Amiloidoza srca, jesmo li je zaboravili?

Cardiac amyloidosis, did we forget it?

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Uvod: Amiloidoza je heterogena skupina bolesti koju karakterizira nakupljanje proteinskih depozita. Neki od njih zahvaćaju srce, a među najčešćima su laki lanci IG i transtiretin.[1-3]

Prikaz slučaja: Bolesnica u dobi od 60 godina javlja se u hitnu službu zbog bolova u prsištu u naporu. Od komorbiditeta liječi dislipidemiju. U 12-kanalnom elektrokardiogramu se bilježe negativni T valovi u odvodima V4-V6. RTG-om se kod bolesnice opisuje uvećano srce. Ehokardiografijom se utvrdi očuvana istisna frakcija lijeve klijetke uz zadebljane stijenke klijetki. Koronarografijom se isključi aterosklerotske promjene na epikardijalnim žilama. Daljnjom obradom kod bolesnice verificira se povišeni NT-proBNP od 4055,4 pg/ml uz 24 satnu proteinuriju od 1,52g. Elektroforeza proteina s imunofiksacijom ukazuje na povišene lambda lance. Punkcijom koštane srži verificiraju se umnožene plazma stanice (15%). Kod bolesnice se zbog zahvaćenosti bubrega odlučilo na biopsiju istog uz pozitivan patohistološki nalaz amiloidoze s lambda lancima. Druga bolesnica u dobi od 57 godina javlja se u hitnu službu zbog znakova popuštanja srca. Od komorbiditeta bolesnica navodi Hashimotovu bolest. RTG-om se opisuje miopatsko srce. U EKG-u se bilježe nespecifični poremećaji provođenja. Učinjenom ehokardiografijom se utvrdi blago snižena istisna frakcija lijeve klijetke uz zadebljane stijenke klijetki. Daljnjom obradom kod bolesnice verificira se povišeni NT-proBNP > 8000 pg/ml. Elektroforeza proteina s imunofiksacijom ukazuje na povišene kappa lance. Punkcijom koštane srži verificiraju se umnožene plazma stanice (9%). S obzirom na to kako se bolesnica prezentira s kožnim lezijama lica uzeta je biopsija istih te je kod bolesnice patohistološki potvrđena dijagnoza amiloidoze s kappa lancima.

Zaključak: Pravovremeno prepoznavanje srčane amiloidoze od izrazite je važnosti za liječenje i prognozu bolesti. Pravovremeno otkrivena bolest omogućava bolesnicima adekvatno liječenje i značajno produljenje života.

Background: Amyloidosis is a heterogeneous group of diseases characterized by the accumulation of protein deposits. Some of them affect the heart, and the most common are the immunoglobulin light chains and transthyretin.[1-3]

Case report: A patient at the age of 60 is admitted to emergency care due to chest pain. She has been taking medicines for dyslipidemia. Chest X ray in ER (emergency room) described a heart enlargement. As a result of the echocardiography, a preserved left ventricular fraction was observed and thickened wall of both ventricles. Coronarography excluded atherosclerotic changes in epicardial vessels. Further treatment of the patient verified an elevated NT-proBNP of 4055.4 pg/ml and a 24-hour proteinuria of 1.52 g. Electrophoresis of proteins and immunofixation indicates elevated lambda light chains. Bone marrow puncture is performed and multiplying plasma cells (15%) were observed. Due to kidney involvement, the kidney biopsy was performed with a positive pathohistological finding of amyloidosis with lambda light chains. The second patient at the age of 57 is admitted to emergency care due to signs of heart failure. He has a history of Hashimoto's disease. Chest X ray in ER has described myopathic heart. 12-lead ECG is characterized by nonspecific conduction disorders. As a result of the echocardiography, slightly lowered left ventricular fraction was observed and thickened walls of both ventricles. Further tests verified an increased NT-proBNP > 8000 pg/ml. Electrophoresis of proteins and immunofixation indicates elevated kappa light chains. Bone marrow puncture is performed and multiplied plasma cells (9%) were observed. Due to skin lesions of the face, the biopsy was performed with a positive pathohistological finding of amyloidosis with kappa light chains.

Conclusion: The timely recognition of cardiac amyloidosis is of extreme importance for the treatment and prognosis of the disease. The timely detection of illness provides patients with adequate treatment and significant life extensions.

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