Polymyositis Presenting with Respiratory Failure

Jerryl Maclean¹, Raj B. Singh¹ and Zaheer Ahmed Sayeed²

Departments of Respiratory Medicine¹ and Neurology², Apollo Hospitals, Chennai, India

ABSTRACT

Polymyositis is a systemic autoimmune disorder characterised by inflammatory myopathy of the skeletal muscles predominantly affecting the proximal muscles and associated with extra-muscular manifestations like dysphagia and skin involvement. In this case report, we describe the occurrence of diaphragmatic weakness and respiratory failure due to polymyositis with relatively well preserved power in limb muscles. [Indian J Chest Dis Allied Sci 2011;53:229-231]

Key words: Polymyositis, Myopathy, Diaphragmatic weakness, Respiratory failure.

INTRODUCTION

Polymyositis is a type of idiopathic inflammatory myopathy (IIM) which is characterised by proximal muscle weakness and non-suppurative inflammation of skeletal muscle, often accompanied by extra-muscular manifestations. We present a patient with polymyositis who unusually had profound respiratory muscle weakness with relatively well preserved power in limb muscle.

CASE REPORT

A 70-year-old female was admitted with complaints of intermittent fever and cough with scanty sputum for four days. She did not report any other symptom. She was known to have systemic hypertension for the preceding two years and blood pressure was well controlled by medication. In 2008, she had experienced some generalised muscle weakness transiently but had not been investigated. She had been empirically started on oral corticosteroids with which she had apparently improved. The dose of oral prednisolone was gradually tapered and she was receiving 5mg prednisolone per day at the time of her present evaluation. On examination, she was febrile (temperature 102 °F). Her blood pressure was 140/90 mmHg. There were a few crackles in the left infra-scapular region. Central nervous system examination revealed power of 4/5 in all four limbs with no sensory deficit. Other systems were normal. Chest radiograph showed consolidation of the left lower lobe. Laboratory examination revealed neutrophilic leukocytosis. Sputum examination was negative for any pathogens. Urine showed no Legionella antigen. She was diagnosed to have a community-acquired pneumonia and was treated with co-amoxiclav and azithromycin. As the fever settled and chest radiograph done after seven days showed partial clearing of the pneumonia, she was discharged.

Four days after discharge she was re-admitted with breathlessness on lying down and difficulty in swallowing. Power in her arm and leg was 4/5. Her neck muscles were weak but she was able to lift her neck off the bed. Bedside observation revealed hypoxaemia only on lying down. Diaphragmatic weakness was considered likely. This was confirmed by ultrasound examination which showed severely restricted diaphragmatic movements on both sides. Arterial blood gas (ABG) analysis on room air showed partial pressure of oxygen (PaO₂) – 52mmHg; partial pressure of carbon dioxide (PCO₂) – 48mmHg, pH 7.38. Echocardiogram showed normal left ventricular function. Computed tomography (CT) of the chest showed that she still had a consolidation in the left lower lobe (Figures 1 and 2). Neurologist’s advice was obtained. Rheumatoid factor, antinuclear antibody (ANA), anti-double stranded deoxyribonucleic acid (anti-ds DNA) antibody and anti-smooth muscle antibody were negative. Nerve conduction studies in both upper and lower limbs were normal. Electromyogram was not done. Creatinine phosphokinase (CPK) level was 24,300 IU/L. Her hypoxia worsened. The ABG analysis showed PaO₂ – 58 mmHg, PCO₂ – 78mmHg, pH 7.28 indicating type II respiratory failure. Non-invasive ventilation was tried but due to persistent hypoxia, she was intubated and ventilated. She was started on high dose intravenous methyl prednisolone 1g once daily for three days. Her respiratory effort and diaphragmatic movement improved and she was...
weaned off the ventilator and extubated. The ABG analysis reverted back to normal. The CPK level came down to 239IU/L. Intercostal and deltoid muscle biopsies were done. Histo-pathological examination showed degeneration of muscle fibers by inflammatory cells and increase in fibrous tissue which was suggestive of polymyositis (Figures 3 and 4). Along with corticosteroids she was also started on intravenous cyclophosphamide pulse therapy (750mg once every 45 days for 6 doses). The patient was discharged while still on bi-level positive airway pressure (BiPAP) at night. Three weeks later the respiratory effort had improved considerably and BiPAP was discontinued.

**DISCUSSION**

Polymyositis from a clinicopathological perspective is classified under idiopathic inflammatory myopathies which is a heterogeneous group of immune mediated disorders that may present in an isolated form or in association with another autoimmune or connective tissue disease, a malignancy or rarely an infection. Polymyositis is presumed to be an autoimmune-mediated disease secondary to defective cellular immunity. The antibodies being implicated in the pathogenesis are the anti-Jo-1, anti-Mi-2 and anti-signal recognition particle antibodies. The other pathogenic mechanism proposed is T-cell mediated cytotoxic process directed against unidentified muscle antigens. Supporting this mechanism is the presence of CD8+ T-cells, which along with macrophages, initially surround healthy non-necrotic muscle fibers and eventually invade and destroy them. The average overall annual incidence rates for inflammatory myopathy vary from $2.18 \times 10^{-6}$ to $7.7 \times 10^{-6}$. In general, the incidence rates of polymyositis are higher in women. Annual incidence
rates of idiopathic inflammatory myopathies increased with age, ranging from $2.5 \times 10^{-6}$ in people under 15 years of age to $10.5 \times 10^{-6}$ in people over 65 years of age. The characteristic clinical features include non-selective painless proximal muscle weakness which may be of subacute onset in early adult life; associated systemic dysphagia and interstitial lung disease. Muscle usually occurs in the following descending order: pelvic girdle is usually the earliest to be involved, followed by, shoulder, neck muscles (flexors), distal muscle (in severe cases) and pharyngeal muscles. Diaphragm is rarely involved. The characteristic laboratory features include elevated serum CPK levels, electromyography showing the features of myopathic motor unit potentials with or without spontaneous discharges and muscle biopsy evidence of a necrotising inflammatory myopathy. Corticosteroids remain the agents of choice for the initial empiric treatment of inflammatory myopathy. A regimen of oral prednisolone (60mg per day) which is tapered over a period of four weeks until a maintenance dose of 5mg is reached is often used. Immunosuppressive agents are considered in patients with polymyositis who show a partial response to corticosteroid treatment. These agents control myositis or the extra-muscular features of this disease and are effective as steroid-sparing drugs. In case of refractory myositis combination of immunosuppressive agents is used. Prognosis for polymyositis is usually good. A 20-year follow-up study of a 46-patient cohort seen from 1978 to 1999 revealed 5- and 10-year survival rates of 95% and 84%, respectively. In this case report, we document the rare occurrence of early diaphragmatic paralysis and the consequent respiratory failure in one patient with polymyositis.

REFERENCES

Fellowship/Membership Fee

Membership (Annual) : ₹ 500.00
Membership (Life) : ₹ 5000.00
Fellowship : ₹ 5000.00
Enrolment (new members only) : ₹ 1000.00

Payments should be made by Cheque/Demand Draft, drawn in favour of the Secretary, National College of Chest Physicians (I). Please add ₹75/- to all out-station cheques.

Annual membership is valid for one financial year (April-March) only.

Sd/-
Secretary

Fellows and Members of the College are requested to mention their old address also while informing for change of address alongwith their Fellowship/ Membership number to the Secretary, NCCP (I) and/or to Publication Department, IJCDAS to avoid any mistake/delay in mailing the Journal's copy as well as College's information.

Sd/-
Secretary