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Dermatomyositis - A state of agony

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Abstract

Dermatomyositis is an inflammatory idiopathic myopathy (1) which affects proximal muscle, skin and blood vessel leading to proximal muscle weakness and rashes. It is usually associated with underlying malignancy. It is to be diagnosed based on clinical features, muscle enzyme elevation and biopsy findings (2). We report a case of 32 year old presenting with weakness of all four limbs and skin rashes. On further evaluation patient was diagnosed as Dermatomyositis. There was no underlying malignancy found which is rare, along with that patient was tested negative for antibodies.

Keywords: Myopathy, Weakness, Creatine Kinase.

Introduction

Dermatomyositis is an inflammatory myopathy of idiopathic cause mostly is characterised be inflammatory and degenerative changes of muscle and skin. It may affect any age group but most commonly between 40-60 years of age. Females are affected twice as often as males.

Case Report

A 32 year old male with no significant past history presented with complaints of weakness of bilateral upper limb and lower limb for last 20 days, which was sudden in onset, gradually progressive, more affecting the proximal muscles and no aggravating and relieving factors. History of patchy areas of hypopigmentation present over forehead, arm, forearm, legs for 20 days. These hypopigmented patches were also present in the neck region, burning and itching on sun exposure. Patient gives history of insect bite prior to the initiation of symptoms. No history of high coloured urine/reduced urine outputor dark coloured stools. Not associated with any sensory loss or paraesthesia. On examination, patient was vitally stable. All peripheral pulses were felt equally. Central nervous system examination revealed reduced power of 3/5 in all four limbs. Tone, Deep tendon reflexes were normal. Other systemic examination was also normal. Lab investigations - Hemoglobin 18.7 gm/dL, PCV 55.3%, ESR 3mm/hr,CRP 0.7 mg/dL,LDH

657 IU/L, D-Dimer 2210 ng/ml, CPK total - 8868 U/L , serum Aldolase - 66.60 U/L . Rheumatoid factor was negative. EMG was done which showed myogenic QMVPs, positive sharp waves and myogenic potential - suggestive of Dermatomyositis. Skin biopsy was done from hypopigmented areas and showed peri vascular mononuclear cell infiltration. MRI of thigh was done which showed hyper intense signals in bilateral thigh muscles - likely inflammatory Myositis. ANA , anti-Mi-2 antibodies were found to be negative. In view of elevated CPK, LDH ; skin biopsy, EMG and MRI thigh - shows features suggestive of Myositis, diagnosis of Dermatomyositis is made even in the absence of positive antibodies which can be negative in few patients. Rheumatologist opinion was also sought. Muscle biopsy was not done. Patient was started on IV steroids, mycophenolate mofetil, hydroxychloroquine. Muscle charting was started. Patients symptoms improved within few days of starting treatment. HRCT chest and CT abdomen was done to rule of suspected malignancy and was found to be normal.

Discussion

Dermatomyositis belongs to a group of idiopathic inflammatory myopathy. It is found to involve muscle, skin, blood vessel. Aetiology is usually idiopathic , paraneoplastic or immune mediated. Characteristically the patient present with proximal muscle weakness, cutaneous findings. The hallmark skin manifestation include heliotrope rash on upper eyelids, face, neck, anterior chest (V sign), back or shoulders (shawl sign). These rash may be exacerbated on sun exposure. Gottron rash maybe present which is characteristic , defined as raised violaceous rash present at knuckles especially at MCP, IP joints. Dilated capillary loops at the fingernail base, thick and distorted cuticle is also characteristic.

Other manifestation include constitutional symptoms, dysphagia, interstitial lung disease, hard calcium deposits, micro abscess, Raynaud phenomenon, cardiac involvement like myocarditis, AV block and heart failure.

Various malignancies associated with Dermatomyositis include carcinoma of lung, stomach, colorectal and ovarian, non Hodgkin lymphoma.

Number of criteria has been set for the diagnosis of Dermatomyositis like EULAR criteria, Peter and Bohan criteria.

Peter And Bohan criteria: characteristic skin manifestation and more than or equal to 3 additional features:

- Symmetric proximal muscle weakness
- Elevated muscle enzymes
- Electromyogram suggesting features of inflammatory myopathy
- Evidence of Myositis on muscle biopsy

There are various subclasses of Dermatomyositis : Classic, Amyopatic, Hypomyopathic, Classic amyopathic,etc. (3,4)

Lab investigations reveal leucocytosis; elevated creatine kinase, LDH, aldolase. Antinuclear antibodies may be positive but can be negative in about 1/3rd of patients. Myositis specific antibodies to be tested. Anti-Mi2 antibody is found to be specific for Dermatomyositis.

Muscle biopsy is gold standard in diagnosing Dermatomyositis. It shows atrophy of fibre near border of fascicle, inflammatory cells around fascicle, between fibre and round blood vessel.

Electromyography shows myopathic motor unit action potential, spontaneous activity like positive sharp waves and fibrillations.

Evaluation of extra muscular manifestation include chest X-Ray, imaging for cancer screening like mammography, CT abdomen, colonoscopy.

Treatment include initiation of corticosteroids and steroid sparing immunosuppressive agent like Methotrexate/Azathioprine. Side effects of long term steroids use results in myopathy which in turn worked the patient symptoms. Steroid induced myopathy vs dermatomyositis can be differentiated based testing power of neck flexor, which is preserved in terroir induced myopathy. In such cases, Methotrexate should be tarted and steroids to be slowly tapered off (5). In case of severe refractory disease, intravenous immunoglobulins can be used. Sun protective measures also play an important role.

Our patient was diagnosed as dermatomyositis based on clinical features, elevated CPK and LDH, skin biopsy and EMG findings. Our patient tested negative for both antibody testing and any underlying malignancy.

Conclusion

We conclude this case report by stressing on few points to be remembered like the need for timely diagnosis and initiation of treatment. Dermatomyositis may need not every time be associated with malignancy and antibody positive, as we have discussed in this case report.

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