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Case Report

Egg-shell calcification in Pick's disease (chronic constrictive pericarditis) ☆☆☆

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ABSTRACT

Egg-shell calcification is rare in cases with constrictive pericarditis. It leads to significant right heart failure and the only treatment is surgical excision of the pericardium. We present a case of a 22-year-old-male who was diagnosed to have severe pericardial calcification on the chest X-ray, which eventually led to a diagnosis of constrictive pericarditis and required an early surgical correction. This case shows that a common diagnostic test such as a chest X-ray does help to diagnose a severe systemic condition.

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Introduction

Pericardial calcification is a rare finding and one of the rarer conditions associated with it is constrictive pericarditis (Pick's disease) [1]. Severe pericardial calcification causes pericardial plasticity and reduced filing, leading to systemic congestion [2]. We present a case of constrictive pericarditis with egg-shell calcification, which was confirmed during subsequent surgery.

Case report

A 22-year-old-male, presented with predominant right heart failure (with edema feet, ascites) and the chest X-ray showed radiopaque shadow surrounding the cardiac silhouette (Fig. 1 Panel A). This is classically referred to as “egg-shell calcification.” There was a history of untreated tuberculosis in childhood. The patient was then evaluated for his right heart failure and severe pericardial calcification.

Two-dimensional echocardiogram was done which showed biatrial dilatation and hyperechoic shadow surrounding both the ventricles. Medial annular tissue velocity was higher than lateral annulus (annulus reversus). Inflow Doppler revealed an inspiratory increase in tricuspid inflow velocity with a corresponding decrease in the mitral inflow velocity. In view

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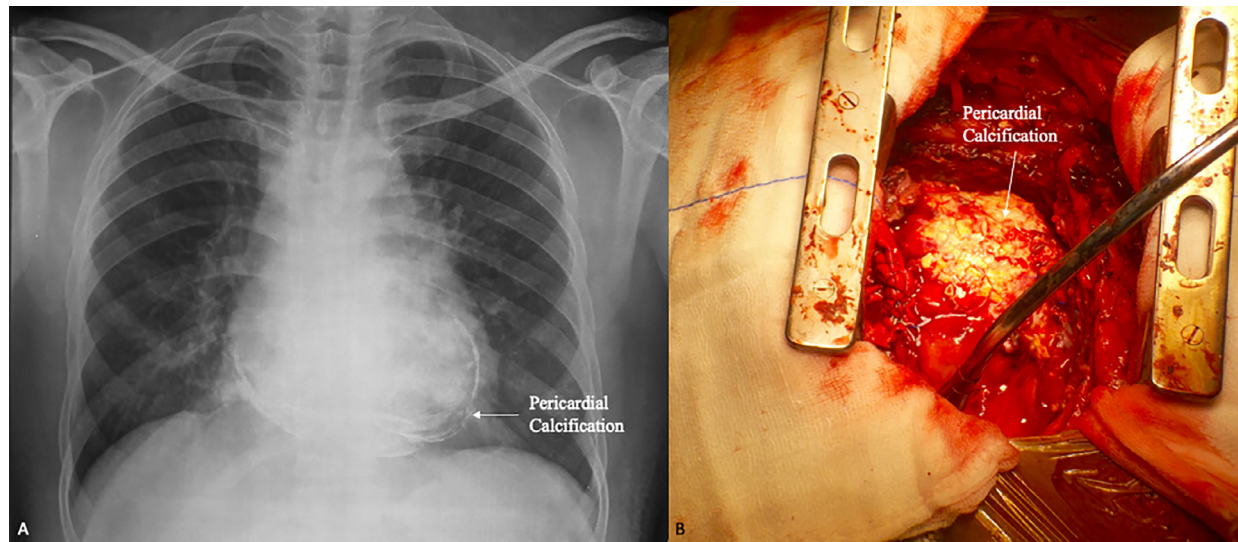


Fig. 1 – Pericardial calcification. (A) Chest X ray at presentation; (B) intraoperative.

of significant restrictive physiology on echocardiogram, a diagnosis of chronic constrictive pericarditis was made and the patient was taken up for invasive hemodynamic assessment to confirm the diagnosis. During right heart catheterization, right atrial mean pressure was elevated. Simultaneous left and right ventricular pressure tracings showed diastolic equalization of the pressures and ventricular interdependence. A typical “square root sign” of ventricular filling could be seen. It signifies that the early diastolic filling is rapid but it is halted abruptly when the volume limit is reached due to the stiff pericardium [3]. Given these typical features on invasive hemodynamic assessment, a diagnosis of chronic constrictive pericarditis (Pick’s disease) with severe calcification was made.

The patient was then taken up for surgical excision of pericardium as the treatment of chronic constriction. During the surgery, it was noticed that some part of the pericardial calcification was adherent to the myocardium (Fig. 1 Panel B). Hence, only a part of pericardium could be removed. The patient succumbed to the illness after around 1-year postsurgery.

Discussion

Pericardial constriction is a result of scarring and it leads to loss of elasticity of the pericardium [4]. The association of calcification with this condition has been previously reported, which can be global or regional [5]. Egg-shell calcification is a rare finding on X-ray.

The evaluation of right heart failure with severe pericardial calcification requires certain protocol, which included echocardiography, invasive hemodynamic assessment, and further imaging with computed tomography (CT) [5]. In our patient, CT could not be done due to financial constraints on the part of the patient. But we could conclusively diagnose the condition with the help of other investigational modalities.

Pick’s disease was described in 1896 to describe the patients with constrictive pericarditis with ascites and hepatomegaly [6]. The predominant cause in the western world is viral pericarditis followed by surgical pericarditis but in the developing world, tuberculosis remains a major cause [7]. The common symptoms are in the form of right heart failure (ie, ascites, peripheral edema, hepatomegaly, etc). [8] Pericardiectomy with complete decortication (whenever feasible) is a reasonable surgical correction and the long-term outcomes of this were found to be dependent on the cause of the disease, with idiopathic disease and successful decortication having the best prognosis postsurgery [9].

The only treatment of calcified pericardial constriction is surgical correction [10]. The 20 years follow up study showed overall mortality of pericardiectomy was 7.9%. It was significantly higher in patients with constrictive pericarditis [10]. Our patient was taken up for surgery on a timely basis but was found to have deeper myocardial involvement which precluded complete pericardiectomy.

Conclusion

Egg-shell calcification is a rare radiological finding and these patients should be evaluated for constrictive pericarditis, which if left untreated can be fatal.

Key teaching points

Although rare, the association of pericardial calcification with constrictive physiology should always be ruled out with a structured investigational protocol.

Patient consent statement

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2021.01.042](https://doi.org/10.1016/j.radcr.2021.01.042).

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