Dermatomyositis with scleroderma-overlap syndrome and its anaesthetic implications

Sir,

Dermatomyositis is an autoimmune disease involving the skin and muscles, and may occur alone or in overlap syndromes mostly with scleroderma. It mainly involves thickening of the skin and fibrosis of the connective tissue, leading to dysphagia due to oesophageal dysmotility, pulmonary pneumonitis, cardiomyopathy, glomerulonephritis and Reynaud's phenomenon along with progressive muscle weakness.[1] We report the successful management of a patient of overlap syndrome suffering from hiatus hernia posted for laparoscopic repair.

A 40-year-old female, known case of overlap syndrome, presented with complaints of chronic regurgitation. She was diagnosed with hiatus hernia and was posted for laparoscopic repair. The patient gave a history of gradual-onset bilateral proximal muscle weakness over 4 years. The muscle biopsy showed vacuolisation and hyalinisation, and electromyography reported a primary generalised muscle disease. Creatinine phosphokinase levels were raised marginally. She was started on Tab. Prednisolone 15 mg thrice daily (TDS), which she took for 2 years and discontinued after partial relief. Currently, she had muscle weakness of grade IV/V, slight restriction of neck extension, dysphagia, sclerodactyly and Reynaud's phenomenon. The remainder of the systems were unaffected. Routine investigations were normal. Antacid treatment was started. On the pre-operative night, she was given antacid and pro-kinetic drugs. After confirmation of nil by mouth status, written informed consent and consent for post-operative ventilation, Inj. Glycopyrrolate 5 μg/kg was given intramuscularly. Pre-medication included Inj. Ranitidine 1 mg/kg I.V., Inj. Metoclopramide 0.1 mg/kg, Inj. Midazolam 0.03 mg/kg and Inj. Pentazocine 15 mg I.V. as analgesic. Inj. Hydrocortisone 100 mg and Inj. Dexamethasone 8 mg I.V. were given as the patient was on steroids initially. A nasogastric tube was inserted and, after aspiration, was withdrawn up to the nasopharynx. After adequate pre-oxygenation, the patient was given Inj. Fentanyl 50 μg and induced with Sevoflurane 0–8% while maintaining cricoid pressure and intubated. Anaesthesia was maintained on O_2:N_2O (50:50), Sevoflurane 1.5–1.8% and Propofol infusion 4–6 mg/kg/h without the use of any muscle relaxant on assisted ventilation. She was covered well to prevent hypothermia.

The duration of surgery was 2 h, and the muscle relaxation was adequate. Post-surgery recovery was quick and the post-op period was uneventful. The major problems in this rare case include difficult airway due to skin thickening and contractures, unknown response to muscle relaxants, cardiomyopathy, Reynaud's phenomenon, chances of aspiration of the stomach contents and side-effects of prolonged steroid treatment. The hiatus hernia could also be due to the scleroderma of the visceral organs but, because there was no improvement in the reflux with treatment, surgery was the only option. The chance of aspiration was high, but adequate pre-operative prophylaxis with pro-kinetic drugs, antacids, nasogastric tube aspiration and cricoid pressure during induction decreased the risk. Taking into account the cardiac depressant action of the anaesthetic agents, titrated doses were given. Inj. Propofol was used to avoid the neuroendocrine response associated with laparoscopy.

There are documented cases with the use of skeletal muscle relaxants in dermatomyositis for laparoscopic surgeries as, after pneumoperitoneum, it was difficult to ventilate. But, in our case, we did not face this problem probably because of the use of Sevoflurane.[2]

Others studied the effect of muscle relaxants in patients of dermatomyositis and found that the duration of action of Pancuronium increased from an average of 1.8 h to 3.1 h.[3,4]

Some studies showed an increased duration of action with the use of Inj. Suxamethonium in dermatomyositis, possibly due to atypical cholinesterase.[5] The 5-year survival rate for dermatomyositis is 95% and the 10-year survival rate is 84%. Most deaths occur due to respiratory and cardiac complications.[1] Hence, if the pulmonary and cardiac component is taken care of, these patients have a very fruitful life. Our aim was to highlight the possible complications associated with anaesthesia for these patients and ways to avoid them.

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