Calcific Constrictive Pericarditis "Horseshoe": Two Case Reports

Selman Dumani^{*}, Laureta Dibra, Ermal Likaj , Stavri Llazo, Fjorba Mana, Alessia Mehmeti, Adelina Musliu, Ali Refatllari, Altin Veshti

Service of Cardiac Surgery, University Hospital Center "Mother Theresa", Tirana, Albania

Abstract

Constrictive pericarditis (CP) is an uncommon disease. Defined as a cicatricial process leading to thickening, hardening of the pericardium which ultimately results in loss of pericardial elasticity and restriction of filling of the cardiac chambers.

Pericardial calcification (PC) is the sequelae of inflammation, fibrosis, and necrosis, which is commonly found in constrictive pericarditis, although cases with almost fully/completely calcified pericardium are very rare. Therefore, presenting cases with such a degree of calcification are of high importance to the medical community. Often pericardial calcification is an incidental finding noted during chest imagery as such remains asymptomatic. However, during disease progression, patients develop signs and symptoms of congestion as a result of restriction to ventricular filling caused by the rigid pericardium.

Surgery provides the only effective treatment for patients with symptomatic constrictive pericarditis. We report two cases with "horseshoe" calcification successfully surgically treated in our institution.

Keywords: Pericarditis, Constrictive, Horseshoe calcification

Address for correspondence: Selman Dumani^{*}, Service of Cardiac Surgery, University Hospital Center "Mother Theresa", Tirana, Albania. E-mail: selmandumani@yahoo.co.uk

INTRODUCTION

Varying degrees of pericardial calcification are present in about 80 to 98% of the cases of constrictive pericarditis, although almost completely calcified cases are rather rare (1). The most common etiologies include viral infections, tuberculosis, chest radiation exposure, and following cardiac surgery (2). Pericardial calcification is more common over the right side, anterior and diaphragmatic aspects of the heart in the atrioventricular grooves (3). The most common X-Ray finding is a curvilinear density at the extreme margin of the cardiac silhouette pressure over the last 9 months. On the ECG persistent atrial fibrillation was confirmed. The chest CT and X-Ray identified a pericardial thickening with multiple calcification sites (fig 1a) as well as minimal bilateral pleural effusion. A thoracentesis was performed which concluded that the pleural effusion was transudative. ECHO revealed a normal ejection fraction with mild to moderate MR. A paradoxal movement of the interventricular septum was found and as well as a restrictive pattern of the LV filling and dilated VCI with 50% collapse during inspiration. Cardiac angiogram subsequently was normal.



Figure 1. (a) lateral X-Ray view, "horseshoe" silhouette; (b) lateral X-Ray post-op

(better on lateral view) also known as the "horseshoe" silhouette (4). We are going to present the clinical/imaging findings as well as the surgery outcomes in the following case reports.

CASE 1

A 45-year-old female patient with no underlying medical illness presented with worsening symptoms of dyspnea, exhaustion, and low blood She was later consulted to our cardiac surgery unit, where a decision to perform a pericardiectomy was taken. Through this procedure was identified that the pericardium was mostly thickened (5-6 mm) around the inferior vena cava and above the right atrium and ventricle. We initially began to remove the calcium around the superior vena cava, pulmonary artery and aorta. (Fig 2) After that we



Figure 2. calcification above RV and RA

went on to perform the phrenic-to-phrenic pericardiectomy, decalcifying around the heavily calcified areas leaving only two small areas above the right atrium and ventricle (where the calcium was strongly adhered) in order not to risk damaging them. The surgery was performed via open median sternotomy. Post-operatively, the patient made an uneventful recovery. The final X-Ray showed an important decrease of calcium around the heart. (Fig 1b)

CASE 2

A 60-year-old male patient presented with worsening symptoms of chest pain and dyspnea over the last 2-3 months. He was admitted to the sanatorium hospital because of severe persistent bronchial asthma. On consecutive ECGs paroxysmal atrial fibrillation was confirmed. The chest CT and X-Ray identified a circumferential diffuse calcification of the pericardium (Fig 3a) as well as left pulmonary cystic bronchiectasis. ECHO revealed an ejection fraction of 55% with mild MR and AR and moderate TR, PSAP 45 mmHg. A twisting of the interventricular septum and restrictive diastolic pattern was shown. The right and left atrium were enlarged. Cardiac catheterization showed the "deep plateau" pattern in the left and right ventricle pressure curvature (5).



