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Pericardiectomy for Managing Constrictive Pericarditis. A Case Report of A 30-Years-Old Saudi Female.

Nora Alsomali[1], Afrah Alsomali[2], AftabTuri[3], Atli Eyjolfsson[4]

[1] College of Medicine, Alfaisal University, [2] College of Medicine, Alfaisal University, [3] King Faisal Specialist Hospital and Research center, [4] King Faisal Specialist Hospital and Research center

ABSTRACT

Background: constrictive pericarditis (CP)is the final stage of a chronic inflammatory process that leads to the thickening and calcification of the heart's pericardium, restricting its natural elasticity and diastolic filling. CP has nonspecific symptoms; hence, requires a high index of suspicion and complete investigations. **Aim of the work**: this is a case of a young Saudi female presented with a 3 years history of shortness of breath associated with abdominal distention**. Patient and Methods**: multimodal imaging, lab workup and cardiac catheterization were done to confirm the diagnosis of CP. **Results**: the patient was managed by total pericardiectomy; during which her calcification was found to be encasing the heart at its length dimension and invading deeply into the myocardium. The patient experienced complicated postoperative course; however, now is doing fine. **Conclusion**: to our knowledge there are no similar cases reported in the literature from Saudi Arabia. Therefore, the aim of this case was to highlight the importance of early diagnosis and management of CP for better prognosis.

Key words: constrictive, pericarditis, pericardiectomy, calcification, Saudi Arabia

INTRODUCTION

Constrictive pericarditis (CP) is a product of an ongoing chronic inflammatory process that result in the thickening and calcification of the heart's pericardium; impairing its diastolic filling **(1,2).** The etiology of CP can differ (Idiopathic, post viral, tuberculous, post-surgical, radiation-induced, etc.); however, they all have the same end results **(1,3).** Tuberculous pericarditis remains to be the most common etiology in both developing and underdeveloped countries, while idiopathic pericarditis, previous cardiac surgery and chest radiotherapy are the common etiologies in the developed countries.

Although the true prevalence of CP remains unknown, it is known to occur in 0.2-0.4% of patients who had undergone cardiac surgery, where idiopathic pericarditis occurs in less than 1% of the patients **(3-5).** Early diagnosis is essential, since CP is considered a potentially reversible cause of diastolic heart failure, but continues to be a challenging clinical diagnosis; due to its nonspecific symptoms, thus, often requires a high index of suspicion **(4).**Echocardiography is proven to be a useful initial imaging method, but cardiac CT and MRI has higher accuracy in detecting pericardial thickening and calcification. The gold standard for diagnosis is cardiac catheterization and pericardiectomy is the only definitive treatment option for patients with definitive constrictive pericarditis **(1).** The CP etiology and management has been well documented in the Western literature, however, we lack adequate information and recent updates regarding CP management in Saudi Arabia **(6).** The case report aimed to highlight the importance of early diagnosis and management of CP for better prognosis and outcome.

CASE REPORT

This is a case of a 30-year-old Saudi female with a 3 years history of shortness of breath (SOB) that got worse on January 2018. In addition, it was associated with abdominal distention, for which she was admitted at a local hospital. During her stay, she underwent CT scan and echocardiography imaging which were suggestive of calcific constrictive pericarditis and was managed with diuretics.

The patient afterwards referred to our hospital for further intervention and investigation. History was also positive for intermittent palpitation and weight loss for 5 years. There was no history of cough, chest pain, fever, loss of consciousness, night sweats, joint pain or urinary tract infection symptoms. No contact with TB patients, previous surgeries or radiations. On physical examination, she had irregular pulse. Chest showed bilateral

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vesicular breathing with decreased breath sounds and dullness over the lower lobes of the lungs. Cardiovascular exam showed audible first and second heart sounds with an added diastolic sound suspicious of pericardial knock, raised jugular venous pressure (JVP), with present abdominal ascites. The rest of the examination was unremarkable. On February 2018, her hospital course was complicated by a new onset of atrial fibrillation that was treated with rate control strategy. Therapeutic thoracentesis was also done to alleviate her respiratory distress. Pericardiectomy was the suggested plan for her pericardial constriction in the multidisciplinary team meeting; However, the procedure was postponed for 1 months due to the family's request.

