‘Egg-cased heart’: a multimodality imaging approach to chronic constrictive pericarditis with egg-shell calcification: a case report

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Background
Constrictive pericarditis (CP), also known as Pick’s disease, is a sequela of chronic inflammation of the pericardium. Pericardial calcification is a common occurrence in CP; however, extensive egg-shell like calcification is rare. Our case, highlights, how a multi-modality imaging in a middle aged female helped to diagnose chronic constrictive pericarditis (CCP) with egg-shell like calcification encasing the heart.

Case summary
Middle aged female with features of right heart failure, was diagnosed as CP based on two-dimensional echocardiography and cardiac catheterization. Computed tomography (CT) scan chest showed extensive egg-shell like calcification encasing the heart, suggestive of calcific CP. Subsequently, she underwent pericardiectomy, through median sternotomy approach and is currently on follow-up with asymptomatic cardiac status.

Discussion
Extensive pericardial calcification encasing the heart like an egg-shell is rare in CCP. Likelihood of incomplete pericardial resection is high in calcific CP and hence a median sternotomy is preferred over anterolateral thoracotomy. A preoperative non-contrast CT scan defines the thickness, anatomic extent the calcification and its adherence to myocardium and surrounding structures. A reconstructed volume-rendered image delineates the extent of calcification precisely, thus determining the optimum surgical approach. A multi-modality imaging in CP, especially in calcific CP, is thus of paramount importance.

Keywords
Case report • Egg-shell calcification • Chronic constrictive pericarditis • Multi-modality imaging • Pericardiectomy

ESC Curriculum
2.2 Echocardiography • 2.1 Imaging modalities • 2.4 Cardiac computed tomography • 6.6 Pericardial disease

Learning points
• A multi-modality imaging approach in chronic constrictive pericarditis is of paramount importance to diagnose, risk stratify, and plan surgery.
• Median sternotomy approach is preferred for pericardiectomy in presence of extensively diseased pericardium.
Introduction

Constrictive pericarditis (CP), also known as Pick’s disease, is a sequela of chronic inflammation of the pericardium. The result of scarring is impaired diastolic filling of ventricles, leading to elevated filling pressures and systemic venous congestion. Thickened calcific pericardium, encasing the heart, is a rare occurrence in CP. Delineation of the exact location of pericardial calcification and thickness is essential as it facilitates risk stratification and surgical planning.

Our case was diagnosed as chronic calcific CP (egg-shell like calcification) and underwent successful pericardial resection. This case highlights the need of multi-modality imaging in the diagnosis and management of chronic calcific CP.

Timeline

Case presentation

A 45-year-old female, with no significant past medical history, presented with features of refractory right heart failure. Jugular venous pressure (JVP) was 18 cm with prominent Y descent. Kussmauls sign, increase in JVP or failure to fall on deep inspiration, was present. An early diastolic sound characteristic of pericardial knock was best heard at left sternal border. Her baseline biochemistry was normal with no evidence of hepatic or renal dysfunction. Chest X-ray showed cardio-thoracic ratio of 0.5 with radiopaque density suggestive of calcium around cardiac silhouette which was better appreciated on fluoroscopy. Transthoracic two-dimensional echocardiogram showed mildly dilated atria with a near normal left ventricular systolic function, normal ventricular wall thickness, and hyperechoic shadow surrounding the ventricle suggestive of thickened pericardium. Septal bounce, exaggerated respiratory variation...
Figure 1  (A) Chest X-ray in PA view shows calcified pericardium (black arrows), which is better delineated in B: lateral cine-fluoroscopic frame of the heart.

Table 1  Salient echocardiographic features of constrictive pericarditis in our patient

| Features                                    | Mechanics                                                                 | Comments                                                                 |
|---------------------------------------------|---------------------------------------------------------------------------|--------------------------------------------------------------------------|
| Abnormal septal motion:                     | i. Exaggerated interventricular interdependence.                          | Equivalent to discordant relationship of ventricular systolic pressures during respiration on cardiac catheterization. |
| Septal bounce/shudder/diastolic check-ing   | ii. Rapid cessation of filling during early diastole due to pericardial constraint. | Equivalent to square root sign/dip and plateau pattern of ventricular filling pressures on cardiac catheterization. |
|                                             | (2D echocardiography and M-mode)                                          | Clinically equates to rapid y descent on JVP (Friedreich’s sign) and pericardial knock. Can also be measured in outflow tract, pulmonary vein, or hepatic veins. |
| Respiratory variation during filling:       | Increase in peak e velocity across a. Tricuspid valve is >40% in inspiration. | Dissociation of intra-thoracic and extra-thoracic pressures causing exaggerated interventricular interdependence. |
|                                             | b. Mitral valve is >25% in expiration.                                    |                                                                          |
| Augmented longitudinal motion of the heart. |                                                                             |                                                                          |
| Annular tissue velocity (e’):               | Tethering effects of disease pericardium limits lateral expansion of the heart resulting in medial e’ >lateral e’. | This phenomenon is called annulus reversus. Reversal of normal phenomenon where lateral annular tissue velocities are more than medial. |
| Medial e’ is normal or even elevated.       | Haemodynamic studies revealed elevated filling pressures.                 | This phenomenon is called annulus paradoxus. Normally, the filling pressures are in direct correlation with E/e’ |
| Lateral e’ is decreased.                    | However, due to compensatory exaggerated longitudinal movement of heart, E/e’ not elevated. |                                                                          |
| Inverse correlation between filling pressures and E/e’. |                                                                            |                                                                          |
| Systemic venous congestion:                 | Impaired ventricular filling                                               | Clinically evident by features of right heart failure.                   |
| Congested and non-collapsing IVC            | i. Thickened pericardium with or without calcification.                   |                                                                          |
| Other parameters                            | ii. Normal ventricular wall thickness.                                    |                                                                          |
|                                             | iii. Normal or near normal ventricular function.                          |                                                                          |
|                                             | iv. Absence of severe bialtrial enlargement.                               |                                                                          |
in both tricuspid and mitral inflow velocities (e) and annulus reversus (medial e’ > lateral e’) were other features of chronic constrictive pericarditis (CCP) that were demonstrated in our patient (Table 1, Figures 2 and 3). Right heart catheterization revealed elevated and equalization of diastolic pressure, classical dip and plateau pattern with discordance of right and left ventricular systolic pressures during respiration. Pulmonary artery pressures were 32/10 (m22) mmHg. Epicardial coronaries were normal on angiogram with no external compression or pinching detected.

