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Emergency caesarean section complicated by paternal susceptibility to malignant hyperthermia

M Watanabe, E Yarnold, P Dewan
Department of Anaesthesia, Milton Keynes University Hospital NHS Trust

Introduction

Malignant hyperthermia (MH) is a rare, autosomal-dominant disorder in which a life-threatening reaction occurs during general anaesthesia (GA).

We describe a case where paternal susceptibility to MH was identified at advanced gestation and the plan made to reduce the potential risk to the baby during delivery.

Case Report

A 28-year-old primigravida presented at 38 weeks gestation to the Antenatal Day Assessment Unit following raised blood pressure (BP) readings in the community.

She had no significant medical history. Her obstetric history included breech presentation with failed external cephalic version, and a foetal cardiac echo showing a normal variation of dominant/persistent left superior vena cava draining to a dilated coronary sinus, but an otherwise structurally and functionally normal heart. An elective caesarean section (CS) had been booked for her at 39 weeks.

Clinical examination revealed bilateral oedema of the hands and feet, brisk reflexes, BP 170/100 mmHg, and nil else of note. Blood tests showed haemoglobin 137 g/L, platelets 216 x 10^9 /L, normal clotting function, and mildly deranged liver function tests. Urinalysis indicated a protein : creatinine ratio of 122 mg/mmol. Based on these findings a diagnosis of pre-eclampsia was made.

The patient was transferred to the Labour Ward for additional investigations, continuous monitoring, ongoing treatment, and to expedite delivery.

During a routine anaesthetic assessment, the patient's partner revealed that he was susceptible to MH. This was the first instance during the patient's care this information had been identified.

At this point, a multidisciplinary team discussion was held involving the obstetric, anaesthetic, and midwifery teams, and a plan was made for the safe delivery of the baby.

Management Plan:

1. Category 3 CS the following morning under spinal anaesthesia.
2. Neonatal team informed of potential risk to foetus should an accidental exposure to a trigger agent occur.
3. A “trigger-free” theatre, anaesthetic machine, and infusion pumps were prepared and kept on standby in the event a GA was required.
4. Further advice was sought from the specialist MH unit based in St James’ University Hospital, Leeds.

The patient proceeded to deliver a healthy female infant by category 3 CS under spinal anaesthesia without any complications.

Although the maternal anaesthetic history forms a normal part of a pre-operative assessment, the anaesthetic history of the partner does not. This case highlights the importance of early recognition and planning for a potentially MH-susceptible foetus as delay may lead to serious harm.

Discussion

Malignant hyperthermia is a pharmacogenetic disorder characterised by a potentially life-threatening hypermetabolic crisis in susceptible individuals. Trigger agents are drugs commonly used in general anaesthesia, namely succinylcholine and volatile gases.

The incidence in the UK is estimated to be 1:50,000 - 1:70,000 and the prevalence of susceptibility is reported to range from 1:3,000 - 1:100,000.[1] Mutations in the RYR1 gene on chromosome 19 account for the majority of cases of MH susceptibility.[2]

Clinical manifestations include a rise in end-tidal CO_2, tachypnoea if spontaneously breathing, tachycardia, muscle rigidity, metabolic acidosis, hyperkalaemia and cardiac arrythmias.

Current mortality is estimated to be below 5% due to an increased understanding of the condition and the introduction of therapeutic agents such as dantrolene.[3]

Both the European Malignant Hyperthermia Group (EMHG) and the Malignant Hyperthermia Association of the United States (MHAUS) have published recommendations for the provision of anaesthesia for caesarean sections in cases with MH-susceptibility in the mother or the foetus.[4,5]

SUMMARY OF KEY RECOMMENDATIONS BY EMHG & MHAUS

1. Discuss anaesthetic management plan with patient in the antenatal period.
2. Inform anaesthetist as soon as patient booked into labour ward, ensure ability to provide trigger-free anaesthesia.
3. Consider early epidural analgesia once labour in progress (can be converted into epidural anaesthesia).
4. Use of neuraxial anaesthesia for delivery (unless contraindicated or where GA is required for obstetric indications).
5. If GA is required for there to be direct supervision by an experienced anaesthetist.
6. Intravenous agents to be used for both the induction and maintenance of GA.
7. Use of rocuronium for muscle relaxation to facilitate tracheal intubation.
8. Use of a combination of non-triggering drugs which the anaesthetist is most familiar with.
9. No consensus on use of succinylcholine in cases of possible MH-susceptibility in the foetus only.

References

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