ANAESTHETIC HAZARDS IN DERMATOMYOSITIS

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Dermatomyositis is a relatively rare diffuse connective tissue disorder which is characterized by muscle weakness and inflammatory changes in muscles and skin[1]. It affects females twice more commonly than males, with a peak onset between 30 and 60 years of age. Frequency is not known but the incidence is roughly one new case per 300,000 persons per year[2]. The aetiology is obscure, but genetic, immunologic and viral factors have all been incriminated. The diagnosis is based on the clinical picture of muscle weakness and skin rash, myelography, raised serum C.P.K. levels and muscle biopsy. There is a paucity in the world literature on the anaesthetic management of these patients. The disease presents some unique problems for anaesthetists in the preoperative, intraoperative and the postoperative period. We present a case history which illustrates some of the difficulties which may be encountered.

CASE HISTORY

A forty six years old female presented as an emergency for drainage of abscess on her back. The patient had presented to our medical out patient department six weeks ago for evaluation of muscle weakness of three months duration, and a skin rash on the face, chest and dorsum of the hands of five months duration. She also gave a history of hypertension controlled with Aldomet for the past twenty years, and diabetes mellitus controlled by oral hypoglycenucs for the past ten years. On examination, she had a puffy face and hands, On neurological assessment , she was found to have gradc 4/5 strength in the upper limbs and 3/5 in the lower limb joints, The gait was waddling. A difficulty in swallowing was also ascertained at this stage. Haematological and serological studies showed anaemia (Hb 9.1 mg%) and raised levels of serum alkaline phosphatase, aldolase, and C. P. K. (17, 416 iu/L) R.A., A.N.A. and L.E. cells were all negative. E.C.G. revealed a left bundle branch block and chest x-ray showed border-line cardiomegaly. Muscle biopsy was diagnostic of dermatomyositis. She was started on 60 mgs of oral prednisolone daily to which she responded. Six weeks later this patient presented with multiple large abscesses on her back, uncontrolled diabetes, and septicemia. Arterial blood gases showed aPaO2 of 77mm of mercury, PaCo2 of 29 mm of mercury and an oxygen saturation of 95% on air. Her diabetes was managed with insulin dextrose infusion and for her low cardiac output status she was started on dopamine infusion. Broad spectrum antibiotics were given to cover the septicemia. Two days later she presented to the anaesthetist for drainage of multiple abscesses on her back as an emergency. Her blood pressure and pulse were stable at this stage. No premedication was given except for hydrocortisone one hour before surgery. She was preoxygenated and induced with thiopentone 100 mgs intravenously and suxamethonium, and intubated with cricoid pressure. Intubation turned out to be difficult due to prominent incisors and a some what anteriorly placed larynx and a size 7.5 tube was passed with the help of a stylet. She was mechanically ventilated and anaesthesia was maintained on nitrous-oxide, 50% oxygen, 0.5% halothane, and 0.4 mg/kg of atracurium. Twitch response was monitored throughout with aMinistem (PrGfessional industries, Houston, Texas) by visual estimation of thumb adduction after ulnar nerve stimulation for two seconds at 2 Hz (Train of Four). The intraoperative period was uneventful. At the end of procedure the patient displayed all four thumb twitches in response to train of four stimulation and did not require neostigmine. Extubation was performed when she was fully conscious, and maintaining an adequate tidal volume. The ETC02 was checked by a capnograph before extubation and was within normal limits. Postoperatively she was kept

on the Intensive Care Unit. Analgesia was provided with intravenous pethidine in titrated doses. She maintained her PaO2 on 4 litres per minute oxygen. She was discharged from the Intensive Care Unit on the second day. The same patient was re-anaesthetised six times over a period of nine months for drainage of recurrent abscesses, change of dressings and finally for partial thickness skin grafting of the debrided area. In subsequent anaesthetics valium was used for premedication and enflurane was substituted for halothane. During one of the anaesthetics the patient received pancuronium 0.06 mgs/kg and recovered normally.

