Clinical Communication

Congenital long QT syndrome: The masquerader

INTRODUCTION

Long QT syndrome (LQTS) is identified by QT interval prolongation on electrocardiogram (ECG) with a predilection to ventricular tachyarrhythmias. It can either be congenital or acquired. We report a case of congenital LQTS which presented with recurrent ventricular tachyarrhythmias intraoperatively.

CASE DESCRIPTION

A 45-year-old gentleman weighing 65 kg presented to the emergency department with loss of consciousness following trauma and multiple lacerated wounds over the face and scalp. On examination, his blood pressure was 80/50 mmHg and Glasgow Coma Scale score was 14/15. Focused assessment with sonography for trauma was negative. Haemodynamic stability was achieved with fluid resuscitation and whole body computerised tomography showed an undisplaced frontal bone fracture. His initial laboratory results were unremarkable except for serum potassium of 2.6 $mEql^{-1}$, serum glutamic oxaloacetic transaminase of 63 IUl^{-1} and albumin of 2.6 $mgdl^{-1}$.

After potassium supplementation and obtaining a laboratory value of 3.2 $mEql^{-1}$, he was posted for debridement and suturing of the scalp wound. On pre-anaesthesia evaluation, his family revealed a history of chronic alcoholism, diabetes and infrequent episodes of syncope or seizures and the last episode was about six months ago. He was not on any regular medications. In the operating room, after applying the standard monitors, he was premedicated with intravenous (IV) midazolam (1 mg), ondansetron (4 mg), fentanyl (100 μg), preservative free lignocaine (90 mg) and anaesthesia was induced with propofol 140 mg. Endotracheal intubation was done after obtaining neuromuscular blockade with succinylcholine 100 mg IV. Anaesthesia was maintained with 2% sevoflurane in 50% air/oxygen mixture and with intermittent boluses of fentanyl (40 μg) and atracurium (10 mg).

After a few minutes, he had two episodes of torsade de pointes (TdP) lasting for less than 10 seconds. The surgery was stopped, help was called for and a defibrillator was rolled in. Magnesium sulphate 2 g IV was given. On closer scrutiny of records, it was found that he had a corrected QT interval of 538 ms in the ECG which was overlooked. A third episode of TdP occurred which progressed to ventricular fibrillation and was reverted to sinus rhythm with advanced cardiac life support. He received two shocks of 200 joules each and return of spontaneous circulation was obtained in 5 min. He was started on nor-adrenaline infusion. The ECG recorded after the cardiac arrest showed ST segment elevation in inferior and lateral leads [Figure 1]. Echocardiogram (ECHO) showed an ejection fraction of 30% with features suggestive of stress cardiomyopathy. The surgery was halted and the patient was shifted to the intensive care unit (ICU) for elective ventilation and further evaluation. Postoperatively, all investigations were normal except for troponin I (positive) and corrected serum calcium (7.6 $mEql^{-1}$).

In the ICU, within 12 hours, he had two more episodes of ventricular tachyarrhythmia which reverted to organised rhythm following defibrillation. In view of repetitive ventricular tachyarrhythmias, a coronary angiogram was done which revealed normal coronaries. An overdrive pacing with a temporary transvenous pacemaker was done and he was taken up for secondary suturing under general anaesthesia after initiation of beta blockers. Once his haemodynamics improved, he was weaned off from the ventilator. The ECG, after the removal of temporary pacing and with normal serum electrolyte levels, still showed a corrected QT interval of 510 ms. Hence, a diagnosis of congenital long QT syndrome was made and an automated implantable cardioverter defibrillator (ICD) was implanted. A detailed ECHO prior to discharge showed no regional wall motion abnormality and good left ventricular systolic function. He was advised

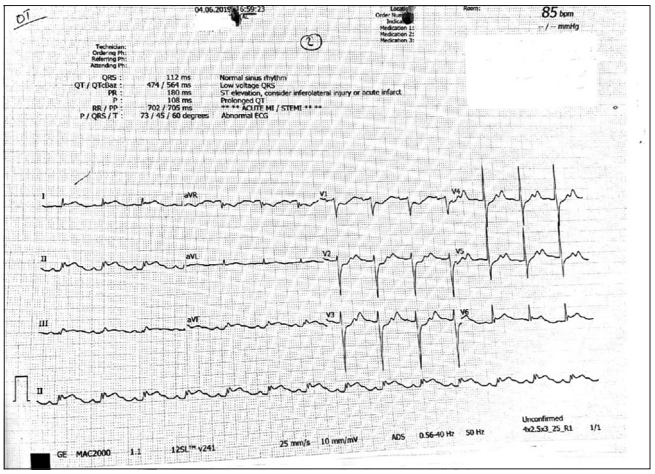

Figure 1: Postoperative ECG shows a prolonged QTc interval

regarding genetic testing and screening of first degree relatives but these recommendations were not followed. He had no further episodes of unconsciousness and is on follow-up every four months.