Figure 3. a) "horseshoe" pattern pre-op; (b) decalcified heart post-op

After consulting it was decided to perform a pericardiectomy, similarly to the first case. After the median sternotomy, we saw that the pericardium was deeply calcified, mostly above the right and left ventricle, with a 6-7 mm thickness. Therefore, we began decalcifying around the aorta and the pulmonary artery working our way down to the diaphragmatic surface and then laterally to the apex. (Fig 4) The goal was to perform a gradual phrenic to phrenic pericardiectomy without perforating the ventricles. At the end of the procedure, we were able to fully free the ventricles from calcium without any complications. The patient made a full recovery and was dismissed 4 days after surgery. The post-operative X-Ray showed successful results of the pericardiectomy. (Fig 3b)



Figure 4. "The birth of the heart"

DISCUSSION

Both patients in the case reports we presented suffered symptoms related to diastolic dysfunction caused by the mechanical restriction of calcified pericardium. Pericardial calcification is in most cases an ongoing process that indicates chronic restrictive pericarditis (6). When symptoms such as shortness of breath, chest pain, elevated jugular venous pulse, pedal edema do not improve after conservative treatment, surgical options are the only remaining solution.

Pericardiectomy can be performed via median sternotomy, anterior thoracotomy or left anterolateral thoracotomy approaches. The aim of pericardiectomy procedure is to remove the calcified or fibrotic pericardium as much as possible. The technical difficulties a surgeon faces during the procedure are related to the degree and the site of calcification. A pericardiectomy can be considered successful when the diastolic filling of the ventricles improves after the procedure and the calcium is removed maximally without damaging the respective surfaces. However, the long-term survival of patients undergoing pericardiectomy is dependent on the etiology of disease that leads to constrictive pericarditis (7).

Although is an uncommon condition, this diagnosis should be kept in mind when patients present cardiac symptoms. Chest X-ray remains a valuable early detector of pericardial calcification. When the onset of the cardiac symptoms has begun and conservative treatment fails to succeed, pericardiectomy remains a gold standard procedure that ensures almost immediate relief from the symptoms of constrictive calcific pericarditis (8).

CONCLUSION

Surgery for extensive calcified constrictive pericarditis remain a challenge for every surgical team and can be performed with excellent results.

Acknowledgements: None declared.

Conflict of Interest Statement: The author declares that have no conflict of interest.

REFERENCES

1. Strang JI, Kakaza HH, Gibson DG, Girling DJ, Nunn AJ, Fox W. Controlled trial of prednisolone as adjuvant in treatment of tuberculous constrictive pericarditis in Transkei. Lancet 1987;2(8573):1418-22.

2. Ling LH, Oh JK, Breen JF, Schaff HV, Danielson GK, Mahoney DW, Seward JB, Tajik AJ. Calcific constrictive pericarditis: is it still with us? Ann Intern Med 2000;132(6):444-50.

3. Wang ZJ, Reddy GP, Gotway MB, Yeh BM, Hetts SW, Higgins CB. CT and MR imaging of pericardial disease. Radiographics 2003:S167-80. 4. Welch TD, Oh JK. Constrictive Pericarditis. Cardiol Clin 2017;35(4):539-549.

5. Talreja DR, Nishimura RA, Oh JK, Holmes DR. Constrictive pericarditis in the modern era: novel criteria for diagnosis in the cardiac catheterization laboratory. J Am Coll Cardiol 2008;51(3):315-9.

6. Lima MV, Cardoso JN, Cardoso CM, Brancalhão EC, Limaco RP, Barretto AC. Constrictive pericarditis with extensive calcification. Arq Bras Cardiol 2011;96(1):e7– e10. doi: 10.1590/S0066-782X2011000100018.

7. Busch C, Penov K, Amorim PA, Garbade J, Davierwala P, Schuler GC, et al. Risk factors for mortality after pericardiectomy for chronic constrictive pericarditis in a large single-centre cohort. Eur J Cardiothorac Surg 2015;48(6):e110–e116. doi: 10.1093/ejcts/ezv322.

8. Haley JH, Tajik AJ, Danielson GK, Schaff HV, Mulvagh SL, Oh JK. Transient constrictive pericarditis: causes and natural history. J Am Coll Cardiol 2004;43(2):271-5.