Idiopatska fibrilacija klijetki – prikaz slučaja

Idiopathic ventricular fibrillation – a case report

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Iznanadna srčana smrt (ISS) definira se kao smrt uslijed neočekivanoga krvotočnog urušaja, najčešće zbog srčane aritmije, unutar jednog sata od nastupa simptoma. Iako joj je najčešći uzrok koronarna bolest srca, u mladih ljudi pretežno se radi o kongenitalnim strukturnim ili aritmogenim bolestima. Idiopatska fibrilacija klijetki (IFK) rijedak je uzrok ISS. Njena incidencija nije u potpunosti poznata. Kroz proteklo se vrijeme, sa sve jasnijim definiranjem i dijagnosticiranjem primarnih aritmogenih sindroma (Brugadin sidrom, sindrom dugoga i kratkoga QT intervala, sindrom rane repolarizacije, kateholaminergična polimorfna ventrikularna tahikardia), njena incidencija smanjivala, a definicija se mijenjala. Dijagnoza IFK postavlja se isključenjem poznatih uzroka FK. FK bez dokazane strukturne bolesti srca ili primarnoga aritmijskog sindroma (sa svojim jasnim fenotipskim osobinama) definira se kao IFK. Karakteriziraju je rijetke pojedinačne ventrikularne ekstrasistole (VES) s uskim intervalom vezivanja (R/T fenomen) i visokom sklonosti za razvoj polimorfne ventrikularne tahikardije (pVT) i FK. Stanični mehanizam vezan je uz Ito kalijevu struju u His-Purkinjeovim vlaknima, što uzrokuje snažan repolarizacijski potencijal s okolnim miokardom i dovodi do pojave VES kratkog intervala vezivanja s posljedičnim kružnim gibanjem u fazi II i nastankom pVT i FK. Nije jasno radi li se o monogenetskom ili poligenetskom entitetu, a do sada je opažena upletenost mutacija za gene CALM1, DPP6, RyR i IRX3. Sklonost recidivu je visoka i kreće se (prema do sada rijetkim ispitivanjima) od 11% do 45% godišnje.[1]

U predavanju prikazat ćemo slučaj bolesnice koja je u mlađoj životnoj dobi preživjela aritmijsku oluju i koju pratimo dugi niz godina.

Sudden cardiac death (SCD) is defined as unexpected death due to circulatory shock, mainly because of cardiac arrhythmias, within one hour of symptom onset. Although coronary heart disease is the most common cause, in young people is mainly due to a congenital structural or arrhythmogenic diseases. Idiopathic ventricular fibrillation (IVF) is a rare cause of SCD. Its incidence is not fully known. Over the past time, with a clearer definition of the diagnosis of primary arrhythmogenic syndromes (Brugada syndrome, long and short QT syndrome, early repolarization, catecholaminergic polymorphic ventricular tachycardia), its incidence decreased, and the definition was changed. The diagnosis of IVF is made by exclusion of known causes of VF. VF without proven structural heart disease or primary arrhythmic syndromes (with its clear phenotypic characteristics) is defined as IVF. It is characterized by rare individual ventricular premature beats (VES) with a narrow interval of binding (R / T phenomenon) and a high propensity for the development of polymorphic ventricular tachycardia (PVT) and VF. The cellular mechanism linked to the Ito potassium current in the His-Purkinje fibers, which causes a strong potential repolarization with surrounding myocardium and leads to VES with short coupling interval bonding with subsequent circular movement in phase II and the emergence of PVT and FK. It is not clear whether it is a monogenic or polygenic entity, and so far has seen the involvement of mutations in genes CALM1, DPP6, RyR and IRX3. Relapse rate is high and ranges (according to the now rare trials) of 11% to 45% annually.[1]

In the lecture we will show case of a patient who is at a younger age survived arrhythmic storm and who we follow up for many years.

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LITERATURE

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