A Unique Case of Dermatomyositis in Young Male Patient

Dr Shyam

Dr Pushpraj Anand

Dr Aswathy Joseph

Dr Arpit Goel

Dr Vinay Yadav

Dr Rajesh Khoiwal

Pacific Institute of Medical Sciences

ABSTRACT

Dermatomyositis is an idiopathic inflammatory myopathy involving proximal muscle weakness and Rash formation.Classification of dermatomyositis for a definitive diagnosis requires characteristic rash and other criteria, such as proximal muscle weakness and muscle enzyme level elevation. Muscle biopsy is central in establishing the diagnosis.In this article, we report a case of dermatomyositis without coexisting cancer which is rare.

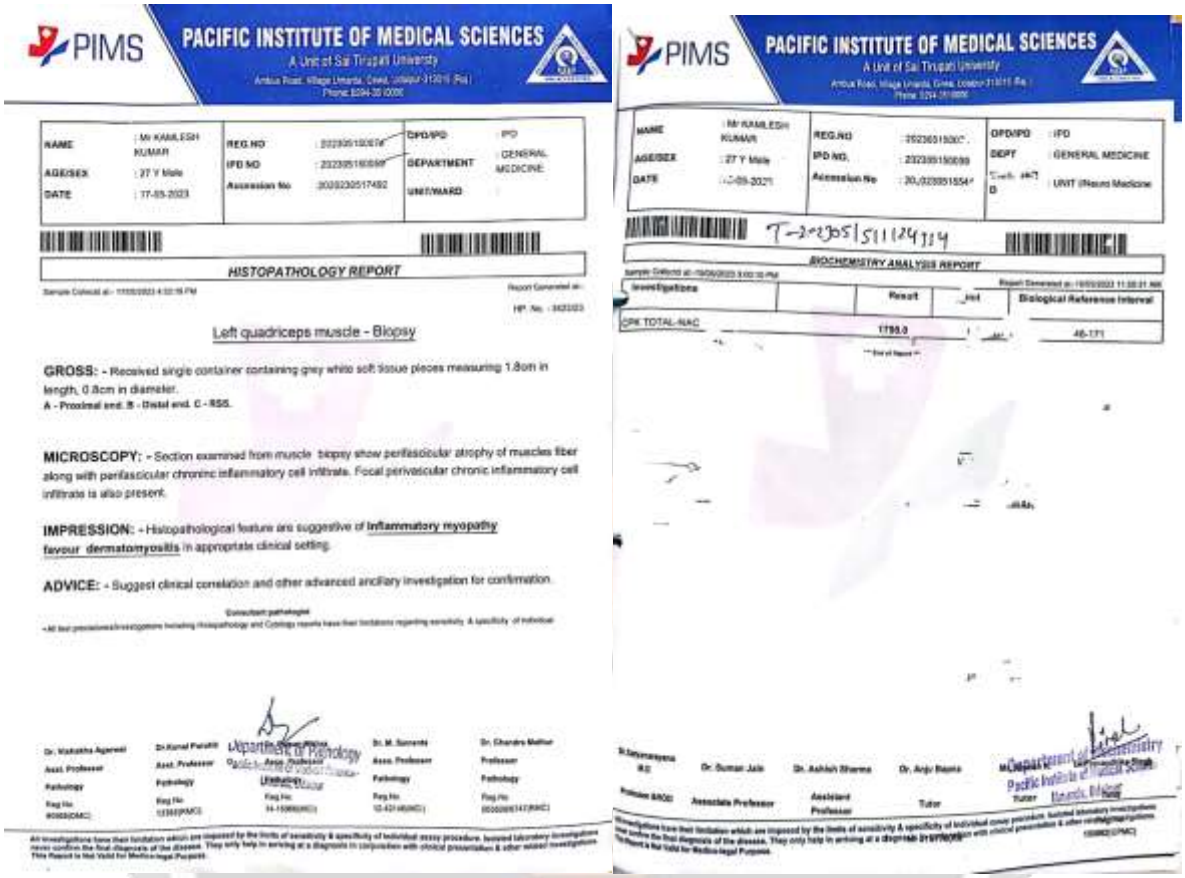
INTRODUCTION

Dermatomyositis is an idiopathic inflammatory myopathy.The average age at diagnosis is 40 years, and almost twice as many women are affected as men, with a prevalence rate of 1 per 100,000 in the general population.The incidence of malignancy is high, especially in the older age groups. Diagnostic criteria include typical cutaneous features, progressive proximal symmetrical muscle weakness, elevated muscle enzymes and abnormal findings from muscle biopsy.We report a male patient with the features of dermatomyositis.

