Anaesthetic management of a child with dermatomyositis

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A two years, ten months old male with dermatomyositis was anaesthetized with enflurane, nitrous oxide and oxygen by mask followed by intravenous succinylcholine to facilitate endotracheal intubation. The evoked thumb twitch in response to succinylcholine demonstrated an abnormal, short-lived contracture. The depresion, duration and return to control of muscle twitch tension and a transient rise in serum potassium concentration followed a normal pattern.

Dermatomyositis is a multi-system disease manifested by nonsuppurative inflammation of striated muscle and characteristic skin lesions. Proximal muscle weakness, impairment of swallowing and respiratory function, and cardiac and cutaneous manifestations require special consideration by the anaesthetist.

We present a case report of the anaesthetic management of a paediatric patient with dermatomyositis in which an unusual response to succinylcholine was demonstrated.

Case report

A Caucasian male, aged two years, ten months, was admitted to the hospital for evaluation of progressive proximal muscle weakness, a scaly erythematous rash and failure to thrive.

On physical examination, the child weighed 10.4 kg, had a height of 78 cm, and head circumference of 48 cm, all less than the 5th percentile for age. Rectal temperature was 38.2° C, blood pressure 104/62, heart rate 116, and respiratory rate 22. Numerous scaly, atrophic, hypopigmented papules as well as telangiectasias were noted over the interphalangeal and metacarpophalangeal joints, knees, and elbows. A violaceous and scaly rash with a primary malar and periorbital distribution was noted on the face. Cardiac examination showed a regular rhythm without murmur or gallop. The chest was clear to percussion and auscultation. There was symmetrical wasting and decreased strength (3/5) in the musculature of the shoulder and pelvic girdles, and the child could not stand or sit from a supine position. The remainder of the physical examination was unremarkable.

White blood count and platelet count were normal. Haematocrit was 31.7 per cent. Serum electrolytes, blood urea nitrogen, and creatinine were within normal limits. Aspartate aminotransferase was 44 $u \cdot L^{-1}$ (normal values 4–35 $u \cdot L^{-1}$) and lactate dehydrogenase was 482 $u \cdot L^{-1}$ (normal values 150–420 $u \cdot L^{-1}$). Other serum skeletal muscle enzyme values were normal.

Electrocardiogram and chest *x*-ray were normal. *X*-rays of the extremities revealed oedema and numerous areas of soft tissue calcification. A barium swallow revealed reflux from the oropharynx into the nasopharynx, normal oesophageal peristalsis, and mild reflux from the stomach into the lower oesophagus. Electromyography of the quadriceps revealed spontaneous fibrillation potentials and reduced amplitude of voluntary contraction potentials.

Anaesthesia was required for a muscle biopsy to confirm the diagnosis of dermatomyositis. The child was brought to the operating room 30 minutes

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after he was premedicated with atropine 0.2 mg IM. Electrocardiogram leads, a blood pressure cuff and an oesophageal stethoscope were applied. Due to an inability to obtain intravenous access while the child was awake, a mask induction was performed with 40 per cent oxygen, 60 per cent nitrous oxide, and 0.5 per cent incremental concentrations of enflurane, every three breaths up to 4.0 per cent inspired concentration. Cricoid pressure was maintained to prevent reflux of stomach contents. During induction a Grass FT-10 force transducer was attached to the hand to monitor twitch tension of the thumb in response to supramaximal ulnar nerve stimulation of 0.15 msec duration at 0.15 Hz. A Gould strip chart recorder run at 5 $mm \cdot min^{-1}$ was used to record the force transducer measurements. A 22 gauge IV catheter was inserted in a foot vein for fluid infusion and a 20 gauge catheter was inserted in an antecubital vein for venous blood sampling. Nitrous oxide was discontinued, and succinylcholine, 1 $mg \cdot kg^{-1}$ IV, was administered.

Approximately 35 seconds after succinylcholine administration, upward baseline shift of the evoked thumb twitch recording began to occur, and this contracture continued to increase in magnitude for 15 seconds, at which time it began to decrease. The recording returned to its original baseline 1.5 minutes later (Figure). Evoked twitch height began to decrease just as contracture began, twitch height falling to eight per cent of control two minutes after succinylcholine administration. Intubation with a 4.5 mm (ID) endotracheal tube was performed 1.5 minutes after succinylcholine administration, without difficulty. No movement of the vocal cords or gross motor activity accompanied intubation, nor were fasciculations seen at any time following succinylcholine. Twitch height returned to 100 per cent of control height 4.5 minutes after succinylcholine administration.

Anaesthesia was maintained throughout the 30-minute surgical procedure with 2.0 per cent enflurane and 50 per cent nitrous oxide in oxygen. Serum potassium levels were determined before and 1, 3, and 15 minutes following succinylcholine administration. Values were 3.9, 4.2, 4.7, and 3.6 $mEq \cdot L^{-1}$, respectively. Blood pressure, heart rate, and the electrocardiogram remained stable throughout. At the end of the procedure, enflurane and nitrous oxide were discontinued. The trachea was extubated in the operating room afer the patient was fully awake. The patient was taken to the recovery room and made an uneventful recovery. Biopsy results were consistent with the clinical diagnosis of dermatomyositis.