Investigations:

During her hospital stay, lab workup and serology were within normal limits. Malignant cells and auto antibody markers were not detected. QuantiFERON (a blood test that detects mycobacterium tuberculosis) was negative. Acid-fast bacilli cultures from both the sputum and pleural tap were negative. Chest x-ray **(Figure 1)** showed mild cardiomegaly with pericardial calcification inferiorly and posteriorly along with enlarged pulmonary vasculatures. Bilateral pleural effusion was also seen with adjacent lung ground-glass opacities. CT chest **(Figures 2 & 3)** showed bilateral loculated pleural effusion with adjacent atelectasis suggestive of empyema or chronic hemothorax. There was extensive pericardial calcification more on the anterior and inferior surfaces with mild cardiomegaly.

Echocardiography **(Figure 4)** showed the presences of constriction and thickened pericardial layer, small Left Ventricle (LV), mildly dilated Right Ventricle (RV) with severely dilated atria, exaggerated RV and LV interdependence and no diastolic reversal in stroke volume. Respiratory variation of transvalvular flow with reverse annular tissue Doppler. Cardiac catheterization **(Figure 5)** showed discordance between LV and RV systolic pressure during expiration and inspiration with equalization of end diastolic pressure along with left ventricular waveforms showing a "square root sign" also known as "dip and plateau sign". The results were found to be consistent with her initial diagnosis of constrictive pericarditis.

Treatment and outcome:

On 8 April 2018, the patient was taken to the operative theatre after acquiring informed consent. Median sternotomy approach was used and the surgery did not require switching the patient on a cardiopulmonary bypass. During surgery, the patient's pericardial calcification was invading deep into the myocardium. The calcification measured 15 cm and it encased the heart at its length dimension of 14 cm, severe calcifications over the right atrium, the diaphragmatic side of the right and left heart and around the left ventricle down to the left atrium was found. Careful dissection of the pericardial calcification was done with the removal of the calcifications anterior to the phrenic nerves. There was no cannulation performed and the patient was operated without heparinization. Specimen of the calcified pericardium was sent to the lab for analysis. It consisted of multiple pieces of firm tissue with dark heterogeneous tan brown outer surface as shown in **fig. 6.** The cytopathology of the specimen revealed calcification, chronic inflammation and fibrosis with no granulation tissue. No complications occurred during surgery apart from a small laceration of the right lung, as an attempt of therapeutic evacuation was made. However, it failed to drain due to the severe pleural adhesions. At the end of the surgery, the sternum was closed with separate steel wires and chest tubes were placed in the right pleura and anterior pericardium. The patient was hemodynamically stable and moved to the Intensive care area for monitoring. The patient's postoperative course was complicated by bilateral lung atelectasis, pleural effusion, multiple episodes of atrial fibrillation and oxygen desaturation. Although patient's condition was stabilized, she was in continuous need of oxygen to maintain her saturation in 90s. Hence, she was discharge on home oxygen and given a close follow up appointment. The patient had multiple ER visits and admissions after her discharge, due to symptoms consistent with fluid overload such as increased exertional SOB, increased oxygen requirement, abdominal distention, and bilateral lower leg edema. It has been noticed that the

