Anaesthetic-induced ventricular tachyarrhythmia in Jervell and Lange-Nielsen syndrome

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A four-year-old deaf girl with a history of convulsions developed polymorphous ventricular tachycardia during induction of anaesthesia. The arrhythmia reverted to sinus rhythm spontaneously. Post-anaesthetic ECG showed marked prolongation of the QTc interval (570–690 msec). Deafness and prolonged QTc interval in association with microcytic-hypochromic anaemia confirmed the diagnosis of the Jervell and Lange-Nielsen syndrome. This case report highlights the potentially lethal complication of halothane anaesthesia in patients with long QTc interval syndrome.

Une fillette âgée de quatre ans atteinte de surdit avec une histoire de convulsions a dvelopp une tachycardie ventriculaire polymorphe lors de l'induction de l'anesthsie. L'arythmie s'est convertie spontanment un rythme sinusal. L'ECG postanesthsique a dmontr une prolongation marque de l'intervalle QTc (570–690 msec). La surdit et la prolongation de l'intervalle QTc en association avec l'anmie hypochromique-microcytaire a confirm le diagnostic du syndrome de Jervell et Lange-Nielsen. Cette histoire de cas met l'emphase sur la complication potentiellement lthale de l'anesthsie l'halothane chez les patients avec un syndrome d'intervalle QTc long.

Key words

BRAIN: convulsions;

COMPLICATIONS: arrhythmia;

HEART: arrhythmia, tachycardia;

SYNDROMES: Jervell Lange-Nielsen.

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The Jervell and Lange-Nielsen syndrome (JLNS) is characterized by congenital deafness, long QTc interval and microcytic hypochromic anaemia. It is an unusual familial cause of syncopal attacks and sudden death during infancy and childhood.¹ It is now recognised that the syncopal attacks in these patients are due to polymorphous ventricular tachycardia.^{2,3} However, these recurrent syncopal attacks in children with JLNS may be misdiagnosed as epilepsy^{4–6} and general anaesthesia may trigger a potentially fatal arrhythmia.^{7–9}

We report a child, previously diagnosed as epileptic, in whom halothane anaesthesia induced polymorphous ventricular tachycardia. This led to the diagnosis of JLNS.

Case report

A four-year-old girl with a history of recurrent epileptiform attacks was scheduled for brain scan under general anaesthesia on an out-patient basis. There was no previous history of anaesthesia. She was deaf and showed no other neurological deficit. Apart from mild microcytic hypochromic anaemia (Hb 9.2 g · dl⁻¹, Hct 29.7%, MCH 19.5 pg · dl⁻¹, MCHC 31%, MCV 62, anisocytosis), all other laboratory investigations were normal. No outpatient pre-anaesthetic ECG was done. Pre-induction ECG appeared normal. An iv infusion of dextrose 0.5% was started and atropine 0.2 mg given iv. There was a moderate sinus tachycardia. Anaesthesia was induced with nitrous oxide in 50% oxygen and halothane given in incremental concentrations up to 4%. At laryngoscopy, and prior to insertion of the endotracheal tube, the ECG showed polymorphous ventricular tachycardia (Figure 1A) which spontaneously reverted to sinus rhythm. A size 4.5 mm endotracheal tube was inserted and anaesthesia was maintained with 1% halothane in 100% oxygen. No convulsion occurred during anaesthesia. The CAT scan revealed no brain pathology. The patient was transferred to the paediatric cardiology clinic where a diagnosis of Jervell and Lange-Nielsen syndrome was made on the basis of clinical features and ECG characteristics (Figure 1B).

FIGURE 1A Anaesthesia-induced arrhythmia. Note run of bizarre-shaped, broad QRS complexes suggesting polymorphous ventricular tachycardia.

Discussion

Our patient was previously misdiagnosed as a case of epilepsy. However, the history of recurrent syncopal attacks in a deaf child should have raised the possibility of long QT interval syndrome (LQTS). A 12-lead ECG would have established the diagnosis. The case supports the contention that LQTS should be considered in the differential diagnosis of seizures, and an ECG should be obtained in these patients.⁴⁻⁶

The role of ventricular arrhythmia as the basis for syncope and sudden death in patients with LQTS is now well recognised. General anaesthesia may trigger ventricular dysrhythmia in patients with prolonged QT interval,⁷⁻⁹ and anaesthetic guidelines have been suggested for these patients.^{10,11} It is recognised that halothane is arrhythmogenic.¹² By contrast, isoflurane increases the ventricular arrhythmia threshold¹³ and hence is the anaesthetic agent of choice for patients with LQT syndrome. Since the rate adjusted QT interval relationship is abnormal, atropinization or excessive catecholamine release may precipitate R-on-T phenomenon with ventricular dysrhythmia.¹⁴ Thus atropine should be avoided and premedication should be given to allay fear and minimize catecholamine release.

Treatment with beta-blocking drugs before general anaesthesia may reduce fatal arrhythmias substantially.¹⁵

In summary, this report highlights the potentially lethal complication of halothane anaesthesia in a patient with undiagnosed Jervell and Lange-Nielsen syndrome, masquerading as epilepsy. It also emphasizes that ECG examination is mandatory for patients who are deaf and present with a history of recurrent seizure disorders.

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FIGURE 1B Resting ECG shows bizarre, inverted T wave and marked prolongation of QTC interval (570-690 msec).

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