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Reactive amyloidosis and end stage renal disease: Two important factors for rapid evolution of constrictive pericarditis

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Abstract

A 61 years old male patient with reactive amyloidosis was admitted for worsening renal function, signs and symptoms of heart failure and diuresis contraction. Compared to year ago, echocardiogram showed worsening valvular pathology: aortic and mitral insufficiency become severe while aortic and mitral stenosis moderate. There was also a restrictive diastolic pattern with elevated filling pressure but medial mitral annulus tissue-Doppler demonstrated elevated early diastolic velocities (e'). Severe pulmonary hypertension and right ventricular dysfunction were also present. The pericardium was hyperechogenic and there was partially organized pericardial effusion too. Suspecting constrictive pericarditis, the patient underwent cardiac catheterization. It confirmed diagnosis.

The most likely mechanism of this rapid progression may lie in the elevated blood values of phosphate and calcium secondary to worsening renal failure. Hyperphosphatemia has been described as a key factor in vascular and other extra skeletal calcification and a risk factor for cardiovascular mortality. Pro-inflammatory cytokines that are increased in chronic kidney disease and in reactive amyloidosis have pro-osteogenic potential.

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Keywords: Reactive amyloidosis; Amyloidosis; Constrictive pericarditis; Cardiac valve calcification; Dialysis; Dip-to-plateau sign; End-stage renal disease; Hyperphosphatemia; Hypercalcemia.

Introduction

We describe a rare case of a 62-years- old Caucasian man with severe kidney failure caused by reactive Amyloidosis (AA) and multiple rapidly evolving cardiac disease, especially constrictive pericarditis.

Case presentation

One yes ago, a 61-years- old Caucasian man with moderate renal failure and arthritis admitted at hospital for dyspnea, fluid retention and rapid deterioration of his renal function; a renal reactive Amyloidosis (AA) was diagnosed. During hospitalization an echocardiogram showed a normal biventricular function with mild left ventricular hypertrophy, mild aortic stenosis, mild mitral regurgitation and mild elevated filling pressure.

One year after, the patient admitted at the hospital for worsening dyspnea and diuresis contraction: his renal function was further deteriorating with a need for dialysis. His blood pressure was always low (about 80-70/40 mmHg) but his was asymptomatic for dizziness.

At the blood exams serum phosphate and corrected calcium values were respectively 7 mg/dL (n.v. 2,5-4,5 mg/dL) and 9,1 mg/dL (n.v. 8,8-10 mg/dL).

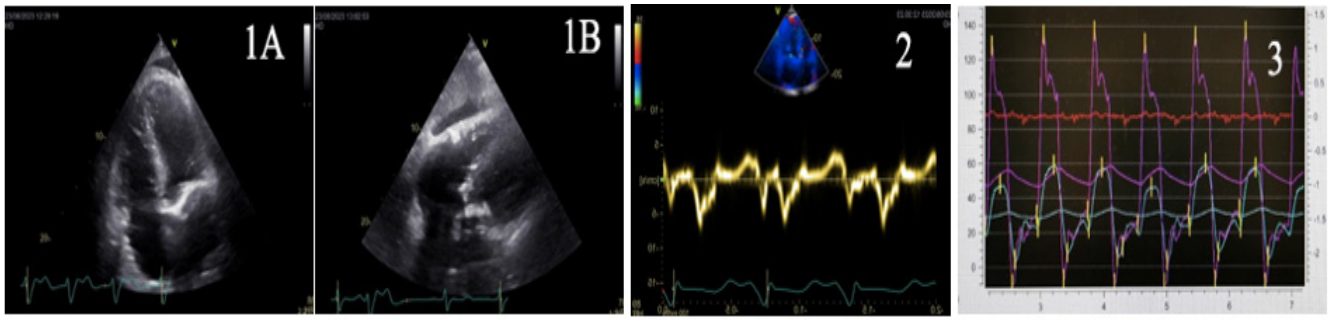

Figure 1: (A-B): Pericardial thickening, pericardial effusion; aortic and mitral calcification.