patient's health improved temporarily during admissions, but soon started to worsen upon her discharge. Eventually patient was admitted because of persistent worsening symptoms and was treated by multidisciplinary team due to the complexity of her symptoms. In addition, upon thoracic surgery recommendation on the 20th of June, right side thoracotomy with decortication was done to decrease her O2 requirements. However, it was complicated by right side pneumothorax and hemothorax with recurrent empyema, which grew *Staph aureus*. Despite all the lung drainage trials, her oxygen demand continued to increase and she became ventilation dependent in critical care unit. As a last resort, thoracic surgery created Clagett window (Which is an opening in the lateral side of the chest created by resecting the posterolateral aspect of the lower rib) to allow continuous drainage of her re-accumulating loculated empyema. After Which, the patient experienced significant improvement and started to maintain her O2 saturation on room air without any supplemental O2 except during sleep. Her tracheostomy was later decannulated and she started to tolerate total oral diet after which the g-tube was removed. The patient was discharge home on per need oxygen therapy whenever the SpO2 drops below 90%.

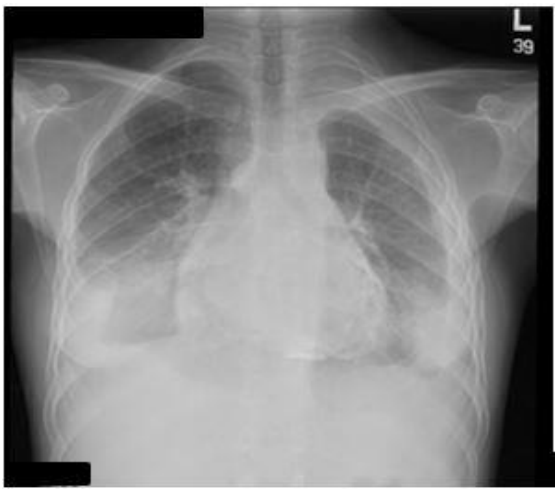

Figure 1: chest X-ray, AP view showing pericardial calcification

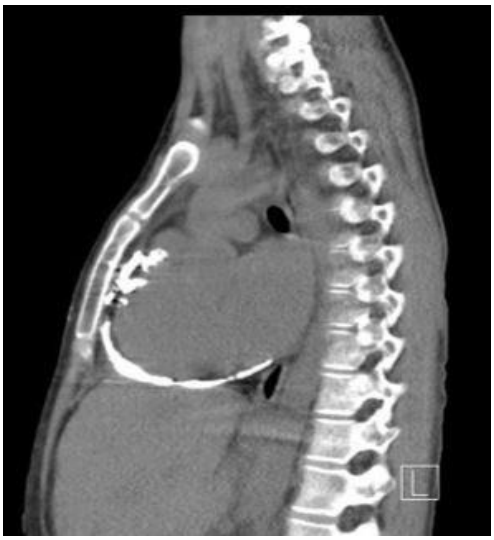

Figure 2: CT scan of the chest lateral view showing extensive hyper density all around the heart extending posteriorly

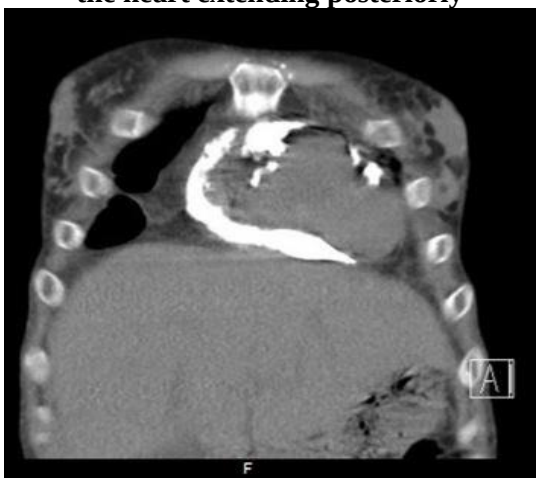

Figure 3: CT scan of the chest anterior view showing extensive hyper density all around the heart extending posteriorly

