An episode of paroxysmal atrial fibrillation with unexpected ending

Nieoczekiwane rozpoznanie ustalone u chorego z napadem migotania przedsionków

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INTRODUCTION

Light-chain deposition disease (LCDD) is characterized by deposition of immunoglobulin light chains (LCs) in multiple organs. The most common manifestation is kidney involvement, including proteinuria and renal failure [1]. Heart involvement is one of the extra-renal manifestations [2]. Described cardiac manifestations are i.e. atrial fibrillation and restrictive cardiomyopathy [3, 4]. We present a case in which paroxysmal atrial fibrillation (PAF) and previous diagnosis of hypertrophic cardiomyopathy (HCM) conduced to diagnosis of LCDD.

CASE REPORT

52-year-old Caucasian man was admitted to our Cardiology Department with yet another episode of PAF, hemodynamically unstable, with rapid ventricular rhythm. The history revealed several episodes of PAF, treated with electric cardioversion 39 times so far, and twice with radiofrequency ablation (RFA), ineffectively. At admission he underwent fortieth electric cardioversion. Apart from supraventricular arrhythmia the patient had also a history of HCM without left ventricular outflow tract obstruction and with ventricular arrhythmia. Due to this condition, he underwent cardioverter-defibrillator implantation a few years earlier, as a primary prevention of sudden cardiac death. Patient's mother was also reported to have HCM.

Considering previous medical history, a Maze procedure was proposed. As a candidate for Maze surgery, the patient underwent several diagnostic procedures. Echocardiography, an important one among them, showed: left atrial chamber enlargement, normal sizes of other chambers, ventricular septum thickness of 2 cm and the rest of left ventricular wall moderately hypertrophied.

As the echocardiography’s results were not fully specific of HCM (more typical of restrictive cardiomyopathy than HCM) and considering the family history of HCM, further investigations were planned. Despite already established diagnosis of HCM, we wanted to exclude other causes of heart muscle hypertrophy i.e. amyloidosis, that would change patient prognosis and influence further treatment choices. Blood samples for free light chains were obtained. Results revealed light chains lambda of 15,4 and light chains kappa of 23,2 (slightly elevated). Lambda to kappa ratio (L/K) was within normal range.
That spoke in favor of cardiac amyloidosis instead of HCM diagnosis.

Couple of weeks later patient underwent uneventful Maze procedure, during which endomyocardial left ventricle biopsy was obtained. The histopathological examination did not reveal signs of amyloid depositions (no staining with Congo red), what resulted in final diagnosis of observation towards LCDD.

DISCUSSION

Atrial fibrillation is the most common sustained cardiac arrhythmia in clinical practice [5], often associated with underlying heart disease [6]. Hypertrophic cardiomyopathy is diagnosed on the basis of otherwise unexplained hypertrophied, nondilated left ventricle. A septal wall thickness ≥ 1,5 cm has been commonly used to diagnose HCM [7]. Restrictive cardiomyopathy is characterized by nondilated ventricles with impaired ventricular filling [8]. The echocardiographic findings in our patient were ambiguous, as the septal thickness of 2 cm would support the diagnosis of HCM. However, the left atrial enlargement was more specific of restrictive cardiomyopathy. Although typically absent in restrictive cardiomyopathy, left ventricular hypertrophy may be triggered by infiltrative diseases like amyloidosis or LCDD [9]. Kidney involvement usually dominates the clinical course of LCDD, though LCs may be deposited in other internal organs, including liver and heart. The clinical manifestations depend on which tissues are involved and the consequent organ dysfunction. The clinical picture is therefore extremely heterogeneous [10]. Tissue deposits in LCDD are usually composed of kappa LCs [1], resulting in low L/K. In this case, the normal L/K increases suspicion of cardiac amyloidosis. However the deposits do not stain with Congo red, which is the pathognomonic feature of amyloidosis [8].

CONCLUSION

In conclusion, this case suggests that PAF connected with restrictive cardiomyopathy may be related to LCDD. This disease should be taken into consideration in patients with suspicion of restrictive cardiomyopathy and cardiac arrhythmias resistant to RFA. Especially the mild to moderate left ventricular hypertrophy may be the sign of infiltrative disease of the heart rather than HCM, as shown in this case.

ABSTRACT

We present a case of 52-year-old male with restrictive cardiomyopathy as well as with history of paroxysmal atrial fibrillation in whom detailed diagnostic process led to suspicion of light-chain deposition disease as a reason for left ventricular hypertrophy.

Key words: light-chain deposition disease, restrictive cardiomyopathy, Maze procedure

STRESZCZENIE

Prezentujemy przypadek 52-letniego pacjenta z kardiomiopatią restrykcyjną oraz z wywiadem napadowego migotania przedsionków, u którego w toku szczegółowej diagnostyki wysunięto podejrzenie choroby łańcuchów lekkich jako potencjalnej przyczyny przestoru mięśnia lewej komory.

Słowa kluczowe: choroba łańcuchów lekkich, kardiomiopatia restrykcyjna, procedura Maze
References:


