Case Report

A Case of Polymyositis

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ABSTRACT

Polymyositis (PM) is an idiopathic inflammatory myopathy (IIM) clinically characterized by chronic muscle inflammation accompanied by muscle weakness. PM patients have poor quality of life. There is no specific regional data available about incidence and prevalence of PM. IIMs is largest group of potentially treatable myopathies. Steroids are the first line of treatment in such patients. Most patients with PM improve with immunotherapies, but usually require life-long treatment.

Introduction

The idiopathic inflammatory myopathies (IIMs) are a group of rare, acquired disorders with primary features of muscle weakness and inflammatory lesions identified in skeletal muscle specimens and the largest group of potentially treatable myopathies in children and adults. The incidence together is greater than 4 cases per 100,000 with prevalence in the range of 1432 per 100,000. There is no specific regional data available. The most commonly mentioned types of idiopathic immune-mediated myopathies (IIMs) are : dermatomyositis (DM), polymyositis (PM) and inclusion body myositis (IBM). PM is an idiopathic inflammatory myopathy clinically characterized by proximal muscle weakness, elevations of serum muscle enzymes and additionally in DM by skin abnormalities[1].

Case Report :

A 45 year old female patient, resident of Madhya Pradesh brought by relatives with complaints of insidious onset of weakness in both lower limbs 6 months back followed by gradual increase in weakness in upper limbs, initially patient was able to do routine activities and walk with support, weakness progressed to such an extent that patient was bedridden since last 3 months.

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Patient started complaining of difficulty in swallowing, initially for solid followed by liquid food, since last one and half month she also noticed nasal regurgitation of food. She also developed muscle pain all over the body during last 3 months, when she was bedridden. She was investigated at Jabalpur with nerve conduction studies and serum creatine phosphokinase (CPK) which was 3770 U/L Thereafter she was treated with intravenous steroids for 2 days, following this treatment patient had come to us. She gave no history of hypertension, tuberculosis, fall, trauma, headache, fever, back pain, skin lesion, bladder or bowel involvement, fasciculation, involuntary movements, waxing or waning of symptoms. On general examination patient was thin built, muscle wasting was present in all the four limbs, had pallor, blood pressure was 110/70 mmHg, pulse was 76/min, weight 38 kg, skin rash was absent. Neurological examination revealed normal higher mental function, except for pharyngeal weakness, no other cranial nerve involvement was seen. Hypotonia was present in all four limbs. Power in upper limb was 2/5 proximally and distally it was 3/5. Power in lower limb was 1/5 proximally, distally it was 3/5. Deep tendon reflexes were depressed in all the four limbs. Both plantar responses were flexor. Cardio respiratory and abdominal system examination was within normal limits.

Investigations :

Complete blood count, liver and kidney function test were within normal limits (WNL). Erythrocyte sedimentation rate (ESR) - 20 (Normal 0-22 mm/hr).

Serum Creatine phosphokinase (CPK) value (Normal 26 - 192 U/L) was in falling trend (10th may) 3770 U/L subsequently it was 597 U/L on 7th June, cerebro spinal fluid (CSF) study was done to rule out other causes.

CSF study routine microscopy was WNL.

CSF Total protein	19.4 mg/dl (normal 15 to 60 mg/100 mL)
Glucose	67 (normal 50 to 80 mg/100 mL)
ADA	1.01 (normal <10 IU/L)
Cytology	No significant cytopathology seen

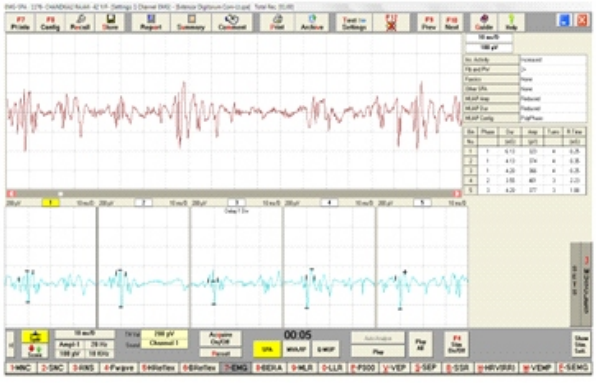
HIV, HBsAg, HCV were negative. Chest X-Ray PA view WNL. Sonography of abdomen was normal. Echocardiography suggestive of type 1 diastolic dysfunction, rest of echocardiogram was WNL.

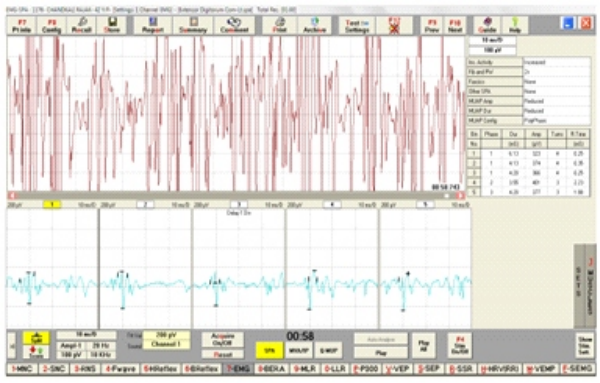
MRI brain (plain) was normal, MRI cervical spine was suggestive of degenerative changes in cervical, thoracic, lumbar vertebrae, no spinal cord compression or spinal canal stenosis was seen. Electromyography (EMG) and nerve conduction velocity (NCV) studies showed increased insertional activity with fibrillation potentials in distal and proximal muscles. Low amplitude, short duration polyphasic motor unit potentials with full interference pattern was seen in left Tibialis anterior, extensor digiticommunis and left biceps brachii muscles, suggestive of myopathy most likely inflammatory type. (muscle biopsy was planned but could not be done as patient went against medical advice because of covid19 pandemic issues).

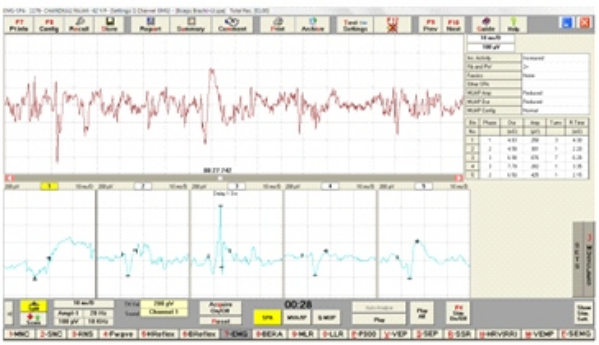
On the basis of patients history, clinical features, raised serum CPK levels and EMG-NCV study findings suggestive of inflammatory myopathy, diagnosis of idiopathic inflammatory myopathy-polymyositis was made and patient was started on oral steroids at the time of discharge but we lost the follow up of patient.

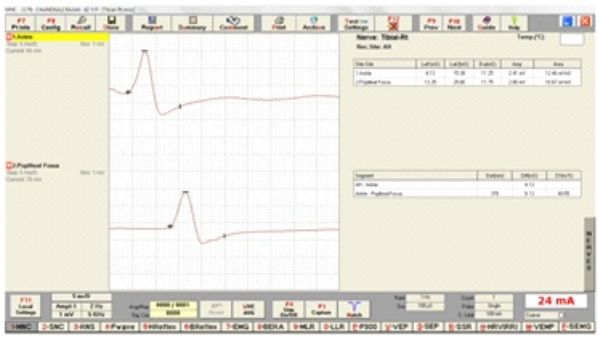
Discussion :

Polymyositis is a diagnosis of exclusion with features of subacute proximal myopathy in adults who do not have rash or other cutaneous manifestations, family history of neuromuscular disease, exposure to myotoxic drugs (statin, penicillamine, zidovudine), involvement of facial or ocular muscles, any endocrinopathies, clinical









phenotype of inclusion body myositis[2]. Although the inciting event of PM is unknown, it has been postulated that some microvascular injury may lead to the release of muscle auto antigens, which are then presented to T-lymphocytes by macrophages in the muscle. T-lymphocytes that have been activated then proliferate and release cytokines such as interferon gamma and interleukin. Interferon gamma promotes further macrophage activation and release of mediators of inflammation such as interleukine-1 (IL-1) and tumor necrosis factor-alpha. The presence of auto aggressive inflammatory cells that surround, enter, and destroy morphologically normal appearing myofibers is the characteristic feature of PM. Most often, patients with non-specific inflammatory cells present in perimysial more often than endomysial locations have been categorized as PM. These inflammatory cells are composed largely of CD8+ T cells and macrophages. When PM is associated with other connective tissue diseases (CTDs) such as Scleroderma, Mixed Connective Tissue Disease (MCTD), Sjögren syndrome, Systemic Lupus Erythematosus it is then known as Overlap Syndrome, it carries good response to treatment. Diagnosis is based on elevations in the blood levels of specific muscle enzymes like CPK, aldolase, transaminases and LDH. CPK levels are always elevated in uncontrolled PM. A variety of auto antibodies may be present in the serum of patients with PM, such as anti-nuclear antibodies (ANA) and antibodies to RiboNucleo Protein (RNP). Magnetic resonance imaging (MRI) can demonstrate early muscle inflammation in PM. Muscle biopsy can demonstrate muscle fibres in various stages of inflammation, necrosis and regeneration, endomysial infiltration by mononuclear cells, capillary obliteration, endothelial cell damage, and increased amounts of connective tissue. PM is commonly associated with myocarditis, interstitial lung diseases (ILD) other CTDs.

