MALIGNANT HYPERPYREXIA: A STUDY OF A FAMILY

Case Report

BY

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SUMMARY

The relatives of a patient who died in 1960 in malignant hyperpyrexia during anaesthesia have been investigated. Grossly raised creatine phosphokinase have been noted among them.

Recently increasing attention has been drawn to the occurrence of fatal hyperpyrexia in patients undergoing general anaesthesia. In some cases the tendency to develop this complication is said to be inherited as an autosomal dominant characteristic (Britt, Locher and Kalow, 1969).

In 1960 a patient died with malignant hyperpyrexia during general anaesthesia in Pontefract General Infirmary.

CLINICAL REPORT

A female patient aged 14 died in 1960 with malignant hyperpyrexia during an operation to correct recurrent dislocation of the patella. Pre-operative examination had revealed no abnormality. At the age of 7 she had experienced an uneventful tonsillectomy under thiopentone-ether-trichloroethylene anaesthesia. Premedication then consisted of morphine 10 mg and atropine 0.6 mg injected 1 hour before induction. Anaesthesia was induced with 2.5 per cent of thiopentone 100 mg, followed by suxamethonium chloride 40 mg. The response to suxamethonium was normal, with no increased fasciculation or rigidity. Intubation was easy and spontaneous respiration returned in a few minutes. Anaesthesia was maintained using nitrous oxide, oxygen and 1.5 per cent halothane. About 10 minutes after the start of surgery, that is 20 minutes after induction, the anaesthetist in charge observed the following.

The pulse was rapid and irregular. There was a marked facial pallor, a circumoral tinge of cyanosis, and the face felt very hot. The oral temperature was 110°F and the rectal temperature 106°F. It was impossible to keep the skin pink in spite of change to 100 per cent oxygen. The operation was rapidly concluded and active cooling with ice packs and sponging carried out. The patient's limbs were rigid, the arms semiflexed and the hands tightly clenched. Respiration became weak, irregular and artificial ventilation with 100 per cent oxygen was used.

Cardiac arrest occurred. Resuscitation was of no avail. She died at 5 p.m., some 2½ hours after the time of induction. Postmortem examination showed non-specific changes indicating anoxia with brain haemorrhages.

The serum creatine phosphokinase and aldolase levels of the members of this family have been studied, following reports of raised levels of these enzymes in a family with three cases of hyperpyrexia (Isaacs and Barlow, 1970).

The members of the family were contacted through their general practitioners and all agreed to the investigation. The subjects were instructed to avoid strenuous physical exercise for at least 48 hours prior to collection of blood. The blood was collected with minimum of stasis; all assays were carried out on fresh serum using U.V. methods (Boehringer Corporation, London). Normal ranges s.c.p.k. 0-50 mU/ml, aldolase 0-6.0 mU/ml.

RESULTS

The family tree, and enzyme determinations are shown in figure 1 and table I.

Our results support the findings of Isaacs and Barlow (1970) in that high levels of s.c.p.k. activity were found amongst asymptomatic relatives. It has been suggested that if high values of s.c.p.k. activity and malignant hyperpyrexia are manifestations of a muscle abnormality, then those asymptomatic patients with the highest values of s.c.p.k. are probably at greatest risk. Serum aldolase levels in all members of the family were normal, showing that in this instance s.c.p.k. activity is a more sensitive test for detection of sub-clinical forms of this muscle disorder. A link between malignant hyperpyrexia and myopathy has received support from other sources. The determination of the enzyme s.c.p.k. may be useful for detection of susceptible individuals, although there is no absolutely proven case for a cause-and-effect relationship between an elevated s.c.p.k. and the development of hyperpyrexia.

We have warned the family under study of a possible increased risk of operation and/or anaesthesia, especially in those members with s.c.p.k. values over 200 mU/ml. In the case of the pregnant lady (s.c.p.k. 83 mU/ml) the elevated value could possibly be a result of her pregnancy. Should anaesthesia be necessary during her confinement, alternative techniques—local, regional, conduction anaesthesia or a combination of nitrous oxide, oxygen and a narcotic mixture—should be considered. We have advised that her s.c.p.k. should be repeated after confinement and when her two children reach a reasonable age their s.c.p.k. should also be determined.

REFERENCES
MALIGNANT HYPERPYREXIA: STUDY OF A FAMILY

HYPERPYREXIA MALIGNE: ETUDE D'UNE FAMILLE

SOMMAIRE
Les membres de la famille d'un patient, décédé en 1960 d'une hyperpyrexia maligne, ont été contrôlés. On observa chez eux un taux élevé de phosphocreatine kinase sérique.

MALIGNE HYPERPYREXIE: BERICHT ÜBER EINE FAMILIE

ZUSAMMENFASSUNG

HIPERPIREXIA MALIGNA: ESTUDIO EN UNA FAMILIA

RESUMEN
Han sido estudiados los parientes de un paciente que murió en 1960 con hiperpirexia maligna durante anestesia. Se observó entre ellos una gran elevación de la fosfocreatina quinasa sérica.

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REVISION COURSE IN BASIC MEDICAL SCIENCES

A revision course in Basic Medical Sciences for the Primary F.F.A. will be held on alternate Saturdays from 10 a.m. to 4 p.m. from October 1971 to March 1972. The course will include lectures, demonstrations and tutorials. Fee £25 for the complete course. (Fees may be recoverable from the employing authority under Study Leave arrangements.) Further details may be obtained from the Secretary, Cambridge University Medical School, Hills Road, Cambridge CB2 1QT.

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INFORMAL JOINT MEETING IN INNSBRUCK

It is proposed to hold a joint meeting on an informal basis with the Austrian anaesthetists between January and March 1972. The programme will be arranged by Professor B. Haid, studying anaesthetic topics in the mornings, and skiing in the afternoons, in Innsbruck. After a few days the party would proceed to a mountain resort for more intensive practice of skiing.

Those interested please contact Dr Jean Horton, Addenbrooke's Hospital, Cambridge, telephone 55671, extension 254.