CASE REPORT

A 27 Year old male patient presented with chief complains of myalgia,difficulty in climbing stairs,difficulty in standing from sitting position,difficulty in combing hair since 4 months.weakness was insidious in onset and slowly progressed in such manner that patient needed stick or one man support for standing from sitting position.He also noticed rash formation over malar prominence and both ears since 4 months which was non-pruritic and no any aggravation of rash when exposed to the sun. There was no history of fever, dyspnea, dysphagia and use of any medications. On general physical examination, his vitals were normal.The ear, nose and throat examination was normal. Neurological examination revealed normal higher mental functions and cranial nerves, power of grade 4 at shoulder and elbow joints, grade 5 in wrist joint, grade 4 at the hip and knee joint and grade 5 at the ankle joint. Tone was normal, all deep tendon refl exes were present and plantars bilaterally flexors.The sensory system was within normal limits.The remainder of the neurological examination was unremarkable.The respiratory, cardiovascular and abdominal systems were normal.Investigations revealed Hb-14.2gm%, WBC-8200/cumm, platelets-2.5 lakhs/ cumm, ESR-28 mm/1h. Urine analysis, renal function tests and blood glucose were within normal limits. The liver function test showed ALT-244 IU/L (reference: 0-40 IU/L).creatinine phosphokinase 1798 IU/L (reference: 24-190 IU/l), S. lactate dehydrogenase (LDH)-2070 IU/L (reference: 40-250 IU/L),thyroid function tests were normal and ANA was borderline positive. Chest X-ray,ECG,ultrasound abdomen and pelvis were normal. Electromyography (EMG) showed early and complete recruitment with polyphasic and low-amplitude motor unit action potential, suggestive of inflammatory myopathic process. Muscle biopsy showed perifascicular atrophy of muscle fibre along with perifascicular chronic inflammatory infiltrates and focal perivascular chronic inflammatory cell infiltrate is also present. Based on the above findings of proximal muscle weakness, rash, elevated creatinine phosphokinase, S. LDH, and findings of EMG and muscle biopsy, a diagnosis of dermatomyositis was made.

He was put on prednisolone 60 mg once a day during hospital stay.At present, he is on prednisolone 10 mg/ day maintenance therapy.



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HISTOPATHOLOGY REPORT

Left quadriceps muscle - Biopsy

GROSS: - Received single container containing grey white soft tissue pieces measuring 1.8cm in length, 0.8cm in diameter.
A - Proximal end, B - Distal end, C - RSS.

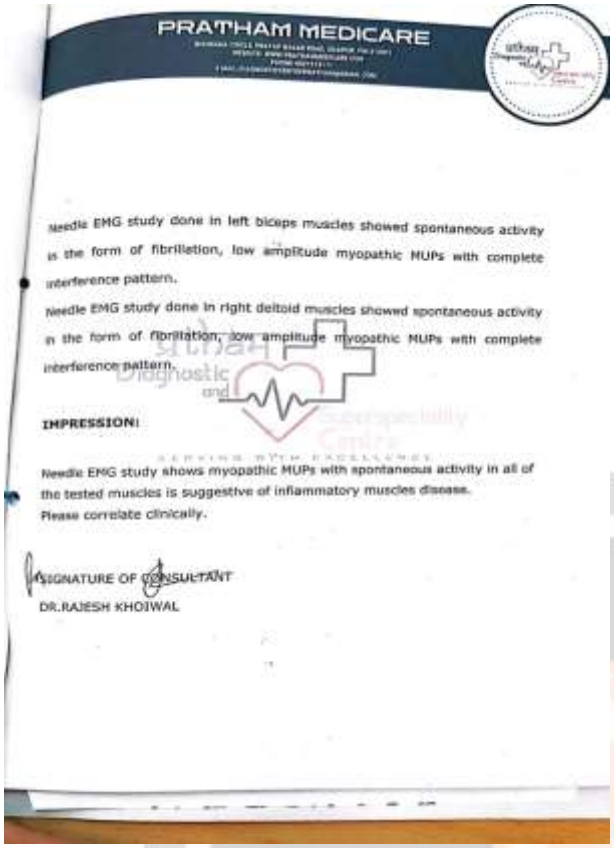
MICROSCOPY: - Section examined from muscle biopsy show perifascicular atrophy of muscles fiber along with perifascicular chronic inflammatory cell infiltrate. Focal perivascular chronic inflammatory cell infiltrate is also present.

IMPRESSION: - Histopathological feature are suggestive of **Inflammatory myopathy favour dermatomyositis** in appropriate clinical setting.

