Amiloidoza srca – možemo li učiniti više?

Cardiac amyloidosis – can we do more?

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KLJUČNE RIJEČI: amiloidoza, transplantacija srca, srčano popuštanje.

KEYWORDS: amyloidosis, heart transplantation, heart failure.

CITATION: Cardiol Croat. 2016;11(10-11):534. | **DOI:** http://dx.doi.org/10.15836/ccar2016.534

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Sistemska amiloidoza je rijetka bolest uzrokovana izvanstaničnim nakupljanjem proteina- amiloida u različitim organima i tkivima, najčešće srcu i bubrezima. Zahvaćanje srca je povezano s povećanim mortalitetom i morbiditetom, posebice u amiloidozi lakih lanaca (AL). Liječenje amiloidoze srca je dvostruko, usmjereno na osnovnu bolest te na srčano zatajivanje i moguće aritmije.[1-3]

Predstavljamo slučaj 61-godišnjeg muškarca s multiplim mijelomom lambda lakih lanaca te amiloidozom srca i bubrega. Pacijent je nakon VAD kemoterapijskog protokola postigao hematološku remisiju, ali s obzirom na amiloidozu srca III stupnja po Mayo klasifikaciji nije bio pogodan za autolognu transplantaciju perifernih matičnih stanica. Razmatrana je mogućnost terapije transplantacijom srca uz naknadnu transplantaciju matičnih stanica što se opisuje u literaturi kao dobro rješenje kod uznapredovale amiloidoze srca u odabranim slučajevima, ali pacijent nije bio prihvaćen za navedeni zahvat budući da nije zadovoljavao kriterije prema postojećim smjernicama. Primjenom uglavnom empirijske medikamentne terapije za liječenje restrikcijske kardiomiopatije i kroničnog kardiorenalnog sindroma te uz terapiju mnogobrojnih infektivnih komplikacija postignuto je preživljenje dulje od 2 godine, što je obzirom na stadij bolesti relativno dobar rezultat.

AL-amiloidoza srca je vrlo rijetka bolest koja se često dijagnosticira kasno. Kontrola klona plazma stanica koji producira lake lance predstavlja temelj terapije, dok je kod pogodnih bolesnika terapija izbora autologna transplantacija perifernih matičnih stanica (ASCT). Iako sporna, transplantacija srca s naknadnom ASCT je jedina opcija za poboljšanje prognoze kod mlađih, pogodnih bolesnika s teškim oblikom AL-amiloidoze srca. Terapija usmjerena na depozite amiloida u tkivima za sada je tek u eksperimentalnoj fazi i još nije u kliničkoj primjeni.

Systemic amyloidosis is a rare disease caused by the extracellular deposition of amyloid fibrils in various organs and tissues, most commonly in the heart and kidneys. Cardiac involvement is associated with an increased mortality and morbidity, especially in primary light chain (AL) amyloidosis. The treatment of cardiac amyloidosis is twofold, oriented both on the underlying disease and heart failure with possible arrhythmic events.[1-3]

We present a case of 61-year-old male patient with lambda light chains multiple myeloma, and heart and kidney AL amyloidosis. Our patient achieved hematologic remission after the VAD chemotherapeutic protocol, but because of Mayo III grade heart amyloidosis he was ineligible for autologous transplantation of peripheral blood stem cells (ASCT). We considered the possibility of heart transplantation, followed by ASCT, which has been described in the literature as good solution in selected cases of advanced cardiac amyloidosis, but the patient was not accepted for the procedure because he did not meet the criteria stated in the guidelines. With mostly empirical medicamentous therapy for restrictive cardiomyopathy and chronic cardiorenal syndrome, and treating many infectious complications he achieved survival of more than 2 years, which is a relatively good result, given the stage of the disease.

Cardiac AL amyloidosis is a very rare condition, often diagnosed with delay. The treatment mainstay is to control the plasma cell clone which is producing the light chains, and in suitable patients the autologous transplantation of peripheral blood stem cells represents the treatment of choice. Although controversial, a heart transplantation followed by ASCT is the only option for improving prognosis in younger, eligible patients with severe cardiac AL amyloidosis. The therapy targeting amyloid deposits in tissues is for now only in experimental stages and is not yet applied in clinical practice.

RECEIVED:
September 22, 2016

ACCEPTED:
October 10, 2016

LITERATURE

1. Banypersad SM, Moon JC, Whelan C, Hawkins PN, Wechalekar AD. Updates in cardiac amyloidosis: a review. J Am Heart Assoc. 2012 Apr;1(2):e000364. **DOI:** http://dx.doi.org/10.1161/JAHA.111.000364
2. Falk RH, Alexander KM, Liao R, Dorbala S. AL (Light-Chain) Cardiac Amyloidosis: A Review of Diagnosis and Therapy. J Am Coll Cardiol. 2016;68(12):1323-41. **DOI:** http://dx.doi.org/10.1016/j.jacc.2016.06.053
3. Grogan M, Gertz M, McCurdy A, Roeker L, Kyle R, Kushwaha S, et al. Long term outcomes of cardiac transplant for immunoglobulin light chain amyloidosis: The Mayo Clinic experience. World J Transplant. 2016;6(2):380-8. **DOI:** http://dx.doi.org/10.5500/wjt.v6.i2.380