Neočekivano ili možda (ipak) ne: prikaz slučaja

Unexpected or maybe not: a case report

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Spužvasta „noncompaction" kardiomiopatija lijeve kljetke je rijetka kongenitalna kardiomiopatija. Razlikujemo izoliranu formu u odraslih ili neonatalnu formu bolesti koja uzrokovana mutacijama gena na X kromozomu i koja je udružena s drugim kongenitalnim kardiomiopatijama i neuromuskularnim bolestima Izolirana noncompaction kardiomiopatija (INMV) prvi put opisana od Chin *i sur.* 1990 g, karakterizirana je prisustvom morfoloških karakteristika spužvaste kardiomiopatije bez prisustva drugih srčanih malformacija. Kod takvih slučajeva duboki recesusi komuniciraju sa šupljinom ventrikula, ali ne i sa koronarnom cirkulacijom za razliku od slučajeva neizolirane neonatalne spužvaste kardiomiopatije gdje takva komunikacija postoji. Tijekom 5 do 8 tjedna embrionalnog razvoja ventikularni spužvasti miokard transformira se u kompaktni, i to od epikarda prema endokardu i od baze prema apeksu klijetke. Ovaj proces istovremeno prati i razvoj koronarne cirkulacije, pri čemu se intertrabekularni recesusi smanjuju i tvore kapilare. Koronarne arterijske fistule (CAFs) predstavljaju abnormalnu komunikaciju između koronarnih krvnih žila i drugih krvnih žila ili šupljine klijetki. Kod 20% bolesnika s CAF zabilježene su druge srčane malformacije, najčešće aortalna i pulmonalna atrezija i otvoreni ductus arteriosus. Fistule između koronarnih arterija i srčanih šupljina su rijetka stanja zastupljena u oko 0,2% pacijenata podvrgnutih koronarografiji. Također vrlo rijetko su opisani nalazi postojanja nekompaktne kardiomiopatije u kombinaciji sa koronaroventrikularnim fistulama.[1-3] Prikazujemo 53-godišnju bolesnicu s INMV i koronarografski prikazanim multiplim koronarno ventrikularnim fistulama. Iako je očuvana ejekcijska frakcija lijeve klijetke, funkcionalno oštećenje potvrđuje se deformacijskim slikovnim metodama (*speckle tracking*).

Left ventricular noncompaction is a rare congenital cardiomyopathy. It can exist in isolated form (INVM) in adults or neonatal form which is caused by mutations gene located on the X chromosome and associated with other congenital cardiac and neuromuscular disease. Isolated noncompaction of the ventricular myocardium (INMV), first described by Chin *et al* in 1990, is characterized by persistent embryonic myocardial morphology without other cardiac anomalies. In such cases, deep recesses communicate only with the ventricular cavity, not the coronary circulation, whereas in non-compaction associated with other congenital heart disease (non-isolated non-compaction), the intertrabecular recesses communicate both with the left ventricular cavity and the coronary circulation. During embryonic weeks 5 and 8, the ventricular myocardium transforms from a hypertrabeculated morphology to a compacted layer, and this process is concomitant with coronary artery development. Myocardial remodeling proceeding from the epicardium to endocardium and from the base of the heart to the apex. The coronary circulation develops concurrently during this process, and the intertrabecular recesses are reduced to capillaries. Congenital coronary artery fistulas (CAFs) are abnormal communications between a coronary artery and any cardiac cavity or great vessel. Approximately 20% of patients with coronary artery fistulae have other cardiac anomalies, most frequently aortic and pulmonary atresia and patent ductus arteriosus. Coronary artery fistulae between a coronary artery and a cardiac chamber is a rare condition and is found in approximately 0.2% of patients undergoing cardiac catheterization. Non-compaction ventricular myocardium (NVM) in combination with multiple coronary artery to ventricle fistulae are rare cardiovascular malformations.[1-3] We present 53-year-old female patient with INMV, with preserved ejection fraction and functional impairment of left ventricle proved with deformation imaging methods (speckle-tracking echocardiography), and existence of multiple coronary to left ventricle fistulae.

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