

## Case Report

# Tuberculous Calcific Constrictive Pericarditis in a child of 13 years

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### ABSTRACT:

#### BACKGROUND:

We are presenting a rare case of a child with tuberculous calcific constrictive pericarditis

#### METHODS:

Investigations revealed presence of the disease and surgery was performed as pericardiectomy.

#### RESULTS:

Recovery after surgery was remarkable and the symptoms ameliorated quickly.

#### CONCLUSION:

A presentation of a very rare condition of constrictive pericarditis in a child without involvement of the lungs and other organs by the disease

**KEYWORDS:** Constrictive pericarditis, pericardiectomy, tuberculous pericarditis

#### INTRODUCTION:

Constrictive Pericarditis (CP), is a chronic inflammatory process that involves both fibrous and serous layers of the pericardium and that leads to pericardial thickening and compression (constriction) of the ventricle and the severity varies according to the pathologic process. The resultant impairment in diastolic filling reduces cardiac function<sup>(1,2)</sup>.

*Historical notes:* Galen in AD 160 described cicatricial thickening of the pericardium in an animal and surmised that the same condition might occur in humans<sup>(1)</sup>. The first formal account of the condition in humans was apparently that of *Lower* who described both acute and chronic CP in 1669. Rehn and Sauerbruck in Germany carried out a successful partial pericardiectomy in 1913<sup>(1)</sup>.

Normally the pericardial pressure is sub atmospheric, similar to the intrapleural pressure, both become more negative during inspiration, but in CP there is an increase in right and left

ventricular diastolic pressures and a decrease in the stroke volume, which is due to obliteration of pericardial space and so increasing the pericardial pressure<sup>(1,2)</sup>. Eventually and in many cases cardiac muscle atrophy may occur or it may appear early in the course of the disease, and in late stages myocardial fibrosis or right ventricular dysplasia may result<sup>(1,2,3)</sup>.

Pericarditis may occur as a primary or as a secondary manifestation of a systemic disease<sup>(4)</sup>. But in general etiology of CP is unknown<sup>(1,2)</sup>.

In 10% of cases, acute pericarditis precedes the development of chronic CP, and less than 3% of patients have evidence of tuberculous pericarditis. Mediastinal radiation has become a prominent cause of CP due to the increase intensity of this treatment<sup>(1)</sup>. Rheumatoid disease, sarcoidosis, trauma or even a cat – scratch are reported as causes of chronic CP<sup>(1,2,5)</sup>.

Cardiac Surgery can be followed by CP, but it occurs in less than 5% of recorded patients<sup>(1)</sup>.

#### Clinical presentation:

*Signs & Symptoms* usually are delayed for several years after the clinical or sub clinical episodes of acute pericarditis<sup>(1)</sup>. So presentation may vary from modest form of CP with early fatigue, with or without modest effort breathlessness and neck vein distension; hepatomegaly with ascitis and intermittent ankle edema<sup>(1,2)</sup>.

These findings may increase progressively to severe cases of orthopnea, paroxysmal nocturnal dyspnea, signs of right sided heart failure with pleural effusion, pulsus paradoxus with reduction in pulse pressure or as massive ascitis especially in children and even generalized edema (anasarca) are reported<sup>(1,2,6,7,8,9)</sup>.

Pericardial Knock, which is an early loud third heart sound due to early ventricular filling in early diastole<sup>(1,2)</sup>. ECG; usually shows diffuse st – segment changes, especially in chest leads. Chest x-ray shows either normal or slight increase in cardiac size with evidence of calcification on the heart shadow in severe cases<sup>(1,2,4,10)</sup>.

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Echo study may show restriction in the movements of the heart chambers with thickening of the pericardium and diminished flow at the outflow large vessels<sup>(1, 2, 4)</sup>.

Cardiac catheterization is usually conclusive of CP; by measuring pressures in all chambers which show evidence of elevated end-diastolic pressure to equal levels in right atrium, pulmonary artery, left atrium and this is considered as the hall mark of chronic CP<sup>(1, 2)</sup>.

Square root sign is also useful and diagnostic for CP, which reveals the dip and plateau of the right ventricular pressure tracing.

Surgery is the definite cure for those patients<sup>(1, 2, 4)</sup>, although some reports mention CP to resolve without surgical intervention by medical therapy<sup>(11)</sup>.

#### **CASE REPORT:**

A child (H. R. S.) of 13 years was referred by a physician to our cardiac center (Ibn Al-Bitar Hospital for Cardiac Surgery) on May 2006. On receiving the patient he

had bilateral leg edema, raised Jugular Venous Pressure (JVP) and was taking aggressive diuretic therapy.

Routine blood investigations were normal, chest x-ray (fig. 1) showed evident calcification rim in the heart shadow, with slight cardiomegally.

