Kazuistika | Case report

Long QT Syndrome and Electrical Storm: Is Implanted Cardiac Defibrillator the Final Destination in Long QT Syndrome Management?

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ARTICLE INFO

Article history:
Submitted: 22. 5. 2020
Revised: 31. 7. 2020
Accepted: 2. 8. 2020
Available online: 22. 10. 2020

Background

Electrical storm is a life-threatening medical emergency and in the context of implantable cardiac defibrillator (ICD) it is defined as three or more appropriate detections of ventricular arrhythmia in prior 24 hours leading to ICD shocks.1,2 Sympathetic stimulation is the forefront player for initiation and maintenance of ventricular arrhythmia especially in long QT (LQT) syndromes.2,3 We are reporting a case of congenital LQT syndrome who underwent ICD implantation and now presented with ICD storm.

Case report

A 17-year-old girl with history of implantable cardiac defibrillator (ICD-VR) implanted for long QT syndrome due to recurrent syncope 10 years ago, she underwent a box replacement for end of life battery 5 years ago, her family history was remarkable for a brother who died suddenly while asleep at age of 13 years. Due to non-availability of the test, she was not genetically classified after being diagnosed of LQT syndrome. The patient was kept on propranolol tab 10 mg thrice daily. Over the last three months she had been admitted frequently for electrical storms. This time again; she was presented with recurrent ICD shocks (more than 6 shocks) over the last 24 hours, all preceded by emotional stress and palpitation, she was admitted to cardiac care unit for further evaluation and management, where electrocardiogram showed long QT interval of 581, as seen in Fig. 1, and ventricular ectopic (PVC), monitor in cardiac care unit showed ventricular tachycardia/ventricular fibrillation episodes treated appropriately by ICD shocks, this was further confirmed by de-
vice programming. During admission; on daily follow-up; the patient was anxious and tearful and more emotional distress was evident with each new ICD shock which further exacerbated her ventricular arrhythmia. Her blood tests including potassium, magnesium, and calcium were all normal, the patient was kept on magnesium and lidocaine infusions in addition to propranolol tab 40 mg thrice daily (considering the important role of beta-blockers in decreasing arrhythmia burden),4 still she had frequent ICD discharges until propranolol increased up to 40 mg four times daily (maximum tolerated dose as further increase caused hypotension), the patient at this dose had no further shocks, magnesium infusion was stopped then lidocaine infusion was also stopped, ultimately patient was on propranolol only with no more ICD shocks.

Considering patient’s impaired quality of life and psychological burden of ICD events both on her and her family and being an adolescent who seeks a normal functioning life and who can face daily stress and challenges especially in her age which might further exacerbate her ventricular arrhythmia, in response; the treating team discussed additional measures that should be undertaken to prevent further episodes of ICD storm especially the episodes were usually triggered by intense emotions, so sympathectomy was recommended according to latest guidelines,5 but unfortunately it was unavailable locally, socioeconomic status of patient deprived her from further attempt to pursue centres abroad for the procedure. Mexiletine also suggested as one of the effective drugs in LQT syndrome which can decrease burden of ventricular arrhythmias, in response; the treating team discussed additional measures that should be undertaken to prevent further episodes of ICD storm especially the episodes were usually triggered by intense emotions, so sympathectomy was recommended according to latest guidelines.6 Despite that genetic testing is the gold standard for subtype recognition, there are certain clinical and electrocardiographic findings that can suggest the subtype in the absence of genotype analysis. For example, the trigger for cardiac event is mainly by exercise, swimming and sympathetic stimulation in LQT1 syndrome, emotional stress or sudden auditory stimuli (like alarm-clock), or less commonly during sleep or exercise, or during the postpartum period in LQT2 syndrome, while cardiac events in LQT3 syndrome usually occur during rest or bradycardia.8–11 ECG also can help in differentiation between LQT subtypes as following:7,9

- **LQT1:** There are broad based, peak T waves producing QT prolongation
- **LQT2:** There are notched or bifid T waves.
- **LQT3:** The late appearance of the T wave after long ST segment will produce long QT interval.

Interestingly, our patient had broad based T wave with generalized T wave inversion and prolonged QT, these changes along with history of arrhythmia triggering by emotional stress can suggest LQT1 syndrome, despite such ECG changes are not classical in long QT syndromes, there is a report of QT syndrome case series12 which described T wave changes like flat T wave, biphasic T wave or deep symmetrical T wave inversions in chest lead V5-V6, however, up to our knowledge, there had been no case report of long QT syndrome with such generalized deep symmetrical T wave inversions involving most ECG leads in the absence of associated cardiomyopathies.

The cornerstone management in high-risk LQT syndrome are beta-blockers and ICD implantation.13 Beta-blockers decreased cardiac events in this population by 50%, they are also effective in decreasing exercise triggered cardiac events,14–17 however, cardiac events can still occur while patients on beta-blockers, particularly in symptomatic individuals as it was reported that 32% of symptomatic persons will have a cardiac event over the next 5 years, and 14% of patients with a prior cardiac arrest will have a recurrence within 5 years.15,18

Another therapeutic option in LQT syndrome is left cardiac sympathetic denervation which is used in refractory ventricular arrhythmias, it is a rarely performed procedure despite effectiveness,19 it can be considered in long QT syndrome patients who suffer from ICD events despite beta-blockers therapy.5,12 Sympathectomy is a treatment modality that can minimize the arrhythmia burden in LQTS patients, but not obliterate it, with low complica-
tion rate.\textsuperscript{20} This option received class I recommendation in cases of recurrent appropriate ICD shocks in long QT syndrome after intensification of medical treatment like beta-blockers or in cases of intolerance to medical therapy.\textsuperscript{5}

With recent progress of mapping techniques and subsequent catheter ablation; there has been broadening in its indications to further include the channelopathies like LQT syndromes, yet it is crucial to notice that ablation of PVC triggers is not curative and not an alternative for ICD insertion in high-risk LQT syndrome cases.\textsuperscript{13}

Current case report highlights other critical point which is the role of psychosocial management and mental health care in young patients with arrhythmogenic syndromes who are treated with ICD, as in emerging countries this service is not provided for those patients despite its necessity especially that patients with arrhythmogenic syndromes are usually young and seek normal quality of life just like their peers. ICD shocks were reported to decrease psychological function and impair quality of life, so ICD benefit in decreasing morbidity can be partially waived with recurrent shocks which can cause depression, anxiety disorders and mood disturbances rendering patients at most need for psychological support.\textsuperscript{21,22}

This case report is a reflection of current status of management of patients with arrhythmogenic syndromes and a call for action to healthcare providers, decision makers and stakeholders to make sympathectomy a feasible option in cardiac facilities and to include psychosocial services in the specialized cardiac centres especially if we consider the high volume of device implantation procedures in young patients that conducted in these centres.

Conclusion

It is of utmost importance that we consider what is the next step after ICD implantation, and we should seriously realize that ICD is not the final destination in managing young population with arrhythmogenic syndromes, it is only the first step of a long journey that we will walk through side by side with our patients, so we need to make it a fruitful prosperous journey with least suffering as possible.

Acknowledgements

None.

Conflicts of interest

None.

Funding

None.

Ethical statement

The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national Iraqi guidelines and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the ethical and scientific committee in Ibn Al-Bitar Cardiac Center. Informed consent to publish the case was obtained from the patient and her parent.

References