The Bohan and Peter criteria for DM and PM : First, rule out all other forms of myopathies

1. Symmetrical weakness, usually progressive, of the limb-girdle muscles with or without dysphagia and respiratory muscle weakness.
2. Muscle biopsy evidence of myositis, necrosis of type i and type ii muscle fibers; phagocytosis, degeneration, and regeneration of myofibers with variation in myofiber size; endomysial, perimysial, perivascular, or interstitial mononuclear cells.
3. Elevation of serum levels of muscle-associated enzymes (CPK, LDH, transaminases, aldolase).
4. EMG triad of myopathy
   A. Short, small, low-amplitude polyphasic motor unit potentials
   B. Fibrillation potentials, even at rest
   C. Bizarre, high-frequency repetitive discharges.
5. Characteristic rashes of dermatomyositis.

Definite PM : all first four elements,
Probable PM : 3 of first 4,
Possible PM : 2 of first 4.

Definite DM : rash plus 3 others,
Probable DM : rash plus 2 others,
Possible DM : rash plus 1 other3.

Goal of treatment : To improve muscle weakness, avoid the development of extra-muscular diseases of the vital organs, improve quality of life.

Treatment : Glucocorticoids are the standard first-line medication for any idiopathic inflammatory myopathy. Initially, prednisone is usually given in a single dose of 1 mg/kg/d, but in severe cases, the daily dose can be divided or intravenous methylprednisolone can be used. The response to therapy should be assessed every 2-4 weeks by monitoring the proximal muscle strength, muscle enzyme levels, and patient functionality. After the initial 6-8 weeks, a slow taper of steroids should begin. Consider a second line agent (usually methotrexate) with glucocorticoids in patients with severe weakness or other organ system involvement (e.g. myocarditis, ILD), those with increased risk of steroid complications (e.g. diabetics, osteoporosis, or postmenopausal women). Intravenous immune globulin (2 g/kg over 25 days; then 1-g/kg every 48 weeks as needed) is often beneficial in patients who

are corticosteroid resistant, especially where there is rapidly progressive or life-threatening progression but its effect is short-lived and repeat infusions are generally necessary every 48 weeks.

Steroid-sparing drugs are used in a steroid responsive patient, the goal is to attain the lowest dose of steroids that will adequately manage the disease or in patients where steroids are contraindicated or are refractory to steroid treatment. Azathioprine (Aza) is usually administered orally at a dose of 1.5-3.0 mg/kg/day. Methotrexate (Mtx) may be administered orally, subcutaneously, or intramuscularly and it is given once a week at doses ranging from 10-40mg.Mycophenolate mofetil is administered orally in doses up to 3g/day. Calcineurin inhibitors: cyclosporine and tacrolimus have both shown some efficacy in the treatment of refractory PM. Cyclosporine is administered orally in doses up to 150 mg twice a day. Rituximab (rtx) is a monoclonal antibody against CD 20+ B-cells, which causes depletion of these cells for 6 months or longer. The optimal dose of rtx is to use 375 mg/m2, infused intravenously once a week for 4 weeks. Cyclophosphamide (ctx) is an alkylating agent that is toxic to lymphopoietic cells. Both T-cells and antibody producing B-cells are affected. This drug may be most useful in patients who have PM with interstitial lung disease. The drug may be given intravenously at 0.8-1.0g/m2/month for several months[4].

Conclusion :

Polymyositis is a life-threatening condition if left untreated, can result in worsening of breathing and swallowing problems. PM is more common in females and most affected muscles are muscles of hips and thighs, upper arms, shoulder, neck. PM can also affect cardiac muscles causing inflammatory myopathy and muscles involved in breathing. The predominant clinical manifestation is proximal muscle weakness. There may be extra muscular involvement such as inflammatory arthritis, Raynaud's phenomenon, myocarditis, and interstitial lung disease. Serum muscle enzymes (CPK) are usually elevated during periods of active disease. A variety of auto antibodies are often found in the serum of PM patients. Characteristic abnormalities are often seen on EMG and muscle MRI. Definitive diagnosis is established by muscle biopsy. The first line agent in treating PM is usually a steroid. Steroid especially prednisolone is very effective in bringing inflammation under control and improving quality of life. Life style modifications like intake of thickened fluids, eat softer or mashed foods, gentle physio exercises are to be followed to improve movement and decrease pain.

References :

1. Greenberg SA, Amato AA. Inflammatory Myopathies. In: Harrison's Principles of Internal Medicine. 20th ed. McGraw Hill LLC; 2000. p. 2590-7.
2. S S. Case Study on Polymyositis. MOJ Biol Med. 2017;1(1):15-6.
3. Leclair V, Lundberg IE. New Myositis Classification Criteria - What We Have Learned Since Bohan and Peter. Curr Rheumatol Rep. 2018;20(4).
4. Hunter K, Lyon MG. Evaluation and management of polymyositis. Indian J Dermatol. 2012;57(5):371-4.