ECG showed diffuse ST – segment changes (fig. 2). Echocardiography gave the following data; Aortic Root (AR) 28mm, Left Atrium (LA) 38mm, Left Ventricular Diastolic Dimension (LVDD) 45mm, Left Ventricular Systolic Dimension (LVSD) 30mm and the Ejection Fraction (EF) was 60%. There was mild functional mitral and tricuspid valve regurgitation and both left and right ventricular dysfunction.

Thick pericardium (6 mm), immobile and calcified, with abnormal flow and motion in the Inferior Vena Cava (IVC), so the picture was suggestive of CP or restrictive cardiomyopathy.

CT scan (fig. 3) then confirmed the presence of thickened calcified pericardium and raises the possibility of CP.

Abdominal Ultrasonography (fig. 4) showed engorged IVC with congested liver.

Cardiac catheterization (fig. 5) was diagnostic and marked the equal pressure between the Right and

Left Ventricular End Diastolic Pressures (RVEDP = LVEDP = 22 mmHg.). Pulmonary Artery pressure was (25/ 12/ 15 mmHg.) and equal to RVEDP. Coronaries were normal and the diagnosis was a CP with obvious calcification.

Surgery was performed at the end of May 2006; through median sternotomy, the pericardium was so thick as one layer and calcified with evidence of caseation (tuberculosis was suggested), so peeling was done starting to release the out flow (aorta and pulmonary) and then the in flow (Superior and Inferior Vena Cava - SVC & IVC) then careful removal was done from the right ventricle, the apex and the right atrium) with an improvement in the contractility and cardiac output.

The patient ran a smooth postoperative period without complications with improvement in his ECG (fig. 6) and chest x-ray (fig. 7).

Then he was discharged home in the 7<sup>th</sup> postoperative day with a marked reduction in his leg edema and ascitis.

The histopathology (fig. 8) showed evidence of chronic inflammatory cells and a picture of old healed tuberculosis.

#### **DISCUSSION & CONCLUSION:**

CP is a rare condition in children, although it is recorded in many places as a chronic calcific CP<sup>(8, 12, 13, 14, 15)</sup>.

However tuberculosis (TB) still is a cause of CP with calcification<sup>(16)</sup>, and still can cause major signs and symptoms especially of right sided heart failure.

Presence of tuberculous constrictive pericarditis with such a severe calcification in a child without any primary lesion in the lung is a very rare incidence<sup>(12, 13, 14, 15)</sup>.

We may conclude that:

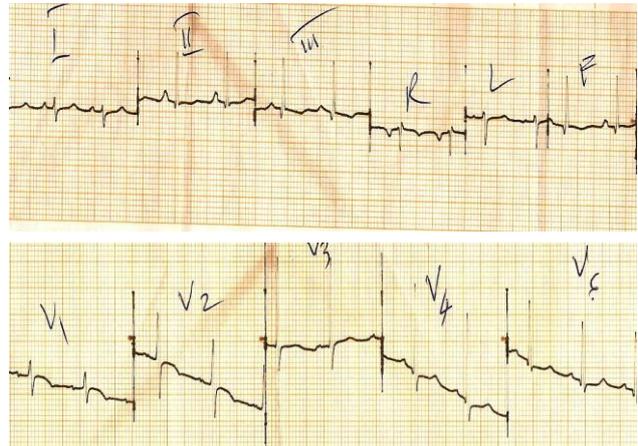
1. TB may come back with different modalities of presentation.
2. Early diagnosis of pulmonary TB is mandatory in controlling the progression of the disease, but the reverse is not true, i.e. TB may manifest in other organs without its classical pulmonary presentation.
3. Calcification should be considered as a possible complication not related to age or to chronicity of the disease.
4. Surgery is the treatment of choice in CP, although medical treatment may suffice sometimes especially in early stages.

## Legends:

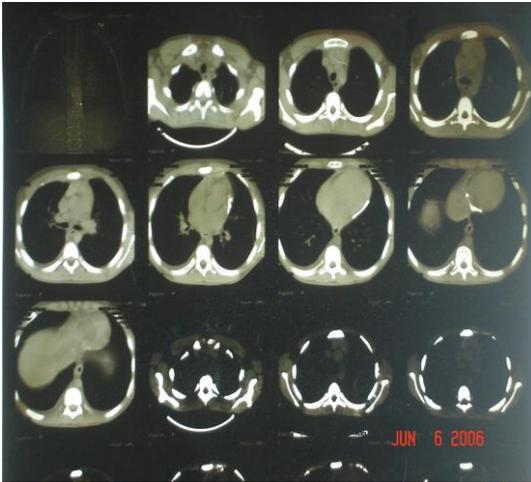
Figure No.	Description	Page No.
Fig. 1	Preoperative CXR showing calcification of the pericardium	10
Fig. 2	Preoperative ECG	10
Fig. 3	Preoperative CT scan	10
Fig. 4	Preoperative abdominal US	10
Fig. 5	Preoperative cardiac catheterization	11
Fig. 6	Postoperative ECG	11
Fig. 7	CXR on discharging the patient	11
Fig. 8	Histopathology	11



