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ANAESTHETIC MANAGEMENT IN A PATIENT WITH DERMATOMYOSITIS – A CASE REPORT

Anaesthesiology

S. Sulekha Senior Resident, Department of Anaesthesiology, ESIC Medical College and PGIMSR, Chennai, Tamilnadu.

R. Arun Kumar* Associate Professor, Department of Anaesthesiology, PSGIMSR, Coimbatore, Tamilnadu. *Corresponding Author

ABSTRACT

Dermatomyositis is a rare autoimmune connective tissue disorder characterised by muscle weakness due to idiopathic inflammatory myopathy. It is a multisystem disorder. Females are more commonly affected. The major concerns of Anaesthesiologists are delayed recovery from muscle relaxant, aspiration pneumonitis, arrhythmias, cardiac failure and steroid supplementation with its complications. It is generally believed that patients with polymyositis are sensitive to Non Depolarising Muscle Relaxant and its use may cause muscle weakness and delayed recovery. We are presenting a case of paraneoplastic dermatomyositis with carcinoma of right breast who underwent modified radical mastectomy.

KEYWORDS

Dermatomyositis, Polymyositis, Muscle relaxant, Anaesthetic concerns, Neuromuscular monitoring

Case Report:
A 43 year old female patient presented with proximal muscle weakness, difficulty in breathing and headache. She had scaly lesions over the scalp, pruritis, periorbital oedema and erythematous rashes on the face, neck and both upper limbs. On examination she had characteristic heliotrope rashes on the neck, upper limbs and upper chest region. She was started on systemic corticosteroid therapy of 500 mg methylprednisolone as pulse steroid therapy by a neurologist. She showed a significant improvement in muscle weakness and regression of skin lesions thereby confirming the diagnosis of Dermatomyositis.
On physical examination a right breast lump was noted. Fine needle aspiration (FNAC) was done, reported as $T_2N_1M_X$ carcinoma of right breast with paraneoplastic association. Patient was posted for right modified radical mastectomy under General anaesthesia.

The complete haemogram, Renal function test, serum electrolytes, coagulation profile and urine routine were within normal limits. Chest X-ray was normal. Electrocardiogram showed sinus tachycardia, echocardiography showed ejection fraction of 66% with adequate LV systolic function and no regional wall motion abnormalities. Creatine kinase (CK-MB) was elevated 8.44Units/L. Lactate dehydrogenase (LDH) was also elevated 421 Units/L. Pulmonary function test showed severe restrictive abnormality.

After obtaining informed consent for General anaesthesia (GA), high risk with post operative ventilation; a large bore Intravenous line was secured and preinduction monitors like ECG, NIBP and SpO_2 were connected in the operating room.

Patient was preoxygenated and induced with Midazolam 1 mg, Fentanyl 100 µg, Xylocard 20 mg , Propofol in graded doses upto 120 mg was given and a non depolarising muscle relaxant (NDMR) Atracurium 30 mg was given after checking adequacy of ventilation. Airway was secured with 7.5mm cuffed endotracheal tube, confirmed by auscultation and $ETCO_2$ monitoring. Neuromuscular monitoring was attached. Muscle relaxants were given based on neuromuscular monitoring to a total of Atracurium 100 mg. Anaesthesia was maintained with oxygen, nitrous oxide and Sevoflurane. Post procedure patient was extubated based on neuromuscular monitoring once fully awake. Post operative period was uneventful.

Discussion:
Dermatomyositis is an idiopathic inflammatory myopathy more common in women 2:1. In adults the incidence peaks between 40-50 years. Clinically characterised by proximal symmetric muscle weakness and cutaneous findings such as Gottrons papules, heliotrope rash, shawl or 'V'sign, periungual erythema, telangiectasia and more rarely vesticulobullous formation. Systemic manifestations include interstitial lung disease, dysphagia due to oesophageal musculature involvement, myocarditis and malignant neoplasia concomitant to disease - Paraneoplastic Dermatomyositis[1].

Diagnosis is by myopathic pattern on muscle and skin biopsy as well as on electroneuromyography, muscle inflammation on Magnetic resonance imaging (MRI), elevation of muscle enzyme - creatine kinase (CK), lactate dehydrogenase (LDH), aldolase, aminotransferase and presence of ANA (80%) and myositis specific autoantibodies such as anti-Jo-1 and anti-Mi-2 antibodies[2].

Association of malignancy with Dermatomyositis was first made by Sterz in 1916. Evidence of malignancy with dermatomyositis is about 5 to 7 times greater than that of the general population mimicking paraneoplastic syndrome. The association between Dermatomyositis and cancer might be related to the expression of auto antigens shared by neoplastic and muscle tissue in some patients. Some possible risk factors for the association with neoplasm are as follows: Leukocytoclastic vasculitis; presence of anti-P^{155}/P^{140} or anti-P^{155} autoantibody; excessive elevation of muscle enzyme; male sex; vesiculobullous Dermatomyositis and advanced age at the time of diagnosis[3].

Treatment consists of administration of glucorticoids at immunosuppression dose – Prednisolone 1mg/kg/day and management of neoplasia if present. Corticosteroids could be accompanied by immunosuppressant drugs – Azathioprine or Methotrexate as glucocorticoid savers. High doses of steroids are used in presence of frequent relapses[2]. Dermatomyositis associated with cancer is generally more resistant to corticosteroids and cytokine therapies than idiopathic myositis. Some reports have described improvement of dermatomyositis without immunosuppressive drugs after resection of the cancer[3].

The major concerns of Anaesthesiologist are delayed recovery from muscle relaxant, aspiration pneumonitis, arrhythmias, cardiac failure and steroid supplementation with its complications[4]. Difficult intubation is not a feature of this disease, airway protection and adequate ventilation are the two primary anaesthetic concerns in these patients[5].

Ukei M et al believed that patients with polymyositis are sensitive to NDMR and its use may cause muscle weakness and severe dysrhythmias. They suggested that muscle relaxant should be given under close monitoring using neuromuscular transmission monitor[6]. Atracurium an intermediate acting muscle relaxant is preferred as it is metabolised by ester hydrolysis and non enzymatic degradation-Hoffman elimination[7].

Conclusion:
Patients with Dermatomyositis are sensitive to the effects of neuromuscular blocking agents causing delayed recovery which makes Atracurium a preferred agent with the use of neuromuscular monitoring to prevent perioperative morbidity.

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