Transplantacija srca kod aritmogene displazije/kardiomiopatije desne klijetke s kasnom prezentacijom – prikaz slučajeva dvoje bolesnika

Cardiac transplantation in arrhythmogenic right ventricular dysplasia/cardiomyopathy with late presentation - two case reports

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Aritmogena displazija/kardiomiopatija desne klijetke (ARVD/C) nasljedna je kardiomiopatija, karakterizirana zamjenom ventrikularnog miokarda masnim i vezivnim tkivom te disfunkcijom desne klijetke, što dovodi do poremećaja ritma srca uključujući supraventrikularne i naročito ventrikularne aritmije.¹⁻³ Česta biva previđena te iznenadna smrt može biti prva manifestacije ove bolesti. Predloženi su standardizirani dijagnostički kriteriji koji se temelje na prisutnosti velikih i malih dijagnostičkih kriterija koji uključuju elektrokardiografske, aritmičke, morfofunkcionalne, histopatološke i genetičke čimbenike.

Prikazujemo slučajeve dvaju pacijenata s vrlo kasnom prezentacijom ARVD/C: jednog 50-godišnjeg muškarca i jedne 49-godišnje žene. Oboje su primljeni u Zavod po dijagnozom terminalnog srčanog zatajivanja (NYHA IV) uslijed dilatativne kardiomiopatije. Klinički i nalazi slikovnih metoda bili su sumnjivi na ARVD/C te iako je u oba pacijenta bila jasna disfunkcija oba ventrikla, ipak su dominirale kliničke slike desnostranog srčanog popuštanja. Isto tako su se oboje prezentirali simptomatskom bradikardijom, uslijed čega je bila potrebna implantacija VVI odnosno CRT-D uređaja. Na kraju su oba bolesnika podvrgnuti uspješnoj transplantaciji srca, dok je klinička sumnja na ARVD/C potvrđena patohistološki.

Neuobičajeno kasna prezentacija ARVD/C-a u ova dva slučaja kao terminalne dilatativne kardiomiopatije sa zahvaćenošću obje klijetke te simptomatskom bradikardijom, u skladu je s njezinom vrlo heterogenom prezentacijom. Uslijed terminalnog srčanog zatajivanja u oba je slučaja bila potrebna transplantacija srca kao krajnja metoda liječenja.

Arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C) is an inherited cardiomyopathy characterized by fibrofatty replacement of the ventricular myocardium, right ventricular (RV) dysfunction, that leads to cardiac rhythm disorders including supraventricular and mostly ventricular arrhythmias.¹⁻³ It is underdiagnosed cardiomyopathy and sudden death may be the first presentation of the disease. Standardized diagnostic criteria have been proposed, based on the presence of major and minor criteria encompassing electrocardiographic, arrhythmic, morphofunctional, histopathologic, and genetic factors.

We present the cases of two patients with a very late presentation of ARVD/C: a 50-year-old man and a 49-year-old woman. They were both referred to department as end stage heart failure (NYHA IV) due to dilated cardiomyopathy. Clinical and imaging findings were suggestive of ARVD/C, and although in the two patients the dysfunction of both ventricles was manifest, signs of right-sided heart failure remained the prevailing finding. Both of them presented also with symptomatic bradycardia as predominant rhythmic disturbance, mandating the implantation of a VVI or a CRT-D device. Ultimately both patients underwent successful heart transplantation, and in both cases the clinical suspicion of ARVD/C with biventricular involvement was confirmed pathohistologically.

The late unusual presentation of ARVD/C in these two cases as end stage dilatative cardiomyopathy with biventricular involvement and bradycardia as predominant rhythmic disturbance is in accordance with its very heterogeneous presentation. In both cases the resulting end stage heart failure required heart transplantation as therapeutic modality.

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