrictive pericarditis appeared in the 1980s, its incidence is variable estimated between 11 and 29%. Chronic pericarditis post radiotherapy can be seen in 30% of certain series.

The other etiologies are rarer: end-stage renal failure (dialysis or not), connective tissue diseases (e.g. rheumatoid arthritis), system diseases (ex: systemic lupus erythematosus), purulent pericarditis (*Staphylococcus*, *Streptococcus*...). Rarely, constrictive pericarditis can also develop after pericarditis as a result of myocardial infarction (Dressler syndrome). And exceptionally, fungal or parasitic pericarditis (histoplasmosis, coccidiomycosis, *Candida albicans*, *Candida tropicalis*, *Echinococcus*...), in particular for groups at risk (drug addicts, immunosuppressed, and broad spectrum antibiotic therapy) [14].

Pericardial hydatidosis is a rare disease; it represents 0.2 to 2% of cases of hydatidosis [15,16]. Cardiac involvement is isolated in 1/3 of the cases; in 2/3 of the cases it is associated with pulmonary involvement [17]. The *Echinococcus granulosus* larva arrives in the left heart chambers after escaping the hepatic filter, reaching the right atrium and from there the left heart through the pulmonary circulation, or even a permeable oval foramen [18].

From the left ventricle, the larvae are expelled into the large circulation and through the coronary arteries; the parasite invades the myocardium [19].

All localizations of the hydatid cyst of the heart are possible. The parasite sits by frequency in the left ventricle in 50 to 60% of the cases, the interventricular septum in 10 - 20% of the cases, the right ventricle in 5 - 15% of the cases; the cardio-pericardial localization constitutes 10 - 15% of the case.

Pericardial localization without cardiac involvement is therefore extremely rare.

The transformation towards constriction of hydatid pericarditis remains exceptionally a mode of revelation of the disease. Thus, our case illustrates a pericardial constriction revealing a pericardial hydatidosis.

The diagnosis of hydatid cysts is confirmed by histology [20]. Chronic constrictive pericarditis commonly presents pathologic changes involving the parietal pericardium, but the visceral pericardium and even the underlying epicardium may also be affected [21]. Typical histological findings include a thick, fibrotic and calcified pericardium. However, the thickness of surgically removed pericardium was reported to be normal in 18% of the cases. Histologically, Hydatid cysts encompass three layers. The inner germinal layer is thin and translucent; scolices develop from an out pouching of this layer. The middle laminated membrane is an avascular, eosinophilic, strongly PAS+, refractile and chitinous membrane. The exterior layer or pericyst is a firm thick protective layer; it consists of fibrovascular tissue with chronic inflammatory cells and variable calcification. The cyst is surrounded by inflammatory fibrous tissue with often foreign body type giant cell reaction [22]. The most significant factor in the diagnosis of hydatid pericarditis is the identification of the organism and/or its various parts. The identification of hyalinized and fibrotic tissue host reaction, can be a highly suggestive of hydatid cysts, although not being diagnostic themselves [23]. In these cases, serology may be of use for the diagnosis confirmation. In our patient, histological findings were not specific, as scolices were absent, but were highly suggestive of hydatid pericarditis. Thus, the diagnosis was confirmed by serology.

In most chronic constrictive pericarditis, the real treatment is surgical pericardial decortication, consisting of a broad resection of the parietal and visceral sheet of the pericardium [24]. The operational risk is significant (> 6% in the most experienced centers) [25]. In some patients, restoration of heart function is not immediate, and may take some time (a few months) to return to normal. Survival is strongly linked to etiology. In the Mayo Clinic's largest series, survival is the best for chronic idiopathic or viral pericarditis and the least good for post-radiotherapy.

Medical treatment is only palliative, allowing the patient to be prepared for the intervention, or reserved for cases where it is rejected because of an absolute contraindication to surgery: significant hepatocellular insufficiency, severe associated myocardial damage.

When the chronic pericarditis is due to pericardial hydatidosis, surgical excision should be performed in the presence of a cyst. Medical treatment represents the treatment of choice for non-operable patients and a complementary treatment to a surgical intervention when there is a risk of dissemination. The most widely used product is Albendazole® at a dose of 10 to 15 mg/kg per day for six months.

The patient's postoperative state, and in particular the level of cardiac output, depends on the extent of the preoperative effusions. If necessary, pharmacological support and/or the placement of an intra-aortic balloon pump can be applied.

The causes of hospital mortality are, above all, in 75% of cases, linked to acute heart failure. Then, mainly, the hemorrhage and respiratory failure. Intraoperative mortality is considered low; less than 1% in the Ling, *et al.* In a recent publication review including 429 patients, hospital mortality was on average 5% ranging from 2.3 to 10.5% [26]. Operational mortality and episodes of low cardiac output are correlated, in McCaughan's study, with the preoperative functional stage [27].

### Conclusion

Chronic constrictive pericarditis is a rare and serious pathology. The diagnosis, which is sometimes difficult, is based on the association of clinical signs of water-sodium retention, specific signs of diastolia and increased ventricular interdependence on ultrasound, and finally an anatomical and functional analysis of the pericardium and the heart by magnetic resonance.



The hydatid origin of constrictive pericarditis is exceptional. The latter is rarely described as the ultimate evolution of pericardial hydatidosis.

