Bolesnik sa sinkopom i "skrivenim" dugim QT intervalom

Patient with syncope and "hidden" long QT syndrome

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Uvod: Sindrom dugog QT intervala je obilježen produženjem QT intervala, koji je duži od 480 ms i povišenim rizikom nastanka sinkope i iznenadne srčane smrti zbog polimorfne ventrikulske tahikardije. Genetski poremećaji u srčanim kalijskim, natrijskim ili kalcijskim kanalima su odgovorni za nastanak sindroma.[1]

Prikaz slučaja: Bolesnik star 49 godina, koji je dosad bio zdrav bio je hospitaliziran zbog sinkope u drugoj bolnici. Na dan prijema, osjetio je palpitacije nakon čega je izgubio svijest na 1-2 minute. U inicijalnom EKG-u je zabilježena kratka polimorfna ventrikulska tahikardija (5 QRS-a, frekvencije 250/minuti). Ultrazvuk srca i koronarografija su bili uredni. Tijekom hospitalizacije u jednom navratu je defibriliran zbog nastanka ventrikulske fibrilacije. U terapiju je uveden amiodaron i bolesnik je premješten u Klinički bolnički centar Zagreb radi nastavka liječenja. U EKG-u je zabilježena jedna ventrikulska ekstrasistola (VES) iz izlaznog dijela desne klijetke (RVOT). QTc je bio normalan (433 ms) s naznačenim U valovima, bez elevacija J točke. Učinjena je elektrofiziološka studija, tijekom infuzije izoproterenola, zabilježeno je produženje QTc intervala (555 ms). S obzirom na navedeno, postavljena je dijagnoza dugog QT sindroma. Kombinacija povremenog produženja QTc intervala i VES iz RVOT-a kod ovog je pacijenta izazvala polimorfnu ventrikulsku tahikardiju i srčani arest. Nakon što je učinjen MR srca kojim je isključena strukturna bolest srca, ugrađen je kardioverter defibrilator i bolesnik je otpušten kući s malom doza beta-blokatora. Na kontrolnom pregledu, broj VES je bio >10%, a bolesnik je postao simptomatičan tako da je na naručen na ablaciju.

Zaključak: VES iz RVOT kod bolesnika sa strukturno zdravim srcem se smatraju benignim stanjem, ali kod nekih skupina ljudi mogu biti fatalne, kao što je naš slučaj. Kod većine malignih RVOT VES, neprepoznata aritmogena displazija desnog ventrikula se smatra odgovornom za smrtnost, ali moramo razmotriti i druge mogućnosti. U ovom slučaju ablacija fokusa VES, može značajno smanjiti učestalost malignih aritmija i aktivaciju kardioverter defibrilatora, ali ugradnja kardioverter defibrilatora je ipak obvezna.

Introduction: Long QT syndrome (LQTS) is characterized by prolongation of the QT interval, with a QTc exceeding 480 ms and an increased risk for syncope and sudden cardiac death secondary to polymorphic ventricular tachycardia (VT). Genetic defects in either cardiac potassium, sodium or calcium channels are responsible for this syndrome.[1]

Case report: 49-year-old patient without previous medical history was admitted to another hospital because of the syncope. On the day of admission, he felt palpitations and after which he transiently lost consciousness for 1-2 minutes. Initial ECG strip recorded short polymorphic tachycardia (5 QRS complexes, 250 beats per minute). Echocardiography and coronary angiography were both normal. During the hospitalization, he developed ventricular fibrillation and was successfully defibrillated. Amiodarone was introduced in therapy and he was transferred to our hospital for further workup. 12-lead ECG showed normal sinus rhythm with 1 premature ventricular contraction (PVC) originating from right ventricular outflow tract (RVOT). QTc interval was normal (433 ms) with prominent U waves and no signs of J wave syndromes. On 24-hours Holter monitoring, PVC burden was around 5%. Electrophysiologic study was performed, during isoproterenol infusion, QTc prolongation was observed (maximum QTc 555 ms). Therefore long QT syndrome was diagnosed. Combination of intermittently prolonged QTc interval and RVOT PVCs in this patient provoked polymorphic VT and cardiac arrest. After MRI which showed structurally normal heart, implantable cardioverter defibrilator (ICD) was implanted and the patient was discharged with low a dose of beta-blocker. In the follow-up PVC burden is increased to 10% and the patient became symptomatic, so ablation was scheduled.

Conclusion: Right ventricular outflow tract PVCs in a structurally normal heart is considered a benign condition, but in some patient populations, they can be fatal, as proved in our case. In most cases of malignant right ventricular PVC-s, unrecognized arrhythmogenic right ventricular dysplasia is deemed responsible for mortality but we have to consider other possibilities. In this case, ablation of culprit PVCs can significantly lower the possibility of fatal arrhythmias and ICD discharges, but nevertheless permanent ICD therapy is still mandatory.

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LITERATURE

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