Myocardial Calcinosis in Chronic Renal Failure

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ABSTRACT

The authors are presenting an 18 year old male with history of end stage renal disease and rejected renal transplant. In his workup echocardiogram and non contract CT of chest revealed diffuse endocardial and myocardial calcifications. Extensive cardiac calcification is a rare but important entity in relation to end stage renal disease as it may cause complications such as valvular dysfunction and fatal arrhythmia.

CASE REPORT

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The patient, an 18 year old Filipino male, first presented to our institution with hematuria and hypertension, with known end stage renal disease secondary to focal segmental glomerulosclerosis. He had undergone a cadaveric renal transplant four years earlier and was found at initial presentation to be in chronic rejection of the renal transplant. The patient subsequently underwent a transplant nephrectomy and was placed on peritoneal dialysis. Two years prior to current admission, the patient received his second cadaveric transplant but rejected it five months later of the same year.

The patient's most current echocardiography imaging showed normal left ventricular (LV) systolic function with severe concentric hypertrophy and decreased cavity size (Fig. 1). The echocardiogram also demonstrated severely elevated pulmonary artery systolic pressure, mild mitral regurgitation, and moderate dilatation of the atria and right ventricle.

The patient underwent a noncontrast CT scan of the chest for suspicion of lung disease, which showed cardiomegaly, LV hypertrophy and hyperdense atrial and ventricular walls due to endocardial and myocardial calcifications (Fig. 2). Pulmonary arteries were moderately dilated with main pulmonary artery measuring 3.7 cm. Atrophic calcified kidneys and dural calcifications were also noted.

DISCUSSION

Disease of the cardiovascular system accounts for about half of all deaths in patients with end-stage renal disease (ESRD) (1). Abnormal calcium and phosphate metabolism in ESRD is thought to account for the majority of heart structure calcification. In patients with hyperparathyroidism secondary to chronic renal failure, calcium is deposited at an increased rate into cardiac tissues because of the increased phosphate Although earlier studies have suggested that the presence of calcification in the heart is not in direct relationship with serum calcium, phosphate or parathyroid hormone levels; others argue that uremia is the main culprit of cardiac calcification and metastatic calcification of previously normal heart tissue is thought to be the result of increased levels of calcium ,phosphorus, parathyroid hormone and alkalosis (2-5). Metastatic myocardial calcifications are commonly (59%) discovered at autopsy in those patients with end stage renal disease undergoing maintenance dialysis (2).

Vascular calcification is common in patients with renal disease, as coronary calcifications can arise from hyperphosphatemia, hyperparathyroidism, and hyperuremia. Therefore, the etiologies of vascular calcification have been studied more broadly than those of cardiac calcinosis. Patients with end-stage renal disease treated by intermittent

hemodialysis have frequent and progressive valve disease. The most common valvular pathologies in ESRD are sclerosis and calcifications of mitral and aortic valves. Mitral annular calcification (MAC) is seen in 36% and aortic valve calcification (AVC) is found in 28% (6). Our case showed calcifications of both aortic and mitral valves (Figure 2). MAC usually starts on the ventricular side of the mitral annulus near the base of the posterior mitral valve leaflet (7). Mitral calcification in its most severe form can include the entire posterior mitral annulus and in some cases the anterior mitral annulus. MAC commonly causes mitral regurgitation and, in severe form, mitral stenosis. A common echocardiographic finding in association with MAC is left atrial dilatation (7).

AVC and resulting stenosis is also commonly seen in patients suffering from ESRD, and in the general, aging population. However, AVC progresses more rapidly in patients with ESRD, with a reported incidence of 30 to 55% as compared to 21 to 29% in the normal population over the age 65 (8, 9). It has been discovered that the hyperphosphatemia component elevating calcium phosphate is one of the main predictors of AVC in the ESRD patient (10). AVC can cause aortic stenosis and aortic insufficiency.