ADVICE: - Suggest clinical correlation and other advanced ancillary investigation for confirmation.

BIOCHEMISTRY ANALYSIS REPORT

Investigations	Result	Unit	Biological Reference Interval
CPK TOTAL-NAC	1799.0		46-171



PRATHAM MEDICARE

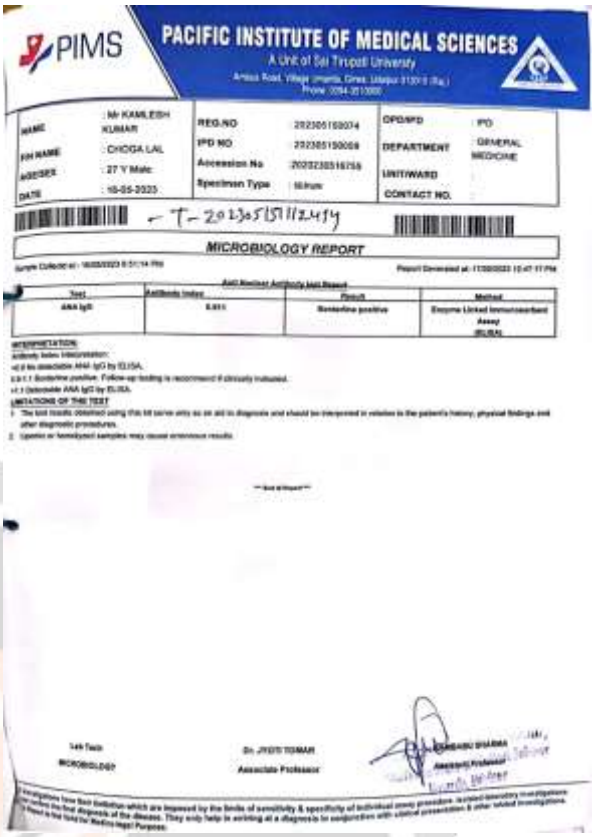
Needle EMG study done in left biceps muscles showed spontaneous activity in the form of fibrillation, low amplitude myopathic MUPs with complete interference pattern.

Needle EMG study done in right deltoid muscles showed spontaneous activity in the form of fibrillation, low amplitude myopathic MUPs with complete interference pattern.

IMPRESSION:

Needle EMG study shows myopathic MUPs with spontaneous activity in all of the tested muscles is suggestive of inflammatory muscles disease.
Please correlate clinically.

SIGNATURE OF CONSULTANT
DR.RAJESH KHOIWAL



PIMS
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NAME : Mr KAMLESH KUMAR
S/H NAME : CHOGA LAL
AGE/SEX : 27 Y Male
DATE : 16-05-2023
REG.NO : 232305150034
IPD NO : 232305150008
Accession No : 2023230516758
Specimen Type : Sputum
OPD/IPD : IPD
DEPARTMENT : GENERAL MEDICINE
UNIT/WARD :
CONTACT NO. :

MICROBIOLOGY REPORT

Test	Antibody Index	Result	Method
ANA IgG	0.811	Borderline positive	Enzyme Linked Immunosorbent Assay (ELISA)

INTERPRETATION:
Antibody Index Interpretation:
<0.8 No detectable ANA IgG by ELISA.
0.8-1.1 Borderline positive. Follow-up testing is recommend if clinically indicated.
>1.1 Detectable ANA IgG by ELISA.
LIMITATIONS OF THE TEST
1. The test results obtained using this kit serve only as an aid to diagnosis and should be interpreted in relation to the patient's history, physical findings and other diagnostic procedures.
2. Lipemic or hemolyzed samples may cause erroneous results.

Dr. JYOTI TOMAR
Associate Professor
MICROBIOLOGY

DISCUSSION

Dermatomyositis is identified by a characteristic rash accompanying or, more commonly, preceding muscle weakness.The hallmark cutaneous findings include a heliotrope rash (blue-purple discoloration) on the upper eyelids in many cases associated with edema and an erythematous rash on the face, neck and anterior chest (often in a V sign) or back and shoulders (shawl sign), knees, elbows and malleoli; the rash can be exacerbated after exposure to the sun and is pruritic in some cases. Characteristic is the Gottron rash, a raised violaceous rash or papules at the knuckles, prominent in the metacarpophalangeal and interphalangeal joints. When chronic, the rash becomes scaly with a shiny appearance. The weakness can be mild, moderate or severe enough to lead to quadriparesis. This patient is classified as classic dermatomyositis. Treatment consists of high-dose prednisone, azathioprine, mycophenolate or methotrexate for steroid-sparing effect, IVIg.

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