Pridruženi učinak spužvaste kardiomiopatije lijeve klijetke i celijakije na razvoj tromboembolije – prikaz slučaja

Combined effect of left ventricular noncompaction cardiomyopathy and celiac disease on the development of thromboembolic events – case report

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Uvod: Spužvasta kardiomiopatija lijevog ventrikula (LV) je kardiomiopatija morfološki karakterizirana ventrikularnim trabekulima, dubokim intratrabekularnim urezima i tankim slojem kompaktnog epikarda. Etiologija poremećaja u potpunosti nije jasna te bolest može nastati tijekom embrionalnog razvoja srca između 5. i 8. tjedna trudnoće ili sporadično tijekom kasnijeg života, najčešće kod sportaša, pacijenata s anemijom srpastih stanica i tijekom trudnoće. Kliničke manifestacije bolesti uključuju: zatajivanje srca, aritmije (atrijske i ventrikularne) i tromboemboliju (21-24%). Celijakija je imunološki posredovana bolest koja zahvaća sluznicu tankog crijeva uzrokujući upalu, hiperplaziju kripti i atrofijsku vilusa, što dovodi do malapsorpcije različitih nutrijenata. Uz gastroenterološke i hematološke simptome, utjecaj celijakije na razvoj kardiovaskularnih bolesti (dilatacijska kardiomiopatija, ishemijska bolest srca, fibrilacija atrija, tromboembolija) je predmet istraživanja s obzirom na procijenjenu incidenciju bolesti od 1% stanovništva u Europi i Sjevernoj Americi.¹⁻³

Prikaz slučaja: Prikazujemo slučaj pacijenta (rekreativni sportaš) u dobi od 34 godine, kojemu je tijekom hospitalizacije verificirana spužvasta kardiomiopatija LV i celijakija. Pacijent je zbog kronične kritične ishemije lijeve noge na bazi tromboembolije (okluzija površinske femoralne i poplitealne arterije) podvrgnut dva puta "limb saving" operativnom zahvatu, međutim dolazi do progresije infekcije i gangrene lijevog stopala, što u konačnici rezultira potkoljeničnom amputacijom. Diferencijalno dijagnostičkom obradom potvrdili smo spužvastu kardiomiopatiju LV (NC/C 2.7 u apikalnom dijelu) i celijakiju (EMA pozitivna; tTG > 200 H RU/ml). S obzirom da obje bolesti različitim mehanizmima mogu dovesti do daljnjeg slabljenja srčane funkcije, nužna je primjena bezglutenske prehrane, redovita kardiološka reevaluacija i adekvatna fizikalna rehabilitacija.

Introduction: Left ventricular noncompaction is a rare cardiomyopathy morphologically characterized by left ventricular (LV) trabeculae, deep intertrabecular recesses and a thin layer of compacted epicardium. The etiology of the disorder is still not clear, and the disease can develop during the embryonic phase of heart development, between 5-8 weeks of pregnancy, or sporadically in adults mostly in athletes, patient with sickle cell anemia and during pregnancy. Clinical manifestations of the disease are: heart failure, heart arrhythmias (atrial and ventricular) and thromboembolic events (21-24%). Celiac disease is an autoimmune disorder that affects the intestinal mucosa causing inflammation, crypt hyperplasia and villous atrophy, which leads to the malnutrition of different nutrients. In addition to hematological and gastroenterological symptoms, celiac disease can affect the cardiovascular system causing dilated cardiomyopathy, ischemic heart disease, atrial fibrillation and thromboembolic events. Due to the significant incidence of disease of 1% in Europe and North America and the increasing incidence of cardiovascular disease, the effect of celiac disease on the cardiovascular system is the subject of much current research.¹⁻³

Case report: We present a patient (recreational athlete), 34 years old, admitted to University Hospital Center Zagreb, Croatia whom we diagnosed as having noncompaction LV and celiac disease. Due to the critical chronic ischemia of the left leg caused by a thromboembolic event (occlusion of femoral and popliteal artery) limb saving procedures were performed twice, but because of infection and eventual gangrene in the left foot, lower limb amputation was indicated. During the episode of care patient was diagnosed with noncompaction LV (MR imaging: NC/C 2.7 LV apical part) and celiac disease (EMA positive; tTG > 200 H RU/ml, DNA analyses homozygous C677T and heterozygous A1298C). As both diseases, acting in concert can, by different pathogenesis, cause further deterioration of the heart function, condition management protocols must include a gluten free diet, regular cardiac function staging and adequate rehabilitation (use of prostheses depending on heart function).

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LITERATURE

1. Arbustini E, Weidemann F, Hall JL. Left ventricular noncompaction: a distinct cardiomyopathy or a trait shared by different cardiac diseases? J Am Coll Cardiol. 2014;64(17):1840-50. DOI: <http://dx.doi.org/10.1016/j.jacc.2014.08.030>
2. Udeoji DU, Philip KJ, Morrissey RP, Phan A, Schwarz ER. Left ventricular noncompaction cardiomyopathy: updated review. Ther Adv Cardiovasc Dis. 2013;7(5):260-73. DOI: <http://dx.doi.org/10.1177/1753944713504639>
3. Ikeda U, Minamisawa M, Koyama J. Isolated left ventricular noncompaction cardiomyopathy in adults. J Cardiol. 2015;65(2):91-7. DOI: <http://dx.doi.org/10.1016/j.jjcc.2014.10.005>