Calcification of the cardiac muscle is more sinister than usual vessel and valvular calcification associated with renal disease and is sometimes difficult to detect prior to autopsy (2, 11). Metastatic calcifications, arising in previously healthy myocardium, are seen in patients with bone disease, hypercalcemia, hyperphosphatemia, renal disease or those on chronic dialysis. Complications arising from cardiac calcification include complex atrial and ventricular arrhythmias, coronary events and sudden cardiac death, with arrhythmia being the most common cause (12). Cardiac calcification was reported before in literature but in much less extend. One report details a 31 year old woman with ESRD secondary to hypertension on long term dialysis for 10 years with diffuse deposition of calcium within the myocardium (13). In addition, there is a case report detailing a 29 year old woman with chronic reflux pyelonephritis undergoing 8 years of hemodialysis with extensive myocardial and left ventricular wall calcification (14). Lastly, a case report describes a 45 year old man suffering from autosomal dominant polycystic kidney disease resulting in end stage renal disease with left and right ventricular myocardial discovered to have numerous tiny hard yellowish nodules, composed of fibrosis and calcinosis, upon autopsy (15). Therefore, it is apparent that such extensive cardiac calcification is a rare entity in relation to end stage renal disease and very few similar cases have been reported in the literature as described above.

TEACHING POINT

Metastatic calcification is an entity need to be considered in patients with bone disease, hypercalcemia, hyperphosphatemia, renal disease or those on chronic dialysis. Complications arising from cardiac calcification include valvular dysfunction, complex atrial and ventricular arrhythmias, coronary events and sudden cardiac death.

ABBREVIATIONS

AA=ascending aorta,
AVC= aortic valve calcification
CT= Computerized tomography
ESRD= end stage renal disease
MAC= mitral annular calcification
LA=left atrium
LK=left kidney
LV=left ventricle
RA=right atrium
RK=right kidney
RV=right ventricle

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FIGURES

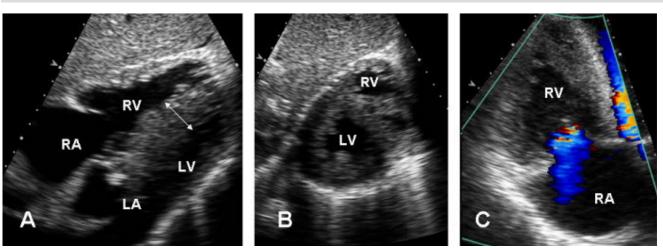


Figure 1: 18 year old male with myocardial calcinosis in chronic renal failure. Transthoracic echocardiography images obtained at four chambers (A, C) and short axis (B) projections demonstrate thickened ventricular walls including interventricular septum (double headed arrow). The interventricular septum measured 1.4 cm. Moderate dilatation of both atria and right ventricle are evident. Moderate tricuspid regurgitation in systole is shown in C. LA=left atrium, RA=right atrium, RV=right ventricle, LV=left ventricle.

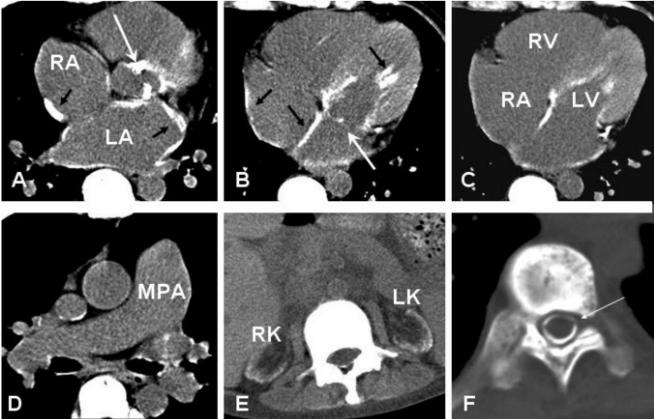


Figure 2: 18 year old male with myocardial calcinosis in chronic renal failure. Axial slices of a noncontrast CT of the chest are presented. Upper row images (A-C) demonstrate calcified endocardial surfaces and myocardium of both atria and left ventricle (small arrows). Calcification of the interatrial septum and papillary muscles are shown in B (small arrows). Concentric hypertrophy of the LV is best shown on C. Note, dilated right and left atria as well as the right ventricle. Calcifications of aortic and mitral valves are shown in A and B (long white arrows). D demonstrates enlarged pulmonary arteries consistent with pulmonary hypertension which was confirmed by echocardiography (80 mm Hg). Also shown are small calcified kidneys (E) and dural calcification (arrow in D). LA=left atrium, RA=right atrium, RV=right ventricle, LV=left ventricle, MPA=main pulmonary artery, RK=right kidney, LK=left kidney.

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KEYWORDS

Myocardial calcinosis, chronic renal failure, calcification, arrhythmias, heart failure

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