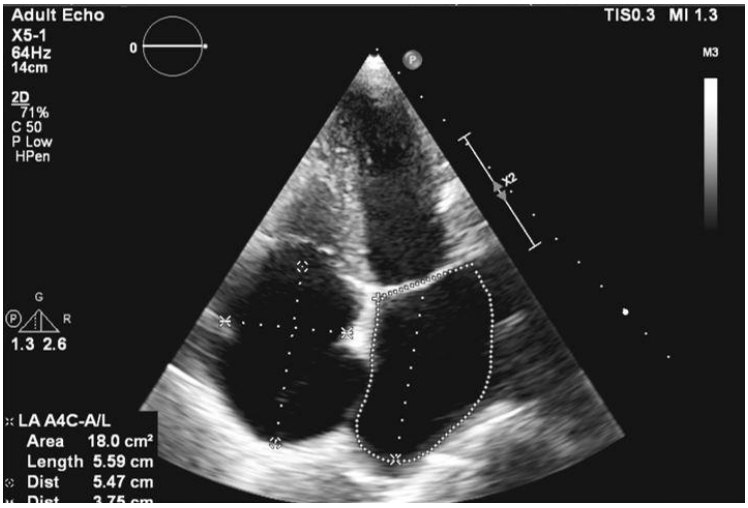

Figure 4: echocardiography showing small left ventricle, mildly dilated right ventricle with severely dilated atria

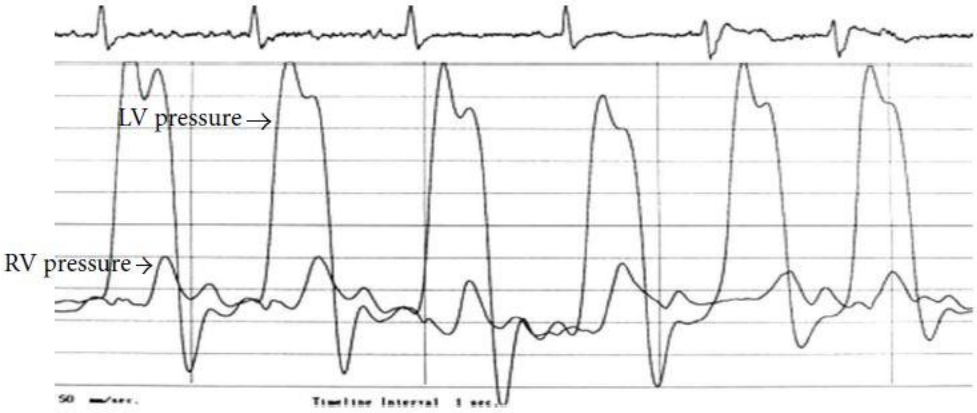

Figure 5: cardiac catheterization showing discordance between LV and RV systolic pressure during expiration and inspiration with equalization of end diastolic pressure

