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Dermatomyositis in An Adult Female Without Malignancy: A Rare Case Report

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Abstract

Dermatomyositis is an idiopathic inflammatory myopathy characterized by proximal muscle weakness and distinct skin manifestations. It is diagnosed by the presence of classical clinical features and elevation of creatine kinase, abnormal EMG findings and muscle biopsy. Timely diagnosis and treatment lead to better lifestyle. We report a case of classical dermatomyositis without malignancy which is rare.

Keywords: Dermatomyositis, Creatine kinase, Myopathy

Introduction

Dermatomyositis is an idiopathic inflammatory myopathy characterized by distinct skin lesions and symmetric proximal muscle weakness. The age of presentation is bimodal with juvenile dermatomyositis presenting between 4-14 years of age and adult dermatomyositis presents between 40-60 years of age.(1) Its prevalence is 1 in 1000 population with female preponderance.(2) Major organs involved are skin and muscles. Cutaneous manifestations include Gottron papules, Gottron sign, Heliotrope rash, Shawl sign, V sign. Muscular manifestations include symmetrical proximal muscle weakness. Involvement of pharynx and esophagus produce dysphonia and dysphagia which is associated with poor prognosis. (1) Systemic manifestations include interstitial lung diseases (ILD) and myocarditis. (1) We report herein a classical case of dermatomyositis.

Case Descriptiona

28-year-old female presented with the complaints of hyper pigmented rash over dorsum of hands, symmetrical proximal muscle weakness of bilateral (b/l) upper and lower limbs and facial puffiness for the last one and a half month. The weakness was progressive and associated with myalgia. The hyper pigmented rashes were present over the hands, abdomen and face. No history of rash over the shoulder, elbow, knee, chest and upper back. No history of fever, weight loss, cough, dyspnea, dysphagia or any sensory loss. On examination, she had peri-orbital edema, malar erythema, hyper pigmented rash over the MCP, PIP joints and periungual area of b/l hands [Figure 1]. Neurological examination –

Muscle bulk was normal, tone decreased in b/l upper and lower limbs, power was 3/5 in proximal muscles of both upper and lower limbs and 4/5 in distal muscles of both upper and lower limbs. Deep tendon reflexes were normal. No cerebellar signs were noted.

Lab investigations

Lab investigations showed raised inflammatory markers and muscle enzymes. Hematological investigations show mild anemia [Table 1] and biochemical investigations show raised liver enzymes [Table 2]. Special Investigations showed CRP – 18.1 mg/l, ESR – 92 mm in Ist hr., CPK-NAC – 12960 U/L, LDH – 4390 IU/L. Thyroid function test was within normal limits (TSH – 2.57 uIU/ml), ECG showed normal sinus rhythm and chest X-ray & HRCT thorax were normal. USG abdomen showed mild hepatomegaly. Urine analysis was normal. Ca-125 was within normal limits. RA factor and Anti-CCP were negative. S. vitamin D was low (13 nmol/L). Anti-HCV, Anti-HAV, Anti-HEV antibodies and HBsAg were negative. HIV was non-reactive. ANA was negative. Immune 17 profile was positive for Mi-2 and PM-Scl antibodies. EMG was suggestive of muscle irritability in the form of increased insertional activity and spontaneous activity with myopathic affection likely inflammatory myositis. Muscle biopsy was taken from left rectus femoris and histopathology showed preserved fascicular architecture, mild variation in fiber size and shape, endomysium showed scattered necrotic fibers invaded and surrounded by macrophages, mild perimysial inflammation, perifascicular atrophic fibers were seen [Figure 2]. The atrophic fibers showed intense NADH activity. All these features are in favor of inflammatory myopathy and dermatomyositis maybe considered in clinical correlation.