### Consent for Publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

### Availability of Data and Materials

Data sharing is not applicable to this article as no datasets were generated or analyzed during the current study.

### Competing Interests

Authors declare they have no competing interests.

### Authors' Contributions

All authors have read and approved the manuscript.

HW, CB: Main authors managed the patient. SD, HB: Co-author analyzed the patient data and was a major contributor in writing the manuscript. JR, RS, and ML: Supervised the management of the patient, and revised the manuscript.

### Bibliography

1. Throughton R., *et al.* "Pericarditis". *Lancet* 363.9410 (2004): 717-727.
2. Lorell BH and Braunwald E. "Pericardial disease". In: Braunwald E editor. *Heart disease*. Philadelphia: WB Saunders (1992): 1465-516.
3. Fowler NO and Harbin AD. "Recurrent acute pericarditis: follow-up study of 31 patients". *Journal of the American College of Cardiology* 7.2 (1986): 300-305.
4. Millaire A., *et al.* "Treatment of recurrent pericarditis with colchicine". *European Heart Journal* 15 (1994): 120-124.
5. Ling LH., *et al.* "Constrictive pericarditis in the modern era: evolving clinical spectrum and impact on outcome after pericardectomy". *Circulation* 100.13 (1999):1380-1386.
6. Afzal A., *et al.* "Myocarditis and pericarditis with tamponade associated with disseminated tuberculosis". *The Canadian Journal of Cardiology* 16.4 (2000): 519-521.
7. Nataf P., *et al.* "Result of subtotal pericardectomy for constrictive pericarditis". *European Journal of Cardio-Thoracic Surgery* 7.5 (1993): 252-255.
8. Schiavone WA. "The changing etiology of constrictive pericarditis in a large referral center". *The American Journal of Cardiology* (1986): 58373-58375.
9. OH JK., *et al.* "Preload reduction to unmask the characteristic Doppler features of constrictive pericarditis. A new observation". *Circulation* 95.4 (1997): 796-799.
10. OH JK., *et al.* "Diagnostic role of Doppler echocardiography in constrictive pericarditis". *Journal of the American College of Cardiology* 23.1 (1994): 154-162.

11. Kojima S., *et al.* "Diagnosis of constrictive pericarditis by tagged cine magnetic resonance imaging". *The New England Journal of Medicine* 341.5 (1999): 373-374.
12. Nishimura RA. "Constrictive pericarditis in the modern era: a diagnostic dilemma". *Heart* 86.6 (2001): 619-623.
13. S Arsan., *et al.* "Long term experience with pericardectomy; analysis of 105 consecutive patients". *The Journal of Thoracic and Cardiovascular Surgery* 42.6 (1994): 340-344.
14. Robert BH., *et al.* "Constrictive pericarditis: clinical and pathophysiologic characteristics". *American Heart Journal* 138.2 (1999): 219-232.
15. Chellaoui M., *et al.* "Hydatidose péricardique: à propos de 3 observations". *World Journal of Radiology* 84.3 (2003): 329-323.
16. Elkarimi S., *et al.* "Tamponnade révélant un kyste hydatique intra-péricardique - un cas". *Annales de Cardiologie et d'angiologie* 63.4 (2014): 267-270.
17. Orhan G., *et al.* "Chirurgie des kystes hydatiques cardiaques: Trente-neuf ans d'expérience". *Annales de Cardiologie et d'Angéiologie* 57.1 (2008): 58-61.
18. Bréchnignac X., *et al.* "Kyste hydatique du Coeur". *La Presse Médicale* 26.14 (1997): 663-665.
19. Rosenberg T., *et al.* "Acute abdominal aorta embolism caused by primary cardiac echinococcus cyst". *European Journal of Vascular and Endovascular Surgery* 7.5 (1993): 582.
20. Sultana N., *et al.* "Primary cervical hydatid cyst: a rare occurrence". *Diagnostic Pathology* 7 (2012):157.
21. Welch TD and OH JK. "Constrictive Pericarditis". *Cardiology Clinics* 35.4 (2017): 539-549.
22. Derqaoui S., *et al.* "Primary Hydatid Cyst of Soft Tissues: Description of 6 Cases". *Journal of Cytology and Histology* 10 (2019): 545.
23. Taxy JB., *et al.* "Echinococcosis". *The American Journal of Surgical Pathology* 41.1 (2017): 94-100.
24. Ling LH., *et al.* "Constrictive pericarditis in the modern era: evolving clinical spectrum and impact on outcome after pericardectomy". *Circulation* 100.13 (1999):1380-1386.
25. Bertog SC., *et al.* "Constrictive pericarditis: etiology and cause-specific survival after pericardectomy". *Journal of the American College of Cardiology* 43.8 (2004): 1445-1452.
26. Ujjwal K., *et al.* "Pericardectomy for Constrictive Pericarditis: A Clinical, Echocardiographic, and Hemodynamic Evaluation of Two Surgical Techniques Cardiothoracic Centre, All India Institute of Medical Sciences, New Delhi, India 81.2 (2006): 522-529.
27. McCaughan BC., *et al.* "Early and late results of pericardectomy for constrictive pericarditis". *The Journal of Thoracic and Cardiovascular Surgery* 89.3 (1985): 340-350.

**Volume 4 Issue 5 May 2021**

**© All rights reserved by Hicham Wazaren., et al.**