DISCUSSION

The anaesthetic problems encountered in patients with dermatomyositis undergoing general anaesthesia will be considered under the following subheadings:

Preoperative

Respiratory System: The patients may have aspiration pneumonia due to weakness of the muscle involved in swallowing[3]. There may be progressive weakness of the intercostal and diaphragmatic muscles resulting in respiratory insufficiency. Lung involvement may occur from the connective tissue disorder itself which results in patchy infiltrates throughout both lungs, interstitial pneumonia or fibrosis[2,4,5]. Carcinoma of the bronchus or lung parenchyma has a higher association with dermatomyositis[6,7].

Cardiovascular system

Cardiac muscle though not severely involved shows changes similar to skeletal muscles. Clinical manifestations are rare, but heart failure and conduction defects have been reported[2].

Haemopoietic system

Anaemia is commonly found in these patients due to the multisystem involvement.

Other systems

A few patients may exhibit signs of some paraneoplastic disorder, e.g., polyneuropathy, subacute cerebellar degeneration, multifocal neuroencephalopathy or myasthenic syndrome. Richardson and Davidson on examining ten patients with dermatomyositis found a myasthenic response in two patients using decamethonium test[7]. This is similar to the findings in myasthenia gravis. These patients are on steroids for treatment of dermatomyositis and therefore require preoperative and postoperative steroid cover. Some patients who do not respond to steroids are given cytotoxic drugs and their haematological status should be examined in more detail preoperatively. Our patient did not have any sign of aspiration pneumonia, though she did give a history of slight difficulty in swallowing at her first admission. Arterial blood gases done on several occasions did show some degree of hypoxemia on air without carbondioxide retention which could be due to the lung involvement owing to the underlying parenchymal involvement. Pulmonary function tests were not done at the first admission as the patient presented in emergency. On chest x-ray cardiomegaly, and on ECG a left bundle branch block was present but could have been secondary to her hypertension. She did have anaemia and was on steroids preoperatively.

Intraoperative

If the patient's ventilatory status is marginal preoperatively then he should not be permitted to breathe spontaneously during surgery. Difficult intubation is not a feature of this disease[8]. Airway protection and adequate ventilation are the two primary anaesthetic concerns in these patients. High incidence of swallowing and vocal cord dysfunction in these patients may lead to pooling of saliva in the pharynx and aspiration into the trachea[9]. Endotracheal intubation is therefore strongly recommended. Muscle relaxant should be used carefully and in smaller doses because of diminished muscle mass and altered sensitivity to the relaxants[10,11]. Narcotics should also be used with caution to prevent any postoperative

respiratory depression. General anaesthesia may be preferred over regional because of airway problems which may be encountered due to sedation in conjugation with the regional technique. In our patient suxamethonium was chosen initially for the speed of onset of muscle relaxation, because of the increased risk of aspiration and anticipated difficulty due to prominent upper incisors. We intubated and ventilated her on all occasions using atracunum for intubation and maintenance of relaxation in subsequent anaesthetics. The twitch response was normal and she did not exhibit abnormal sensitivity to either atracurium in a dose of 0.4 mg/kg or pancuronium in a dose of 0.06mg/kg. The use of atracurium has not been previously reported in literature in this disease. As difficult intubation is not a feature of this disease, in our patient it was probably due to large protruding incisors and an anteriorly placed larynx.

Postoperative

Respiratory insufficiency is the major postoperative complication. These patients may require mechanical ventilation in the postoperative period. Due to the weakness of thoracic muscles the patients may have a diminished cough reflex, leaving them vulnerable to atelectasis. Weakness of pharyngeal muscles may make them more vulnerable to aspiration pneumonia postoperatively. Postoperative pain relief should be with a titrated analgesic regime or regional block if possible. Our patient had an uneventful recovery from the general anaesthesia at all occasions. A plan of anaesthetic management of these patients is presented (Table).

TABLE. Anaesthetic Management Plan for Patients with Dermatomyositis.

PREOPERATIVE
Baseline arterial blood gas
Baseline pulmonary function tests
Treatment of any current pulmonary tract infection
Breathing exercise
Chest x-ray
Steroid cover
INTRAOPERATIVE
Endotracheal intubation
IPPV
smaller doses of narcotics use of muscle relaxants
Smaller doses of narcotics
Monitoring of neuromuscular function
POSTOPERATIVE
Ventilate till adequate minute ventilation
Postoperative pain relief:
a. Titrate analgesia b. Regional blockade
Postoperative chest physiotherapy.

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