Uspješna resinkronizacija u bolesnika s lijevom ventrikularnom ne-kompakcijskom kardiomiopatijom: prikaz slučaja

Successful cardiac resynchronization therapy in a young patient with left ventricular non-compaction cardiomyopathy: a case report

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KLJUČNE RIJEČI: lijeva ventrikularna ne-kompakcijska kardiomiopatija, resinkronizacija.

KEYWORDS: left ventricular non-compaction cardiomyopathy, resynchronisation.

CITATION: Cardiol Croat. 2018;13(11-12):357. | https://doi.org/10.15836/ccar2018.357

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Uvod: Spužvasta kardiomiopatija lijeve klijetke (LVNCC) može biti povezana s dilatacijom ili hipertrofijom lijeve klijetke (LV) te sistoličkom i/ili dijastoličkom disfunkcijom.[1] Simptomi i klinička slika insuficijencije srca variraju, a neki bolesnici mogu biti asimptomatski. Bolesnici su izloženi riziku od lijevostranog i/ili desnostranog zatajivanja srca (ZS), embolizacije i malignih aritmija, kada je implantacija uređaja nužna. Jedna studija je pokazala da bolesnici s LVNCC pokazuju bolji odgovor na resinkronizaciju.

Prikaz slučaja: 36-godišnji muškarac, do sada zdrav, primljen je zbog akutnog zatajivanja lijeve klijetke, NYHA III./IV. stupnja. EKG je pokazao blok lijeve grane sa širinom QRS- kompleksa od 168 ms. Ehokardiografija pri prijemu otkrila je proširen LV (71 mm) s procijenjenom ejekcijskom frakcijom (EF) od 26%, umjerenom mitralnom regurgitacijom, umjerenom dilatacijom lijevog atrija, umjerenom plućnom hipertenzijom s neizravnim znakovima očuvane desne ventrikularne funkcije. Uvedena je standardna terapija s diuretikom, ACE inhibitorima, beta-blokatorom, antagonistima mineralokortikoidnih receptora.i profilaktičkom dozom enoksaparina. Nakon početne rekompenzacije dobio je akutnu embolizaciju desne površne femoralne arterije. Izvršena je uspješna embolektomija i uvedene su terapijske doze enoksaparina uz preklapanje s varfarinom. Kontrolna ehokardiografija nije pokazala tromba u LV, koji je detaljnom analizom pokazao hipertrabekulaciju, ali bez jasnih ehokardiografskih kriterija za LVNCC. Varfarin je titriran do ciljne INR vrijednosti, a nakon optimizacije medikamentozne terapije otpušten je u NYHA II. stupnju i predviđen za MSCT koronarnu angiografiju i MR srca. Isključena je ishemijska bolest srca, a MR srca potvrdio je LVNCC s EF od 24%. Indicirana je implantacija CRT-D pod varfarinom u terapijskoj širini. Sve tri elektrode implantirane su s optimalnim vrijednostima osjetljivosti i stimulacije, a tijekom resinkronizacije imao je visoki R val u V1 i V2 odvodu, s ehokardiografskim znakovima "super-respondera". Nakon 9 mjeseci praćenja, bio je u NYHA I. stupnju bez diuretika Henleove petlje, sa 100% uspješnom biventrikularnom stimulacijom te bez ventrikularne ektopije nakon uvođenja sakubitril/valsartana.

Zaključak: Pacijenti s LVNCC i odgovarajućim kriterijima potencijalni su "super-responderi" na resinkronizaciju s optimalnom remodelacijom lijeve klijetke.

Background: Left ventricular non-compaction cardiomyopathy (LVNCC) can be associated with left ventricle (LV) dilation or hypertrophy, systolic or diastolic dysfunction, or both.[1] Heart failure (HF) symptoms and presentations vary, and some patients may be asymptomatic. Affected individuals are at risk of left and/or right ventricular HF, embolization and malignant arrhythmias when device implantation is necessary. One study showed that LVNCC patients showed better response to resynchronization therapy.

Case presentation: 36-year-old previously healthy male was admitted because of acute left ventricular failure in NYHA III/IV class. ECG showed left bundle branch block with QRS duration of 168 ms. Bedside echocardiography revealed a dilated LV (71 mm in end-diastole) with estimated ejection fraction (EF) of 26%, moderate mitral regurgitation, moderate left atrial enlargement, moderate pulmonary hypertension with indirect signs of preserved right ventricular function. Standard therapy with diuretics, ACE inhibitors, beta-blocker, mineralocorticoid receptor antagonist and prophylactic dose of enoxaparine was introduced. After initial recompenzation he suffered from an acute embolization of the right superficial femoral artery. Successful embolectomy was performed and therapeutic doses of enoxaparine were introduced with warfarin overlap. Control echocardiography revealed no residual thrombi in the LV, that showed trabeculization, but without clear echocardiographic criteria for LVNCC. Warfarin was titrated to optimal INR value, and after optimization of medical therapy he was discharged in NYHA II class and scheduled for MSCT coronary angiography and cardiac MR. Ischemic heart disease was excluded and cardiac MR confirmed LVNCC with EF of 24%. He was scheduled for CRT-D implantation under warfarin in therapeutic range. All three electrodes were implanted with optimal sensing and stimulation values and during resynchronization he had high R wave in V1 and V2 lead, with echocardiographic signs of a super-responder. After 9 months of follow-up he was in NYHA I class without loop-diuretics, and with 100% successful biventricular pacing and no ventricular ectopy detected after introduction of sacubitril/valsartan.

Conclusion: Patients with LVNCC and appropriate criteria appear to be great responders to resynchronization with chances of optimal reverse remodeling.

RECEIVED:
October 25, 2018

ACCEPTED:
November 5, 2018

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