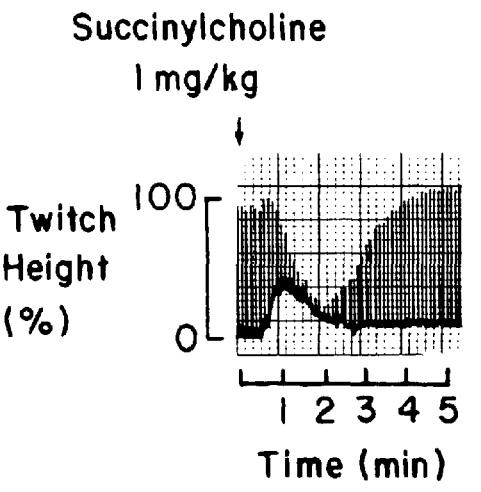

FIGURE Neuromuscular block following succinylcholine 1 $mg \cdot kg^{-1}$ IV in a child with dermatomyositis. Note simultaneous onset of contracture (upward baseline shift) and block approximately 35 seconds after administration of succinylcholine. Peak block of 92 per cent occurred after two minutes, with recovery of original twitch height 4.5 minutes following succinylcholine.

Discussion

Dermatomyositis is a chronic inflammatory disease primarily affecting muscle and skin. Although its cause is uncertain, cellular immune mechanisms have been strongly implicated in its pathogenesis.[1] The onset is insidious, with symmetrical weakness of the limb girdle and anterior neck flexors which is progressive over weeks to months. Soft tissue contractures, muscle atrophy, and subcutaneous calcification may develop with progression of the disease. Severe involvement of the muscles of respiration and deglutition has resulted in respiratory impairment, aspiration, and death.[2]

A characteristic skin rash is common and may be the presenting symptom. It consists of a violaceous heliotrope discoloration of the upper eyelids with periorbital oedema, a scaly erythematous dermatitis in a malar distribution, and symmetric erythema-

tous atrophic changes over the extensor surfaces of joints.[3] A widespread necrotizing vasculitis may be present in the childhood form of this disease.[3] Mucosal ulcers, cutaneous necrosis, and gastrointestinal ulceration with bleeding and perforation have been reported.[1,4] Pulmonary problems may be encountered due to aspiration pneumonia, respiratory muscle weakness, and interstitial pneumonitis.[5] Although 40 per cent of patients will present without pulmonary complaints, complete examination and investigation often demonstrates bibasilar rales, hypoxemia and restrictive lung disease.[6] Cardiac manifestations including arrhythmias, cardiomyopathy, and bundle branch block are well described.[2,4] Death as a consequence of myocarditis or arrhythmias has been reported.[7]

Serum skeletal muscle enzymes are often elevated, particularly creatine phosphokinase and often aldolase, serum alanine and aspartate aminotransferases, and lactate dehydrogenase. Electromyography may reveal a triad consisting of spontaneous fibrillation potentials, reduced amplitude of voluntary contraction potentials, and repetitive potentials on needle insertion. Muscle biopsy adds support to the diagnosis. Pathologically, there is variation in fiber size and necrosis and vacuolization of both Type I and Type II muscle fibres. There may be an inflammatory perivascular infiltration and areas of fibrous replacement of muscle.[1]

While the anaesthetic management of dermatomyositis has been described, little information exists on the use of muscle relaxants in these patients. Sensitivity to the nondepolarizing relaxants[8–10] has been referred to anecdotally, but without elaboration. Cautious titration of nondepolarizing relaxants with the use of a peripheral nerve stimulator is recommended. No published reports on the use of succinylcholine or other depolarizing relaxants in this disease are available.

Use of succinylcholine in our patient resulted in a short-lived contracture response similar to that seen following succinylcholine administration in a patient with myotonic dystrophy reported previously.[11] Depression, duration, and return to control of muscle twitch tension followed a normal pattern. A mild and transient rise in the serum potassium concentration followed the administration of succinylcholine (eight per cent rise at 1 minute, 20 per cent at three minutes, and return to near baseline by 15 minutes), consistent with increases seen in normal children given succinylcholine following halothane induction.[12]

Although there was no evidence of malignancy in this patient, it is important to point out that occult malignancy may be associated with dermatomyositis and that the possibility of malignancy-related neuromuscular weakness and myasthenic syndrome may exist.[10] The anaesthetist should therefore exercise caution in using either depolarizing or non-depolarizing muscle relaxants.

In summary, dermatomyositis is a multi-system disease of importance to the anaesthetist primarily because of its association with impaired airway reflexes, respiratory dysfunction of multiple aetiologies, and myopathy with uncertain response to muscle relaxants. Use of succinylcholine in a patient with dermatomyositis did not appear hazardous. Further evaluation of the effects of succinylcholine in patients with dermatomyositis is warranted.

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Résumé

Un patient âgé de deux ans dix mois atteinte de dermatomyosite a été anesthésié avec l'enflurane, protoxyde d'azote, oxygène, au masque après administration intra-veineuse de succinylcholine afin de faciliter l'intubation endotrachéale. Après la succinylcholine la réponse à la stimulation musculaire était anormale et de courte durée. La dépression, durée et ainsi que le retour au contrôle de la contraction musculaire ainsi que l'élévation transitoire du potassium sérique étaient prévisibles.