**Fig. 1, Preoperative CXR showing calcification of the pericardium**



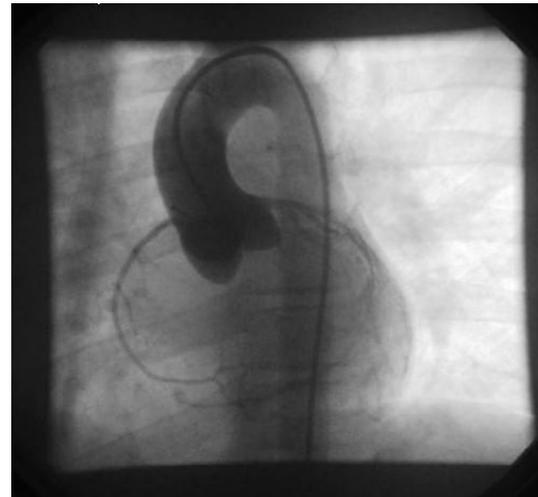
**Fig. 2, Preoperative ECG**



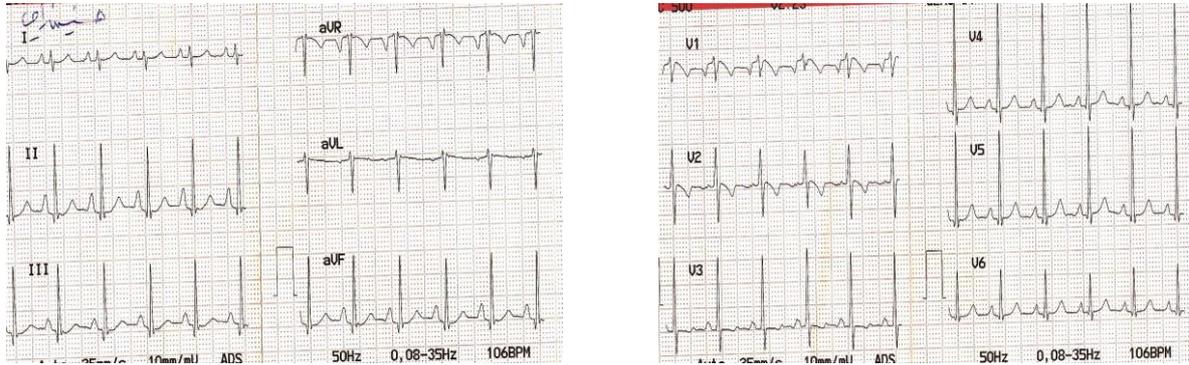
**Fig. 3, Preoperative CT scan**



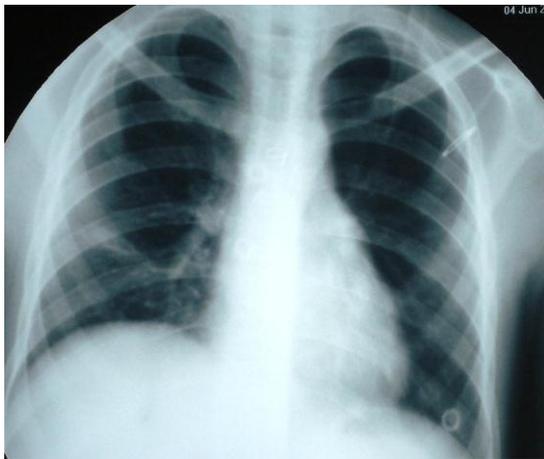
**Fig. 4, Preoperative abdominal US**



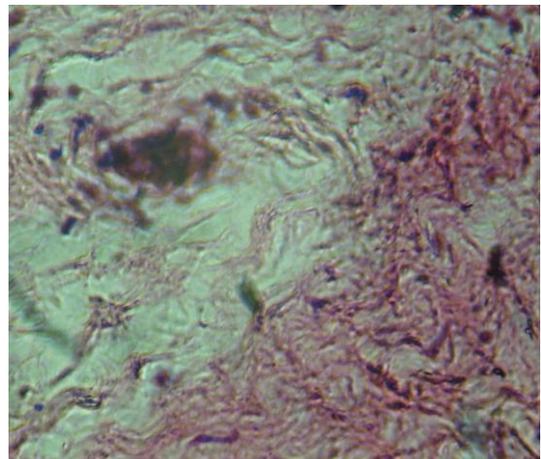
**Fig. 5, Preoperative cardiac catheterization**



**Fig. 6, Postoperative ECG**



**Fig. 7, CXR on discharging the patient**



**Fig. 8, Histopathology**

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