A patient with severe central core disease

Editor—We report a case of a 19-yr-old primigravida who was referred to our tertiary obstetric unit at 37 weeks gestation. She was known to have skeletal muscle weakness since birth and had a history of delayed motor milestones as a child. Myopathy of an unknown aetiology had been suspected since early childhood. Additionally, after sudden cardiac deaths of the patient’s father and brother, concerns of an underlying familial cardiomyopathy or rhythm abnormality had been raised.

Our patient had an unremarkable pregnancy with no clinical history suggestive of cardiovascular disease. On examination, her height was 148.5 cm with the only positive finding being minimally reduced power involving the upper arms. There was no contracture or any spinal deformity. Her gait was normal. Systemic examination revealed no abnormal findings. ECG and 2D echocardiogram were normal with no evidence of dysrhythmia or cardiomyopathy. An elective Caesarean section at 38 weeks gestation was planned as our patient had cephalopelvic disproportion. In view of her history, the operation was scheduled in the cardiac theatre suite.

Before operation, she received aspiration prophylaxis as standard. Compound Ringer’s lactate solution was infused i.v. External defibrillator pads were attached and lateral tilt and calf compressions used. Radial artery was cannulated. Regional anaesthesia was induced using a combined spinal–epidural technique using 2 ml of heavy bupivacaine 0.5% and fentanyl 20 μg. Caesarean section was uneventful with 300 ml of total blood loss. A healthy baby boy was delivered weighing 2.8 kg. A rectus abdominus muscle biopsy was performed and after operation, she was transferred to the high dependency unit. She made a complete recovery and was discharged on Day 10. The diagnosis of central core disease was confirmed on histopathology with biopsy findings suggestive of long standing and severe disease.

Central core disease is a slowly progressive autosomal dominant congenital myopathy. It presents during infancy with delayed motor milestones secondary to generalized hypotonia and muscle weakness. It is a histopathological diagnosis made on finding characteristic central cores on muscle biopsy. The ‘central core’ is an area of central clearing in the muscle with the loss of myofibrils, mitochondria, and glycogen. Central core disease has been consistently associated with malignant hyperthermia (MH). Susceptible patients with central core disease have been shown to have mis-sense mutation in the ryanodine receptor gene on chromosome 19. Genetic screening of all patients with central core disease for MH has been recommended.

Although our patient had an uncomplicated anaesthetic outcome, she had a high risk of developing MH and avoidance of general anaesthesia probably helped in preventing this developing. Evidence from the literature would suggest that any patient with an undefined myopathy that has not been confirmed by histopathology, genetic mutation analysis, or both is a possible candidate for central core disease and therefore of developing MH. This case highlights the subtle clinical features with which patients with central core disease can present. Even in patients with known central core disease, it is safest to avoid triggering agents like volatile anaesthetics and suxamethonium even if they have a history of safe anaesthetics in the past, as patients with central core disease may develop MH as a first episode, despite having no complications on previous general anaesthesia (GA). This susceptibility is thought to depend on the biochemical milieu at the time of the anaesthetic rather than an absolute risk with each GA. Ropivacaine as opposed to bupivacaine has been shown to have lesser motor block in a multicentre study in labour. It has been reported to be a safer agent to use in patients susceptible to MH. If GA is needed, we suggest using total i.v. anaesthesia with a vaporex free anaesthetic machine as has been recently reported. Anaesthetists need to maintain a high index of suspicion for risk of developing MH in any patient with an ill-defined myopathy or non-specific features of muscle disease.

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A LMA CTrach™ for large patients

Editor—The LMA CTrach™ system (CTrach) (The Laryngeal Mask Company, Singapore) enables viewing of the glottis, alignment of the laryngeal mask conduit with the glottis, and tracheal intubation under vision. In earlier work with the CTrach, it was frequently difficult to view the glottis, and epiglottic downfolding was the most common cause. This may limit the CTrach’s usefulness in managing difficult airways and may be a particular problem in tall obese male patients. A CTrach airway modified for use in big patients may be needed and hence the size 5L CTrach airway was recently developed.

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