Figure 2: Medial mitral annulus tissue-Doppler, elevated early diastolic velocities (e').

Figure 3: Dip and plateau pressure trace in constrictive pericarditis.

The electrocardiogram showed sinus rhythm, low ventricular voltage.

The transthoracic echocardiograpy showed a normal left ventricle diameter and function, important aortic valve calcification with moderate stenosis (planimetric area 1.3 cmq, area with equation of continuity 1.05 cmq, maximum gradient 34 mmHg, mean gradient 20 mmHg, stroke volume 68 ml, 37 ml/m^2, CO 12 l/min, CO indicizzato 7 l/min/m^2), severe aortic regurgitation (half pressure time 170 msec), calcific mitral valvular apparatus with severe regurgitation (regurgitan orifice area 0.5 cmq), and mild stenosis (mean gradient 6 mmHg), normal right ventricle diameter but reduced function (TAPSE 11 mm; fractional area change 18%) and hyperecogenic free wall; severe tricuspid regurgitation and severe pulmonary hypertension (70 mmHg) were also present (Figure 1).

Mitral inflow restrictive pattern with respiratory variation and normal e' septal velocity and hyperechogenic pericardial have caught our attention: we suspected a constrictive pericarditis (Figure 2).

The chest computer tomography scan confirmed the pleuric and pericardial calcification while a trans-esofageal echocardiogram the valvulopaties.

The right and left catheterization support the diagnosis of constrictive pericarditis: Dip-and-plateau pattern (or square root sing) of right ventricular pressure with equalization of diastolic pressures (LV diastolic pressure 1 mmHg, RV diastolic pressure 5 mmHg) and remarkable ventricular interdependence were present (Figure 3). Coronarography showed a critical lesion at proximal left anterior descending artery.

The procedure was complicated by resuscitated hyperkinetic cardiac arrest.

Given the prohibitive risk, cardiac surgeons did not declare eligible for surgical solution of heart diseases.

A few days later, after dialysis, the patient died for cardiac arrest due to ventricular fibrillation.

Discussion

The particularity of this case is due to the rapid evolution of heart diseases.

One year before, the patient had mild valvular defects and not mention of pericardial calcification.

The systemic inflammation due to AA amyloidosis and the electrolyte abnormality caused by advanced renal failure have probably played a key role.

Reactive amyloidosis, also known as systemic amyloidosis or secondary amyloidosis, is a disorder which generally occurs as a complication of chronic inflammatory disease.

The protein that forms the fibrils in amyloid deposits is derived from serum amyloid A, a serum acute phase protein. It produced by hepatocytes, play a role in immune regulation. It stimulates pro-inflammatory cytokines, such as TNF, IL-1 and IL-6 [1].

This patient suffered of hyperphosphatemia poorly controlled by the therapy.

Elevated blood values of phosphate were frequently secondary to end stage renal function.

Hyperphosphatemia has been described as a key factor in vascular and other extra skeletal calcification, and also as a risk factor for cardiovascular mortality [2-5].

The exact and complex mechanism by which phosphate collaborates in the formation of extraskeletal calcifications is not yet fully understood and is under study [6].

A crucial event may be a phenotype switch of vascular smooth muscle cells to osteoblast-like cells which is induced through a number of different stimuli like hyperphosphatemia and hypercalcemia.

Pro-inflammatory cytokines that are increased in chronic kidney disease and in reactive amyloidosis have pro-osteogenic potential.

However, even if vascular and aortic calcification is relatively common in patient with ESRD [7-10], this is the first case of rapid progression of multiple calcific valvulopathy, constrictive pericarditis and coronaropathy.

Conclusion

Constrictive pericarditis, although rare, can be a complication of ESRD especially in patients undergoing dialysis treatment. This condition may be due to multiple mechanisms that are not always easy to understand, such as hyperphosphatemia and hypercalcaemia or as a complication of uremic or dialytic pericarditis. A systemic inflammation state can promote this phenomenon that can be very quick.

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