Upper limb compartment syndromes: a complication of malignant hyperthermia in a patient with ill-defined myopathy

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Summary
We report a case of compartment syndrome complicating malignant hyperthermia (MH) in a 12-yr-old girl with a history of myopathy and multiple skeletal deformities; she underwent bilateral Achilles tendon surgery. Marked oedema of both forearms became evident in the immediate postoperative period and resolved after conservative treatment. Compartment syndrome is a rare complication of MH. Early recognition and therapy may prevent the onset of muscle ischaemia and distal neurovascular deficit. The need for urgent surgery and repeated anaesthesia in the early phase of recovery from an acute episode of MH may thus be reduced. (Br. J. Anaesth. 1995; 74: 343-344)

Key words
Malignant hyperthermia Complications, compartment syndrome. Complications, myopathy.

Malignant hyperthermia (MH) is a disorder of skeletal muscle metabolism precipitated by recognized general anaesthetic “trigger” agents. Hypermetabolism and muscle spasm may result in the formation of oedema in the confined space of fascial sheaths and subsequently in the development of a compartment syndrome [1]. Several rare myopathies have been shown to predispose to the development of MH. Central core disease, characterized by muscle weakness and distinct histological appearances, is strongly associated with MH susceptibility [2]. Hypotonia, skeletal deformities and cryptorchidism are clinical manifestations of the King-Denborough syndrome, an uncommon myopathy which has also been reported to predispose to the development of MH [3].

We describe a case of MH in a patient with ill-defined myopathy who developed bilateral compartment syndromes of the forearm after operation. Early conservative treatment may have prevented progression of the condition, thus avoiding the risk of further anaesthetic exposure.

Case history
A 12-yr-old (25-kg) girl with multiple skeletal deformities was admitted to the intensive therapy unit (ITU) after an acute episode of MH which occurred during anaesthesia for bilateral Achilles tendon surgery. Previous muscle biopsies had shown an ill-defined myopathy with non-specific type 1 fibre predominance. There had been no previous anaesthetic exposure and there was no family history of an adverse reaction to anaesthesia.

The patient was premedicated with oral temazepam 10 mg. After uneventful induction of anaesthesia with propofol 2.5 mg kg⁻¹ and insertion of a laryngeal mask airway, controlled ventilation was commenced until spontaneous ventilation resumed. Anaesthesia was maintained with 60% nitrous oxide and 1–2% isoflurane in oxygen using a circle system with a carbon dioxide absorber. The following were monitored: non-invasive arterial pressure, ECG, pulse oximetry, capnography and rectal temperature. Vital signs were stable with an oxygen saturation of 99%, end-tidal partial pressure of carbon dioxide 7.8 kPa, heart rate 94 beat min⁻¹, arterial pressure 110/65 mm Hg and rectal temperature 36.5°C. Twenty minutes after commencing the procedure, with an arterial tourniquet applied to the right thigh to provide a blood-free operating field, the patient developed sinus tachycardia of 138 beat min⁻¹. The anaesthetist noted marked rigidity of the upper limbs which coincided with a sudden increase in end-tidal partial pressure of carbon dioxide to 15.0 kPa and a decrease in oxygen saturation to 80%. Anaesthesia was discontinued, assisted ventilation with 100% oxygen was commenced and dantrolene 40 mg was administered i.v. Arterial blood-gas measurements, obtained 20 min after the event, showed a combined severe metabolic and respiratory acidaemia (PₐO₂ 1.0, pH 7.12, PₐCO₂ 15.3 kPa, PₐCO₂ 6.7 kPa, HCO₃⁻ 14 mmol litre⁻¹, base excess 12.3 mmol litre⁻¹). Vital signs remained stable and the patient was transferred to the ITU awake and breathing spontaneously. At no time during the perioperative period was the patient’s rectal temperature elevated.

Laboratory investigations showed a serum potassium concentration of 6.6 mmol litre⁻¹, creatine kinase (CK) 20000 iu litre⁻¹ with a peak concentration of 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentration of 1.73 mmol litre⁻¹. Haemoglobin concentration remained normal and quantitative laboratory assay for urinary myoglobin concentration 115000 iu litre⁻¹ after 48 h, and a corrected serum calcium concentra...
was negative. The acidaemia resolved gradually over the following 48 h. Dopamine 3 μg kg⁻¹ min⁻¹ and sodium bicarbonate 8.4 % (3 mmol kg⁻¹ in 24 h) were administered to promote alkaline diuresis. Dantrolene 3 mg kg⁻¹ in 24 h was given in divided doses.

Marked swelling of both forearms developed 6 h after admission to the ITU, sufficient to prevent normal fastening of the patient’s wristwatch strap which required loosening by two notches. The patient complained of pain in the flexor compartment of both forearms, which was aggravated on passive finger extension. Movement of the fingers was restricted but peripheral perfusion and sensation were normal. The swelling and discomfort subsided gradually after both arms had been maintained in an elevated position and resolved completely after 48 h. Further recovery was uneventful and the patient was discharged from hospital with arrangements for further investigation of MH susceptibility.

Subsequent muscle biopsies showed histological features of a non-specific myopathic condition consistent with MH myopathy. The diagnosis of MH susceptibility was confirmed by in vitro contracture tests.

Discussion

Compartment syndromes are characterized by increased tissue pressures within the confined space of fascial sheaths. The lower extremities, in particular the narrow tibialis anterior compartment, are affected more commonly than the upper limbs. Loss of integrity of the microcirculation with fluid exudation into the interstitial space results in oedema formation, muscle swelling and raised intracompartmental pressures, eventually leading to compression of blood vessels and nerves [1].

Reperfusion injury, trauma, crush injury and a variety of non-traumatic conditions have been described as aetiological factors in the development of compartment syndrome [4]. These include rhabdomyolysis which is a prominent feature of MH. Hyperkalaemia and elevated concentrations of CK are ominous signs and may herald the development of myoglobinuria [5]. In our patient, laboratory tests suggested substantial muscular injury sufficient to cause rhabdomyolysis, although myoglobinuria was not observed.

The only previously reported case of compartment syndrome in MH shared a number of similarities with our patient [6]. In both instances isoflurane appeared to have been the “trigger” agent and, in both operations a pneumatic tourniquet was applied to one lower limb. In contrast with our patient in which the initial muscle rigidity spared the limb isolated by the tourniquet, Helms and colleagues reported that the compartment syndrome had occurred in both lower limbs.

“Subclinical” compartment syndromes may be a common manifestation of MH but may evade recognition because they reflect a transient response to altered muscle metabolism and resolve rapidly with successful treatment of the condition. Physical examination, including passive stretching of involved muscle groups, should be performed frequently to detect early signs of compartment syndrome. Direct percutaneous monitoring of intracompartmental pressures has been proposed [7]. Progressive increase in muscle compartment pressures may be prevented by early recognition and conservative therapy. The need for fasciotomy and further high risk anaesthesia in the recovery phase from MH may thus be reduced.

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References