Discussion

Dermatomyositis is characterized by specific cutaneous manifestations and symmetric proximal muscle weakness. Cutaneous manifestations are divided into pathognomonic, characteristic, compatible and rare.[1] Pathognomonic manifestations include Gottron papules, Gottron sign and Heliotrope rash. The classical muscular manifestations include symmetric proximal muscle weakness along with the elevation in serum creatine kinase.[1] Esophageal and pharyngeal muscle involvement will produce the symptoms like dysphagia and dysphonia. Dermatomyositis is classified into following subtypes-[3]

i) Classical dermatomyositis – Patient will have cutaneous manifestations with proximal muscle weakness occurring within 6 months of onset of skin disease.

ii) Amyopathic dermatomyositis – There is no muscular weakness even after 6 months of hallmark cutaneous manifestations.

iii) Hypomyopathic dermatomyositis – There is no muscular weakness after 6 months of hallmark cutaneous manifestations but laboratory involvement of myopathy is present in the form of raised muscle enzymes and myopathy on electromyography.

iv) Clinically amyopathic dermatomyositis – It includes both amyopathic and hypomyopathic dermatomyositis.

v) Juvenile dermatomyositis – In this type, onset is before 18 years of age.

Anti- Mi-2 antibody is directed against a nuclear DNA helicase involved in transcription. Generally anti- Mi-2 antibody positive patients presents with classical dermatomyositis. These patients show elevated creatine kinase levels out of proportion to their degree of muscle

involvement. (1) This form of dermatomyositis is responsive to treatment. (4) The prevalence of malignancy in adult dermatomyositis is 20%. Malignancies associated with dermatomyositis are ovarian cancer, colon cancer, melanoma, breast cancer, non-Hodgkin lymphoma. However nasopharyngeal cancer is common in Asian population. Our patient didn't show any evidence of malignancy. (4)Muscle biopsy show perifascicular atrophy, perimysial and perivascular inflammatory infiltrate consisting of macrophages, B-cells, T-cells, plasma cells and dendritic cells.(1)Treatment consists of large dose prednisolone, azathioprine or methotrexate for steroid sparing effect.(3) We started our patient on prednisolone and the patient responded to treatment.

Conclusion

The incidence of dermatomyositis is rare. It can be diagnosed with classical clinical features and myositis specific antibodies. Histopathological examination of the involved muscle confirms the diagnosis. Though disease flares may occur while the patient is on treatment but overall prognosis is better and the patient responds well to steroids and immunosuppressants.

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Legend Tables & Figures:

Table 1: Hematological Parameters

RBCs	4.37 x 10^6 /uL
HB	9.7 g/dl
HCT	33.6%
MCV	76.9 fL
WBC	8.05 x 10^3 /uL
DLC	N-75%, L-14.8%, M-8.4%, E-1.6%, B-0.2%
PLT	315 x 10^3 /uL
INR	1.04

Table 2: Biochemical Parameters

AST	500 U/L
ALT	462 U/L
Bilirubin	0.68 mg/dl
ALP	136 mg/dl
GGT	125 mg/dl
Albumin	3.4 g/dl
T. Protein	6.3 g/dl
Urea	26 mg/dl
Creatinine	0.59 mg/dl
Na	136 mmol/l
K	4.48 mmol/l
Cl	102 mmol/l
Ca	9.4 mg/dl
PO4	4.9 mg/dl
Uric acid	6.6 mg/dl
RBS	78 mg/dl
LDH	4390 IU/L

Figure 1: Gottron sign

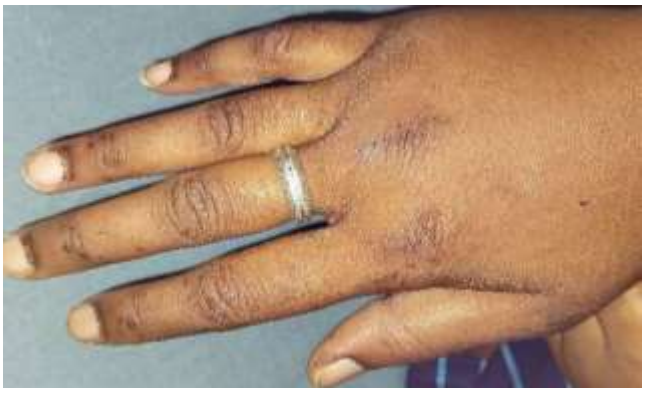

Figure 2: Photomicrograph showing Perifascicular atrophy