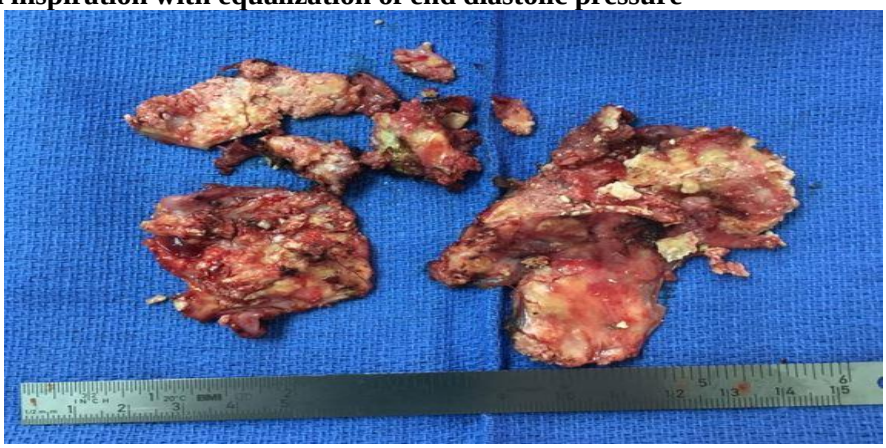

Figure 6: multiple pieces of fibrous pericardium post resection

DISCUSSION

This case illustrated chronic presentation of constrictive pericarditis, which is a rare disease and accounts for 20-40% of constrictive cardiomyopathy cases [3].The true incidence of this disease in general was unknown. Due to its nonspecific presentation CP can go undiagnosed or misdiagnosed[7]. Patients with CP usually presents with symptoms related to fluid overload with variable degrees of severity (Some presents with peripheral edema and some presents with anasarca) or decreased cardiac output symptoms such as elevated JVP, SOB or fatigability[1]. Our patient presented with worsening SOB associated with abdominal distention along with bilateral loculated pleural effusion and chronic hemothorax. Although, there are many common causes of bilateral pleural effusion like renal failure, hypoalbuminemia or hypothyroidism, CP can also result in such presentation. According to results of **Doustkami *et al*.** pleural effusion occurred in about 50% of patient with CP; which was mainly unilateral left sided [8].They added that, bilateral effusion could present; like in their case our patient both had similar distribution of pleural effusion (Bilateral and more on the right side) contraindicating the characteristic presentation of CP.

Since the symptoms of CP are not specific, important other possible diagnosis should be ruled out since they are similar in the clinical presentation; However, they differ in etiology and management. Restrictive pericarditis and congestive heart failure were on the top of the differential list. Restrictive pericarditis is a disease that affect the myocardium via intracellular or interstitial infiltration with or without fibrosis in the absence of LV dilation; resulting in an abnormal LV diastolic filling [3]. There is a similarity between restrictive pericarditis and CP in presentation and reduction of LV compliance to CP. We used multimodal imaging especially echocardiography and cardiac catheterization to confirm the diagnosis of constrictive pericarditis and rule out the other differentials.

According to literature, the most common cause of CP is idiopathic [3,9]. In our case, all the investigations and diagnostic tests for CP etiologies were negative; hence, by exclusion idiopathic CP was thought to be the cause of CP in our patient. Similarly, important causes which are considered endemic in Saudi Arabia such as tuberculosis were ruled out [6].

Pericardiectomy is the preferred and definitive method of treatment in CP patients and relatively safe operation [5,10]. It results in relief of symptoms and satisfactory improvement in function status [7,11]. In addition, a significant improvement was seen in survival rate and quality of life, especially with the idiopathic CP if done early [3,12]. Our patient underwent a successful total Pericardiectomy with median sternotomy approach, which provides good access and exposure of the heart. It also enables good clearance of the disease. The outcome and the postoperative prognosis in this case were compromised due to the patient's concomitant lung disease, but after the creation of the Clagett window and complete evacuation of the puss she had improved significantly. According to studies, CP has an operative risk of 5 to 10% and a late mortality rate of 15-70% depending on underlying etiology and clinical status of the patient [2,11]. Two studies [7,12] conducted in Cleveland Clinic and Mayo Clinic showed that 70% to 80% of patients were free of adverse cardiovascular events 5 years after pericardiectomy and 40% to 50% at 10 years. Moreover, we have found in other studies that long term outcome was predicted by variable factors, such as advanced age, increase NYHA class, post-irradiation exposure, etc., but these factors differed from one study to another [7,12]. Nevertheless, more studies are needed to determine the prevalence of CP and its outcome in Saudi Arabia.

CONCLUSION

Early diagnosis and management of constrictive pericarditis are crucial for better outcome. Long term outcomes after pericardiectomy for CP vary according to the etiology and preoperative clinical condition of the patient; However, pericardiectomy remains to be the best treatment modality and relatively safe operation for CP. Unfortunately, there is no large study done to evaluate the relationship between preoperative risk and postoperative outcome.

Disclaimer:
This case described the management and prognosis of a chronic constrictive pericarditis in a single patient in a tertiary hospital in Saudi Arabia and compared it to the management and outcome to established literature; Thus, the result of this case cannot be generalized, and it is not representative of the Saudi population.

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