EDITORIAL

Genetics and Anaesthesia: Malignant Hyperpyrexia

Dr Shaikha Salim Al Arrayed*

Malignant hyperpyrexia¹ is a potentially fatal complication of general anaesthesia. In the affected individuals Suxamethonium and/or Halothane and other drugs can trigger a sudden rise in body temperature which unless rapidly corrected is followed by convulsion and death in 60% of cases. The reaction does not seem to depend upon any particular anaesthetic agent nor on the type of surgery being performed. It has often developed in previously healthy individuals undergoing relatively minor surgical procedure. The tendency to develop this complication is inherited as an autosomal dominant characteristic.

Soon after induction of anaesthesia the muscles go into massive spasm, the body temperature quickly rises to a high level and the patient becomes acidotic. It is probably the cardiac effect of the acidosis which often causes sudden death.

Malignant hyperpyrexia is accompanied by the following biochemical changes²: serum creatine phosphokinase (CPK) reaches high level within three hours after induction of anaesthesia which is explained by muscle injury and damage; high level of serum glutamic oxalacetic transaminase (SGOT) and lactate dehydrogenase (LDH); high level of serum phosphate which results in a fall of serum calcium; and high levels of serum potassium and serum aldolase.

Denborough et al³ studied the family and three close relatives of a patient who had survived malignant hyperpyrexia. They were found to have a very high level of serum creatine phosphokinase (CPK). Although the patient's muscles seemed normal, two of the three relatives had a mild but definite myopathy, affecting predominantly the lower muscles of the thigh. It seems that malignant hyperpyrexia develops in individuals with a myopathy which is inherited as an autosomal dominant,⁴ and which may be sub clinical. Therefore, all patients with myopathy and their relatives may be at risk of malignant hyperpyrexia (indeed this syndrome was described first in a patient with myotonia congenita).

Fortunately, affected individuals can be detected by a serum CPK estimation, all blood relatives of the patient who had malignant hyperpyrexia should be examined clinically and screened for raised serum CPK levels. It was advised that all individuals having general anaesthesia be screened by a serum CPK estimation because the myopathy causing malignant hyperpyrexia may not be detected clinically, and a family history of this rare disorder is usually not given. Whenever serum CPK level is elevated from some other causes, the body temperature should be monitored during anaesthesia, so that hyperpyrexia can be corrected at an early stage.

Muscle contracture tests used to be the corner stone for diagnosing patients with malignant hyperpyrexia, the test is not easy and it can be done only in few centres in the world, while serum CPK can be estimated in most Medical Centres.

 ^{*} Clinical Geneticist Genetic Unit
Salmaniya Medical Centre
State of Bahrain

Whenever faced with an unexpected attack, all anaesthetic agent should be discontinued at once and Dantrolene administered by rapid IV push, starting with 1 mg/kg and continue until the symptom begin to subside.^{5, 6} It is also advised to reduce the temperatue by vasodilator drugs, cool the patient, correct acidosis and any electrolyte disturbance (hyperkalaemia), and support the circulation.

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