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Case report

Conduction Disorders in a Patient with Transthyretin Amyloidosis: A Case Report

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Abstract Amyloid cardiomyopathy is an important and often unrecognized cause of heart failure and rhythm disturbances leading to syncope or presyncope, especially among the elderly population. For wtATTR amyloid cardiomyopathy, bradyarrhythmia is common, more often due to conduction disorders than sinus node disease itself, which may require the implantation of a permanent antibradycardia pacemaker. This paper presents 75-years old male, with the symptomatic second-degree AV block with a ventricular rate of 35/min, therefore indicating the need for the implantation of a permanent anti-bradycardia pacemaker. Pacemaker implantation can improve symptoms and prevent sudden death caused by severe bradycardia, providing an opportunity for optimal medical therapy of tachyarrhythmia, but not reducing mortality rate of cardiac amyloidosis.

Key-words Transthyretin Amyloidosis, Conduction disorders, Treatment, Prognosis

Introduction

Cardiac amyloidosis is a disorder caused by the build-up of pathological amyloid fibrils in the extracellular space of the myocardium, including the conduction system[1]. Amyloid cardiomyopathy is an important and often unrecognized cause of heart failure and rhythm disturbances leading to syncope or presyncope, especially among the elderly population. Patients with wild-type transthyretin amyloidosis (wtATTR) are most often 60-y-o plus men. In addition to cardiomyopathy, ATTR amyloidosis can also be characterized by peripheral neuropathy, orthostatic hypotension, carpal tunnel syndrome, spinal canal stenosis, and gastrointestinal disorders[2]. Raising clinical suspicion is an important step in making a diagnosis.

Case presentation

A 75-year-old patient with known coronary artery disease was hospitalized for the first time at the Cardiology Department of the Clinical Hospital Centre Zemun due to anginal complaints. He also suffered from chronic kidney disease, arterial hypertension, hyperlipidemia and spinal canal stenosis. Laboratory test revealed persistently slightly elevated value of high-sensitivity troponin, as well as slightly elevated values of natriuretic peptides. Echocardiographic examination showed concentric hypertrophy of the left ventricle, with markedly increased indexed myocardial mass, presence of a discrete difference in regional contractility, moderately reduced systolic function and reduced global longitudinal strain of the left ventricle, with relative sparing of the apex (characteristic

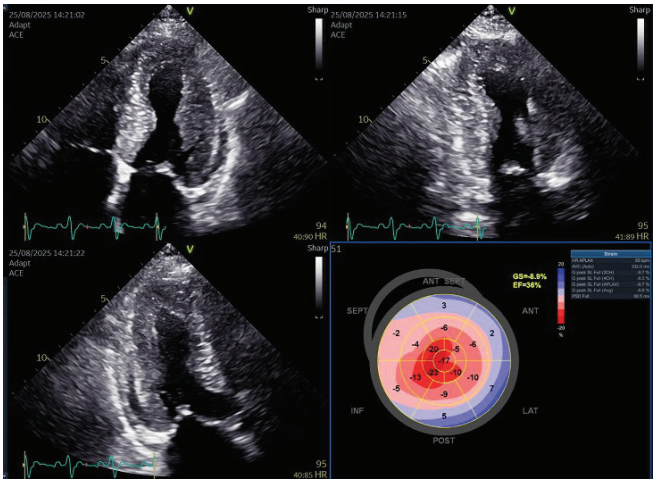


Figure 1. Eho images of patient with cardiac amyloidosis with thypical cherry on the top echocardiographic sign

"cherry-on-the-top sign", see Figure). The typical echocardiographic pattern, raised suspicion of amyloidosis, later confirmed by scintigraphy (Perugini score 3), while genetic testing ruled out the hereditary form of transthyretin amyloidosis. During the outpatient check-up after 3 months, the patient complained of frequent episodes of dizziness, instability, and weakness, along with slow pulse and low blood pressure. Tachycardia-bradycardia syndrome was documented during the 24-hour Holter ECG monitoring: atrial flutter with multiple episodes of slow ventricular response. The beta blocker was discontinued and amiodarone introduced, but without significant improvement. A re-introduction attempt of the lowest beta-blocker dose resulted the same day in symptomatic second-degree AV block developed with a

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ventricular rate of 35/min, therefore indicating the implantation of a permanent anti-bradycardia pacemaker.

Discussion

Reduced voltage of the QRS complex, disproportionate to the left ventricle myocardial mass, pseudo-infarct Q-waves, slow progression of the R-wave, atrial fibrillation, conduction disorders, bundle branch blocks, and prolonged QT interval are frequently observed in patients with cardiac amyloidosis[3]. For wtATTR amyloid cardiomyopathy, bradyarrhythmia is common, more often due to conduction disorders than sinus node disease itself, which may require the implantation of a permanent antibradycardia pacemaker. Approximately 10% of patients with cardiac amyloidosis already have a permanent pacemaker at the time of diagnosis[4]. Pacemaker implantation can improve symptoms and prevent sudden death caused by severe bradycardia, providing an opportunity for optimal medical therapy of tachyarrhythmia, but not reducing mortality rate of cardiac amyloidosis.

Conclusion

The incidence and prevalence of conduction disturbances are high in wtATTR cardiac amyloidosis, and those patients require careful heart rhythm monitoring and close clinical follow-up. On the other hand, in patients with conduction disorders, the existence of cardiac amyloidosis should be considered if there are characteristic clinical, laboratory or imaging parameters that may indicate amyloidosis. In addition to the implantation of a pacemaker, today patients are also treated with specific therapies that can improve their otherwise poor prognosis.

Echocardiography showing hypertrophy of the left ventricle, thickening of the valves, and significantly reduced global longitudinal strain values with relative sparing of the heart apex.

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Sažetak

Poremećaji u sprovođenju kod pacijenta sa transtiretinskom amiloidozom: prikaz slučaja

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Amiloidna kardiomiopatija je važan i često neprepoznat uzrok srčane slabosti i poremećaja ritma, koji dovode do sinkope ili presinkope, naročito u starijoj populaciji. Za wtATTR amiloidnu kardiomiopatiju karakteristične su bradiaritmije, češće kao posledica poremećaja sprovođenja nego bolest samog sinusnog čvora, koje mogu zahtevati implantaciju trajnog antibradikardnog pejsmejkera. U ovom radu prikazan je slučaj muškarca starosti 75 godina sa simptomatskim AV blokom II stepena sa komorskom frekevncom 35/min, i potrebom za implantacijom trajnog antibradikardnog pejsmejkera. Implantacija pejsmejkera može popraviti simptome i prevenirati naprasnu smrt uzrokovanu teškom bradikardijom, te daje mogućnost za optimalnu medikamentnu terapiju tahiaritmija, ali ne redukuje mortalitet od kardijalne amiloidoze.

Ključne reči: *Transtiretinska Amiloidoza, Poremećaji sprovođenja, Lečenje, Prognoza*