Diagnostic Dilemma of Constrictive Pericarditis With Concomitant Mitral Stenosis: A Case Report

Kee Soon Chong¹, Ching Kit Chen², See Lim Lim¹

1. National Heart Centre Singapore, Singapore 169609
2. KK Women’s and Children’s Hospital, Singapore 229899

Corresponding author:
Chong Kee Soon,
Department of Cardiothoracic Surgery
Institute Jantung Negara
No 145, Jalan Tun Razak
50400 Kuala Lumpur, Malaysia.
Email: cason23chong@gmail.com

Dear Editor,

Constrictive pericarditis (CP) is due to impediment of cardiac diastolic filling secondary to an inelastic and fibrotic pericardium [1]. Observation of this phenomenon is paramount in the understanding of the pathophysiology through several mechanisms. Firstly, there is dissociation of intra-thoracic and intra-cardiac pressure during respiration leading to respiratory variation of mitral and tricuspid inflow velocities. Secondly, there is ventricular interdependence causing septal bouncing observed on echocardiography during the respiratory cycle. Thirdly, there is impairment of diastolic filling causing an abrupt pause in mid to late diastole. This can be demonstrated during cardiac catheterization as dip-and-plateau ventricular pressure tracing or square root sign. Pericardial calcification is common in CP. It is classically described as “eggshell” or amorphous calcification in the atrioventricular groove.

A 57 year-old lady had open mitral commissurotomy in 1991 for severe rheumatic mitral stenosis (MS). She remained well until 2014 when she presented with severe heart failure symptoms (NYHA Class III-IV). Further questioning revealed that she had recurrent hospital admissions for past 3 years with diagnosis of fluid overload or pneumonia. On examination, she had gross elevation of jugular venous pressure, hepatomegaly, right pleural effusion, ascites and peripheral edema, findings consistent with severe right heart failure. Transthoracic echocardiography (TTE) showed severe MS with preserved left ventricular (LV) ejection fraction. The severity of right heart failure is out of proportion to the severity of MS. Hence, it raised the clinical suspicion of CP. Transesophageal echocardiography (TEE) showed severe MS with typical rheumatic changes characterized by thickened leaflets, commissural fusion and annular calcification. MV orifice by planimetry was 0.9cm² and inflow mean pressure gradient of 9mmHg. Doppler findings showed severe pulmonary hypertension (PHT) with estimated pulmonary artery systolic pressure of 61mmHg. However, there was no significant respiratory variation of mitral and tricuspid inflow velocities to suggest definitive evidence of CP. The inferior vena cava was plethoric and septal bouncing was demonstrated. Cardiac catheterization showed PHT with mean pulmonary artery pressure of 43mmHg and right ventricular (RV) pressure of 69/20mmHg. There was elevation and equalization of filling pressure of all cardiac chambers and square root sign. Similar to TEE findings, there was no significant respiratory variation in LV and RV end diastolic pressure tracing. Despite convincing echocardiographic evidence of severe MS, there was no diastolic pressure gradient across the mitral valve (MV) on simultaneous pulmonary artery wedge pressure (PAWP) and LV end diastolic pressure tracing. This remained so even after 500mls of normal saline infusion. It seems that the PAWP does not appear to be a true indicator of left atrial (LA) pressure. Chest computed tomography (Figure 1) showed extensive pericardial calcification and thickened pericardium.

Overall assessment suggests CP with severe MS. Due to inconsistent findings, there were multiple debates in regard to the management approach. Finally, she underwent percutaneous transvenous mitral commissurotomy in April 2015, which only resulted in marginal symptomatic improvement. Subsequently she underwent pericardiectomy and bioprosthesis MV replacement. Intraoperative findings showed moderate pericardial adhesions and normal epicardium. There was dense pericardial calcification

* Corresponding author. E-mail: cason23chong@gmail.com

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over the left atrioventricular groove likely accounting for the constrictive physiology showed on the cardiac catheterization. There were multiple isolated pearl-like calcified concretions loosely attached to the pericardium and epicardium ranging from 3mm to 1cm in size (Figure 2). Partial pericardiectomy was performed followed by bioprosthetic MV replacement. Postoperatively, she recovered well with tremendous improvement in her symptoms. However, she developed transient RV failure with persistent right pleural effusion. It resolved spontaneously after optimization of diuretics. She was discharged 30 days post-operation.

CP after cardiac surgery is rare, in the range of 0.025-0.3% [2]. Although it is a well recognized disease, its diagnosis can be challenging. It is particularly true in the presence of concomitant MS; the hallmark diagnostic features of CP may not be demonstrated. In CP, there is dissociation of intra-thoracic and intra-cardiac pressure during respiration. Hence, there is a lack of transmission of respiratory changes in intra-thoracic pressure to the heart chambers. During inspiration, intra-thoracic pressure decreases while intra-cardiac pressure remains high. Therefore, there is a reduced total pulmonary venous return and less pressure gradient created between the LA and LV. This leads to reduced mitral inflow velocity during inspiration and the opposite effect is seen with expiration, which will produce the mitral inflow respiratory variation. This hallmark feature was not demonstrated in the TEE and cardiac catheterization. We believe that high LA pressure from the concomitant severe MS has masked this important feature. There is a published case report [3] which showed similar findings. Interestingly this case report able to demonstrate respiratory variation in the PAWP tracing, but not LV end diastolic pressure tracing. Transient RV dysfunction can occur after pericardiectomy in patient with CP, possibly due to myocardial atrophy after prolonged constriction [4]. The management strategy of this condition starts with excluding other causes of RV dysfunction such as pulmonary embolism. In order to improve the right heart function, we need to reduce RV preload by optimization of diuretic therapy. CP is potentially curable with pericardiectomy. Hence, the first prerequisite to successful treatment of CP is the correct diagnosis. The diagnosis of CP can be challenging sometimes especially in cases with concomitant diseased valve, as illustrated in this case. On literature review, there were some case reports [5] which showed that diagnosis of CP was delayed owing to associated MS. This case exemplified the importance of a high index of suspicion of CP in the setting of severe right-sided heart failure that is out of proportion to the pulmonary involvement of a left-sided heart disease [6]. With clinical acumen and appropriate investigations, correct diagnosis can be reached early and surgical management can be instituted to decompress the cardiac constriction before it deteriorated into an irreversible state.

Declarations of Interest
The authors declare no conflicts of interest.

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References