Subsequent imaging with non-contrast computed tomography (CT) chest detected the presence of a thickened, extensive, asymmetric, and circumferential egg-shell like calcific pericardium encasing the heart (Figure 4). A volume-rendered CT precisely delineated the extent of calcification. A pericardiectomy through median sternotomy approach was performed. However, densely adherent pericardium over posterior surface was not resected. Histopathology revealed moderate fibrosis with calcification with no evidence of granulomas (Figure 5). Postoperative CT revealed denuded anterior and lateral surface of the heart with minimal residual pericardium over the posterior surface of heart (Figure 4E and F). Tissue Doppler velocities revealed improved ventricular filling with no respiratory variation across the tricuspid and mitral inflow (Figure 3C and D). Inferior vena cava size was normal with normal collapsibility although septal bounce was still present on echocardiogram. She is currently on follow-up for more than 2 years with asymptomatic cardiac status. She showed no features of constriction on follow-up transthoracic echocardiogram and continues to have near normal left ventricular function.

Discussion

Chronic constrictive pericarditis, often called as Pick’s disease—Pick described the presence of liver involvement and preponderant ascites in CCP—is a state of ‘inflow stasis’. The pericardial constraint imposed by the inelastic and thick pericardium in CCP, limits the normal diastolic filling. 1 Cardiac filling occurs only in the early part of diastole and patients present with features of biventricular failure. 1 Unless pericardiectomy is performed, symptoms persists and progress in majority of patients. The timing of pericardiectomy is thus of paramount importance. Pericardial resection remains the only causal treatment available for CP and should not be delayed after symptom onset. Removal of the thickened, inflamed, and adherent pericardium is technically challenging and associated with a high perioperative mortality, ranging from 2% to 20%.2-4 New York Heart Association class IV symptoms, concomitant coronary artery disease, reduced left ventricular function, and radiation-induced disease are risk factors for poor outcomes.3-5 Obstructive airway
Figure 3  Doppler spectra across atrioventricular valve showing exaggerated respiratory variation. Increase in peak e velocity across tricuspid valve during inspiration >40% increase (A) and >25% during expiration across mitral valve (B) is characteristic of chronic constrictive pericarditis. Post-pericardiectomy, no respiratory variation is seen across tricuspid (C) and mitral valve (D). Yellow block arrows depict inspiratory and red block arrows depict expiratory phase.

Figure 4  Non-contrast computed tomography sections of the heart at various planes. Thickened, extensive, and asymmetric calcification around the heart (arrow heads) is clearly delineated in pre-operative computed tomography images in axial (A), sagittal (B), and coronal (C) planes. A non-contrast computed tomography reconstructed volume-rendered image precisely delineates the extent of `egg shell’ like calcification as shown in D (blue block arrows). Post-pericardiectomy, axial computed tomography images (E and F) show complete absence of the diseased pericardium over anterior and lateral surface, with minimal residual unresected calcified pericardium (black arrows) at the posterior surface of the heart.
Disease and preoperative renal and or liver dysfunction are other independent non-cardiac risk factors for early and late adverse outcomes post-pericardiectomy. Pericardial decortication in these subgroups of patients should be considered cautiously, as they may not benefit from surgery.

One in four cases of CP are associated with pericardial calcification. However, extensive egg-shell like calcification, as present in the index case, is rare. Likelihood of incomplete pericardial resection is high in calcific CP which portends a poor prognosis. Efforts should be made to excise as much of the diseased pericardium as possible and hence a median sternotomy is preferred over anterolateral thoracotomy. Clinical examination and transthoracic echocardiogram along with haemodynamic catheterization confirm the diagnosis of CCP. Chest X-ray and cine-fluroscopy help to ascertain the presence or absence of calcification. A non-contrast CT scan defines the extent of calcification and involvement of the myocardium and surrounding structures. By delineating the exact location and severity of pericardial thickening (normally less than 2 mm) and calcification, a pre-operative CT scan facilitates risk stratification and surgical planning. Cardiac magnetic resonance (CMR) can be considered as an adjunct to above imaging modalities. Although less sensitive than CT scan to detect pericardial thickness and calcification, CMR provides better information regarding the haemodynamics of pericardial constraint. It is of particular importance to identify transient and reversible form of CP. Late gadolinium enhancement (LGE) with thickened pericardium predict resolution of constriction following anti-inflammatory drugs therapy. In cases of non-calcific CP, demonstration of no residual inflammation (a negative LGE sequence) has recently emerged as an important step before pericardiectomy.

A multi-modality imaging is of paramount importance in management of CCP.

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**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

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