Restrictive cardiomyopathy caused by diffuse calcification of the left ventricle after 20 years of haemodialysis

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Abstract
Valvular and vascular calcifications are common among patients with end-stage renal disease, but diffuse calcification of the left ventricle is rarely reported. We report on a rare case of restrictive cardiomyopathy resulting from severe myocardial calcification and review the literature. A 77-year-old man was diagnosed with end-stage renal disease after having received regular haemodialysis for 20 years. He was referred to our emergency room due to exertional dyspnoea and exacerbated shortness of breath. A chest X-ray revealed severe pulmonary oedema and bilateral massive pleural effusion. Transthoracic echocardiography revealed impaired diastolic function of the left ventricle but preserved systolic function with a 50% ejection fraction. Repeat chest computed tomography demonstrated exacerbation of the calcification from the mitral annulus to the whole circular left ventricle. A coronary angiogram revealed non-significant stenosis, and right heart catheterisation demonstrated elevated pulmonary capillary wedge pressure. He was discharged after two weeks of conservative medication.

Keywords: cardiac calcification, restrictive cardiomyopathy, heart failure

Case report
Our patient was a 77-year-old man with a history of end-stage renal disease who had received regular haemodialysis for 20 years. He was diagnosed with an old cerebrovascular disease with right hemiplegia, diabetes mellitus and coronary artery disease after percutaneous coronary intervention. He was also diagnosed with severe mitral annulus calcification without stenosis or regurgitation two years earlier.

At this visit, he was sent to our emergency room due to symptoms of exertional dyspnoea, exacerbated shortness of breath, palpitation and hypotension during haemodialysis. His heart rate was 164 beats per minute and his blood pressure was 101/63 mmHg.

A 12-lead electrocardiography revealed atrial flutter. The chest X-ray in the emergency room revealed pulmonary oedema and bilateral pleural effusion (Fig. 1). Transthoracic echocardiography revealed impaired diastolic function of the left ventricle but preserved systolic function with a 50% ejection fraction. Chest computed tomography revealed diffuse circular calcification of the left ventricle and massive bilateral pleural effusion (Fig. 2).

Thyroid-stimulating hormone level was 1.50 µU/ml and free T4 level was 1.51 ng/dl, which suggested normal thyroid function. The free calcium level was also in the normal range of 4.93 mg/dl. The coronary angiogram revealed no significant stenosis requiring percutaneous coronary intervention. However,
A right heart catheter revealed a high pulmonary capillary wedge pressure at 28 mmHg. Diastolic congestive heart failure with acute pulmonary oedema was diagnosed based on these findings, which was suspected to result from diffuse myocardial calcification.

The patient was excluded from the heart transplantation waiting list given his age and uraemic state. Furthermore, he refused any invasive surgical procedures and declared ‘do not resuscitate’.

The patient underwent bilateral pleurocentesis to relieve the symptoms. We prescribed inotropes to preserve cardiac function, beta-blockers and amiodarone to control the rhythm, and warfarin to prevent thromboembolism. Albumin supplements were also administered to reduce pulmonary oedema. Eventually, the pulmonary oedema faded, and the inotropes were tapered off under stable haemodynamic status. He was released from our hospital and attended regular out-patient follow-up visits.

Discussion

The aetiologies of myocardial calcification are categorised into dystrophic, metastatic and idiopathic calcification. With underlying end-stage renal disease, patients frequently exhibit calcium deposits throughout the body as metastastic calcification.3 Metastatic myocardial calcification is a frequent cause of heart failure in patients undergoing haemodialysis,4,5 which may cause wall motion abnormalities.

Infection-related myocardial calcification, both bacterial and viral, has been reported to occur within a severe systemic infection or a myocarditis episode. The bacterial pathogens *Pseudomonas* spp. and *Klebsiella pneumoniae* are not uncommon in a septic episode, and hypoperfusion resulting from septic shock may cause the sequela of myocardial calcification.6 Epstein–Barr virus was reported to cause rhabdomyolysis and myocarditis in a clinical case report,7 resulting in dystrophic calcification of the myocardium. We also detected possible related infectious pathogens, but no positive results were confirmed. Myocarditis was excluded given the normal serum levels of creatine kinase (41 U/l) and troponin I (51 pg/ml).

Diffuse massive myocardial calcification can also cause restrictive cardiomyopathy.8 In 1984, Silver et al.9 described the first case of massive endocardial calcification of the left ventricle. They reported that exertional dyspnoea and substernal chest pain were the initial symptoms. The symptoms were exacerbated by calcific deposits in the left ventricle, which became severe.
within five years. Eventually, the patient suffered from recurrent periodic episodes of acute pulmonary oedema. In our case, the patient presented with similar symptoms and arrhythmia during haemodialysis. Pulmonary oedema and bilateral pleural effusion resulted from diastolic congestive heart failure at admission.

No effective management of restrictive cardiomyopathy resulting from fibrosis or calcification has been identified. The current medication strategy is conservative treatment, which could reduce the symptoms of pulmonary and general oedema by reducing the filling pressures and enhancing systolic pump function. We controlled intravascular fluid status through regular haemodialysis and administered inotropes to preserve cardiac function. Furthermore, strict control of the heart rate and blood pressure was critical in preventing arrhythmia events and sequelae as well as unstable haemodynamics.

If optimal medication fails, heart transplantation remains the gold standard for end-stage restrictive cardiomyopathy. Ventricular assist devices represent an alternative treatment. Salas de Armas et al. reported a case of implantation of a left ventricular assist device in a heart failure patient with a heavily calcified left ventricle. They performed the non-excisional technique to preserve the left ventricular cavity. However, the use of ventricular assist devices for end-stage restrictive cardiomyopathy is rarely reported in the literature, and the benefits are limited because the small left ventricle always results in a suction event.

Topilsky et al. analysed 83 end-stage heart failure cases waiting for heart transplantation, including eight restrictive or hypertrophic cardiomyopathies and 75 dilated and ischaemic cardiomyopathies, who received axial-flow left ventricular assist device therapy. The one-year actuarial survival rate did not differ between the two groups. These findings support the option of ventricular assist device implantation in patients for whom heart transplantation is not feasible and management with medication is not satisfactory.

In addition to the use of a ventricular assist device, minimisation of inflammation, avoidance of calcium, correction of phosphate positive balance, and correction of vitamin D and K deficiencies were reported to be effective in the prevention of cardiovascular calcification. Sodium thiosulphate has been used to treat calciphylaxis, but a systematic review and meta-analysis revealed no significant clinical benefit.

Kuzela et al. analysed via autopsy soft tissue calcification in chronic patients who had undergone dialysis, and determined that 78.5% of patients displayed general soft tissue calcification. Notably, our patient had only isolated left-side cardiac calcification, including the mitral annulus, septum and left ventricular myocardium. The calcification surrounding the left ventricle, which restricted the left ventricle in an eggshell, eventually resulted in diastolic dysfunction and pulmonary oedema.

Conclusion

We report on this rare and interesting case of isolated left ventricular calcification in a patient after 20 years of haemodialysis. Although we preserved his cardiac function with optimal medication, the disease may eventually be exacerbated into end-stage heart failure and pulmonary hypertension. Moreover, the prognosis is expected to be extremely poor in the near future. No accurate survival estimates for diffuse left ventricular calcification are reported in the literature. However, the five-year survival rate for all types of restrictive cardiomyopathy is only approximately 50%.

References