Malignant Hyperpyrexia in a Serving Officer

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SUMMARY: A case of Malignant Hyperpyrexia (MH) is described which highlights the need for continued vigilance amongst anaesthetists and raises the question of how these patients should be medically graded.

Case Report

A 26 year old female officer weighing 69 kg was admitted to hospital for extraction of four wisdom teeth the following day. She had received a general anaesthetic two years previously for a MUA of her ankle and had a strabismus correction under general anaesthesia when she was four years old. She reported no problems with either anaesthetic. Her concurrent medication consisted of mefloquine and the oral contraceptive pill. There were no other points of note in the pre-operative history or examination.

Premedication consisted of dexamethasone 8mg intramuscularly, 0.5% ephedrine drops to each nostril, temazepam 20mg and diclofenac 50mg orally. On arrival in the anaesthetic room electrocardiogram (ECG), pulse oximetry, and non invasive blood pressure (BP) monitoring was established prior to induction. The initial baseline pulse rate was 90bpm and the BP normal but not recorded. After preoxygenation, anaesthesia was induced intravenously with fentanyl 100mcg, thiopentone 375mg and suxamethonium 75mg. A 7.0mm Magill red rubber naso-tracheal tube was passed directly into the trachea without need for laryngoscopy. At this point it was noted that the patient had considerable flushing of her face and chest, this was presumed to be due to drug induced histamine release. Her chest was auscultated but there was no evidence of bronchospasm. Anaesthesia was maintained by manual ventilation with oxygen 30%, nitrous oxide 70% and isoflurane 2%. Laryngoscopy was then attempted in order to place a throat pack but was difficult as the patient's jaw was stiff. As the initial dose of suxamethonium could have worn off by this time she was given atropine 0.3mg (this did not produce a tachycardia) and a further 25mg of suxamethonium intravenously. This did not relax her jaw, a throat pack was sited with a little difficulty.

The patient was transferred into the operating theatre where the monitors at induction were reconnected

together with EtCO₂. She resumed spontaneous ventilation. In theatre the monitors displayed; SPO₂ 100%, BP 138/73 mmHg, ECG rate 140 bpm, EtCO₂ "on scale" (>10 kPa). At this point the surgical drapes were removed and the patient examined. She was not hot to touch but she was generally rigid – her knees could not be flexed, even with force. A presumptive diagnosis of malignant hyperpyrexia was made and surgery was abandoned.

Over the next ten minutes her rectal temperature rose from 36.9°C to 37.1°C and arterial blood gases revealed a base deficit of 10.6. Treatment was commenced according to protocol which included hyperventilation with volume free apparatus, active cooling, dantrolene 1mg/kg intravenously, mannitol 500ml of 10% intravenously and invasive monitoring – (CVP line, arterial line and a thermistor). Sedation was maintained with a propofol infusion and vecuronium was used to facilitate ventilation. Within 25 minutes of induction there were definite signs of clinical improvement as shown by a falling heart rate and a reduction in muscular rigidity.

The patient was transferred to the intensive therapy unit (ITU). Further therapy over the next 16 hours consisted of further doses of intravenous dantrolene, a total of 2mg/kg was required to treat a rising temperature. A diuresis over 200ml/hour was maintained and the urine alkalinised using intravenous sodium bicarbonate in order to avoid renal deposition of haematin.

Laboratory investigations during her ITU admission demonstrated a high white cell count of 17.64 10⁹/l with no other haematological or clothing abnormality. Serum lactate peaked at 2.9 mmol/l and creatine kinase at 424 iu/l. There was clear evidence of myoglobinuria. Other significant abnormalities were an AST of 1360 iu/l, ALT of 261 iu/l and a LDH of 1386 iu/l.

She was extubated the following morning and remained in ITU for the next 48 hours as her acidosis resolved slowly. Her core temperature fluctuated between 36.4°C

and 37.6°C for the first 24 hours before settling around 37.3°C. Thereafter she made an uncomplicated recovery on the ward. At follow up six months later she remains well.

Discussion

Malignant Hyperpyrexia is a rare condition usually inherited in an autosomal dominant fashion. It presents in around 1 in 15000 general anaesthetics (1,2). It is triggered by all anaesthetic volatile agents and the neuromuscular blocking drug suxamethonium (Scoline). An attack is characterised by muscular rigidity, hyperthermia and hypermetabolism, it is usually fatal unless treated with dantrolene. MH is associated with Central Core Disease and Muscular Dystrophy and possibly with Heat Stroke, Myotonia and other myopathies. The mortality has fallen in recent years to about 10% due to the introduction of dantrolene, better anaesthetic monitoring and an increased awareness of the disease amongst anaesthetists.

As far as the patient was concerned, her two previous anaesthetics were uneventful. We have since obtained copies of the medical records for both procedures. Anaesthesia for the manipulation under anaesthetic of her ankle was a total intravenous anaesthetic without inclusion of suxamethonium or any volatile agent. There is no anaesthetic record for the squint correction twenty-two years previously but the surgeon commented in the notes that the "patient remained stiff after scoline". The significance of this information was not appreciated and thus not passed on to the patient's parents. This serves as a reminder that previous uneventful anaesthetics are no guarantee against the development of MH on a subsequent occasion.

Current Ministry of Defence Policy states that a potential recruit known to be MH positive should be rejected for service. For serving personnel a medical

grade of P = 3, PES = LE and a medical warning tag with the words "Malignant Hyperpyrexia" is recommended (3). Serving personnel grade P3LE may of course be deployed in an area where medical cover is provided by a Field Hospital, Medical Support Troop or a Field Surgical Team. None of these units carry a stock of dantrolene, nor do they have on their inventory: vapour free apparatus, internal temperature probes, capnographs or other items considered necessary for the provision of safe anaesthesia to MH susceptible individuals (4). It is therefore the opinion of the authors that the appropriate medical grade for MH susceptible personnel is P = 7, PES = HO. We also recommend that field units on deployment carry a stock of dantrolene so that cases of MH arising *de novo* may be treated according to current protocols.

Addendum

Report received from the Malignant Hyperthermia Unit, St James's University Hospital, Leeds: Neuropharmacological results: susceptible to Malignant Hyperthermia. Neuropathology; muscle histology within normal limits.

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