DISCUSSION

Long QT syndrome can be asymptomatic or can present with various symptoms like palpitations, syncope, seizures or even sudden death. In our patient, the initial investigations showed hypokalemia which itself is a cause of long QT. The diagnosis was missed as no ECG was repeated after correction of serum potassium levels. Also, the history of seizures obtained, might have been due to episodes of ventricular tachyarrhythmia which terminated on their own. The stress of surgery, certain drugs administered in the perioperative period like ondansetron, succinylcholine, sevoflurane, etc., and even the intra-hospital transfer of the patient might have triggered the multiple episodes of subsequent ventricular tachyarrhythmias.

Congenital LQTS is an inherited channelopathy characterised by prolonged QT interval and polymorphic ventricular tachycardia. Schwartz risk score is used for diagnostic evaluation of congenital LQTS and a score of >3.5 is considered as significant.[1,2] The score of our patient was more than 3.5. Factors like exercise, acute emotion or sudden auditory stimuli like an alarm and bradycardia that occur during sleep are known to trigger ventricular arrhythmias in these patients.[3] The management includes beta adrenergic blockade, left cardiac sympathetic denervation, ICD and avoiding triggering factors like exercise, auditory stimuli, etc.[3-5] Shortening the QT interval with overdrive pacing using a transvenous pacemaker decreases the frequency of early after depolarisation type premature ventricular complexes.[3] Before pursuing a diagnosis of congenital long QT syndrome, the causes of acquired QT prolongation must be excluded [Table 1].[2]

Anaesthesiologists must be able to diagnose congenital LQTS and manage the ventricular tachyarrhythmias associated with it. Drugs that cause QT prolongation should be avoided whenever possible.[2,3] Beta blockers should be continued in the perioperative period. Serum levels of potassium, magnesium and calcium should be checked and optimised.[2,3,4] A quiet operating-room equipped with a defibrillator and IV magnesium sulphate is essential before induction of anaesthesia.[2,4] Care should be taken to avoid perioperative pain, hypoxia, hypothermia, hypercarbia, hypovolaemia, hypoglycaemia, raised intrathoracic pressures and electrolyte disturbances.[2,3] The perioperative management of patients with an ICD or a pacemaker should be modified accordingly.[4] Total IV anaesthesia is the ideal anaesthesia technique.[3,4] Among muscle relaxants, succinylcholine and pancuronium need to be avoided.[2,3] Sugammadex can be used but anticholinesterases should be used with caution.[3,4] Regional anaesthesia techniques can be safely used in these patients.[4,5] Dexamethasone, cyclizine, low dose propofol or metoclopramide can be used and ondansetron and droperidol are avoided as antiemetics.[2,4] Postoperatively these patients should be nursed in a quiet environment with adequate monitoring.[2]

Table 1: Causes of acquired LQTS

Factors	Conditions
Metabolic factors	Hypokalaemia, hypomagnesaemia, hypocalcaemia, hypothyroidism, hypothermia
Other clinical factors	Myocardial ischaemia, intracranial disease
Drug class	**Examples**
Anaesthetics and sedatives	Chloral hydrate, ketamine, thiopentone, sevoflurane, desflurane
Antiemetics	Droperidol, ondansetron
Neuromuscular blockers	Succinylcholine, pancuronium
Opioids	Buprenorphine, hydrocodone, methadone, sufentanil
Antianginal drugs	Ivabradine, ranolazine
Antiarrhythmic drugs	Disopyramide, hydroquinidine, procainamide, quinidine, flecainide, propafenone, amiodarone, ibutilide, sotalol
Fluoroquinolones	Ciprofloxacin, gatifloxacin, levofloxacin, moxifloxacin,
Macrolide antibiotics	Azithromycin, clarithromycin, erythromycin, roxithromycin
Azole antifungals	Gluconazole, voriconazole
Antimalarial drugs	Chloroquine, hydroxychloroquine, quinine
Antihistamines	Astemizole, hydroxyzine, terfenadine
Antineoplastic drugs	Aclarubicin, arsenic trioxide, ceritinib, crizotinib, dasatinib, encorafenib, gilteritinib, oxaloplatin
Antipsychotics	Amisulpride, chlorpromazine, thioridazine, clozapine, flupentixol, haloperidol, olanzapine, quetiapine, risperidone
Antidepressants	Citalopram, clomipramine, doxepin, escitalopram, imipramine
Miscellaneous	Mifepristone, oxytocin, terlipressin, organophosphate intoxication

CONCLUSION

Perioperative QT prolongation can be managed by correcting electrolyte disturbances and removing offending medications. Persistence of prolonged QT interval despite these measures warrants a cardiology

evaluation. Measures should be taken to prevent and revert any malignant arrhythmias in the perioperative period.

Declaration of patient consent

The patient's consent was obtained prior to submission of case report.

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Conflicts of interest

There are no conflicts of interest.

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