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Familial long QT syndrome presented as ventricular tachycardia during anesthesia

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Case report

The diagnosis of long QT syndrome (LQTS) is usually made by the presence of QT interval prolongation on electrocardiogram (ECG) obtained during clinical evaluation of syncope episodes or screening of family members of patients known to have the disease(1–3). We describe an adolescent who during a total thyroidectomy had episodes of ventricular tachycardia and was subsequently diagnosed with familial LQTS.

A 13-year-old girl with Graves' disease presented for thyroidectomy. There was no known history of heart disease or syncope. Preoperative medications included potassium iodine, propylthiouracil, and calcitriol. Physical examination revealed a goiter (14 × 4 cm) with palpable thrill and the patient was euthyroid. Anesthesia was induced with propofol and succinylcholine and the trachea was intubated. Anesthesia was maintained with sevoflurane, fentanyl, and cisatracurium. The procedure was remarkable for difficult mobilization of the goiter, estimated blood loss of 1400 ml, and recurrent episodes of ventricular ectopy including multifocal premature ventricular complexes, couplets, triplets, and nonsustained ventricular tachycardia (Figure 1). Intraoperatively, serum level of magnesium was 0.61 mmol·L⁻¹ (normal 0.75–1.0 mmol·L⁻¹), potassium 3.7 mmol·L⁻¹ (3.3–5.1 mmol·L⁻¹), and calcium 1.92 mmol·L⁻¹ (2.05–2.50 mmol·L⁻¹). Intravenous lidocaine, magnesium, and calcium were administered and after correction of serum magnesium and calcium, lidocaine was discontinued, and propranolol was started.

An ECG obtained upon completion of thyroidectomy revealed sinus rhythm, QT interval of 443 ms, QTc of 542 ms, and juvenile pattern of T wave inversion from V1 to V4. An echocardiogram yielded no structural abnormalities. In retrospect, it was discovered that unknown to surgeons and anesthesiologists, 'a routine ECG' had been obtained in the evening prior to surgery. This ECG was reviewed by the cardiologist consulted after the intraoperative episode of ventricular tachycardia and showed sinus rhythm, QT interval of 439 ms, QTc of 494 ms, and T wave inversion from V1–V4. In addition, after further inquiry, the child's parents revealed that the patient had an episode of syncope at the age of seven, and that both mother and maternal grandmother had episodes of syncope. After the patient's diagnosis of LQTS, maternal ECG confirmed prolonged QT interval.

We describe a patient whose diagnosis of familial LQTS was made during the evaluation of ventricular arrhythmias during anesthesia. To our knowledge, this is the first report of familial LQTS diagnosed during clinical evaluation of ventricular arrhythmias observed

during general anesthesia which is unlike the usual presentation of LQTS – syncope episodes associated with QT interval prolongation. What factors might have contributed to the occurrence of ventricular arrhythmias in our patient? It is known that a number of drugs including anti-arrhythmics, antibiotics, and anti-depressants can prolong the QT interval (1). In our patient, the drugs administered preoperatively (potassium iodine, propylthiouracil, and calcitriol) are not known to be associated with prolongation of QT interval. However, it is possible that, thyroid disease, hypomagnesemia, hypocalcemia, and volatile anesthetic agents contributed to the occurrence of ventricular tachycardia in this patient with previously unknown LQTS (1,4,5).

This case illustrates a few points worthy of consideration. The first and most important is that intraoperative ventricular arrhythmias can be the initial presentation of familial LQTS. The second is that a thorough evaluation of intraoperative ventricular arrhythmias should be conducted and such evaluations can yield previously unknown familial cardiac diseases in the pediatric population. The third is that while not current practice in pediatric anesthesia, a preoperative ECG can possibly reveal previously unknown cardiac disease with anesthetic implications.

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Figure 1.
Initial presentation of a patient with familial long QT syndrome: ventricular arrhythmias during anesthesia.