

Classical Dermatomyositis: A Case Report

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Abstract: Dermatomyositis is an idiopathic myopathy involving progressive muscle weakness with skin manifestation. Diagnosis is based upon the progressive muscle weakness, skin rashes, elevated serum muscle enzymes, muscle biopsy, and abnormal electromyogram. The incidence of dermatomyositis is rare. In this case study, we report a case of classical dermatomyositis without incidence of malignancy. Timely diagnosis and administration of steroid led to better prognosis of the patient.

Keywords: dermatomyositis, myopathy, creatinine phosphokinase

Introduction

Dermatomyositis (DM), a connective tissue disorder, is an idiopathic inflammatory myopathy characterized by skin manifestation. It occurs in both juvenile and adult forms. The prevalence of dermatomyositis is 1 in 100,000 while women are more affected than men.^{1,2} The diagnosis of dermatomyositis is based on rashes on the skin, progressive muscle weakness, elevated serum muscle enzymes, abnormal electromyogram, and abnormal findings on muscle biopsy.³

Dermatomyositis has a typical cutaneous feature such as erythema over the face, neck, and upper trunk as “neck-line V-shaped.”⁴ Photosensitivity and pruritis are other common cutaneous symptoms. Muscle symptoms include mild to severe weakness, muscle cramps, and fatigue. Dyspnea, dysphagia, and weight loss are frequent signs of disease progression or underlying malignancy.⁴ To the best of our knowledge no cases are reported from Nepal to date. There are no typical signs and symptoms for the diagnosis of dermatomyositis. We reported a female patient with the features of dermatomyositis.

Case Description

A 44-year-old female patient living in Purano-baneshwor, Kathmandu, Nepal was referred from the National Center for Rheumatic disease (NCRD) for laboratory investigation in the Modern Diagnostic Laboratory and Research Center on 24 July 2019, presenting with hyperpigmented erythematous rash all over her body along with V-shaped rashes on the neckline since 2 months. Initial symptoms as mentioned by the patient included itchy rashes and photosensitivity. She had no history of headache, fever, and weight loss with a normal vital test. In addition, she had no surgical as well as medical history of chronic disease. She had progressed symptoms like muscle pain (especially in the left arm and thigh), unable to walk upstairs, inability to stretch hands, unable to turn neck, and difficulty in swallowing food.

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Laboratory Findings

Physical examination showed signs of inflammation supported by laboratory findings, i.e. erythrocyte sedimentation rate (ESR) done by Wintrobe's method found to be 30 mm/1st hour (normal 0–20 mm/1st hour), leukocytosis (14,920 cells/ μ L) with neutrophilia 13,080 cells/ μ L (normal range 2500–8000 cells/ μ L), and carbohydrate reactive protein (CRP) done by nephelometry found to be 25.3 mg/L (normal <3 mg/L). Hematological parameters were measured using Sysmex XN-350 and are presented in Table 1. Biochemical parameters (Table 2) were measured by a fully automated Dimension RxL Max integrated chemistry analyzer that revealed an elevated level of serum creatinine kinase 17,160 IU/L (normal 26–192 IU/L), and normal plasma glucose level and renal function test. Her liver enzymes revealed an elevated level of alanine aminotransferase (ALT), i.e. 370 IU/L (normal 5–45 IU/L), and aspartate aminotransferase (AST) 616 IU/L (normal 5–45 IU/L). Nailfold capillaroscopy of 8 fingers findings showed variant and simple Raynaud's. Urine analysis, chest X-ray, and thyroid function test were normal. Serum lactate dehydrogenase (LDH) 1657 IU/L (reference 25–250 IU/L), Jo-1 antibodies, and dsDNA antibodies were negative while anti-Mi-2 antibody was positive. Raynaud's phenomenon and

Table 1 Hematological Parameters of the Dermatomyositis Patient During Diagnostic Work-Up

Parameter	Result	Reference Range
Hemoglobin	13 g%	(11.9–14.6) g%
Red blood cells	4.55 million cells/ μ L	(4.44–5.61) million cells/ μ L
Packed cell volume	37.7%	(40–50) %
Total leukocyte count	14,920 cells/μL	(4000–11,000) cells/ μ L
Differential leukocyte Count		
Neutrophils	88%	(40–70) %
Lymphocytes	9%	(20–45) %
Eosinophils	2%	(1–6) %
Monocytes	1%	(2–10) %
Basophils	0%	(0–1)%
Platelet count	429,000 cells/ μ L	(150,000–450,000) cells/ μ L
ESR	30 mm/1st hour	(0–20) mm/1st hour

Note: Bold face indicates the abnormal level of hematological parameter.

Table 2 Biochemical Parameters of the Dermatomyositis Patient During Diagnosis

Parameter	Result	Normal Range
Random blood sugar	89 mg/dL	(70–110) mg/dL
Renal function test		
Urea	27.00 mg/dL	(15–45) mg/dL
Creatinine	0.6 mg/dL	(0.6–1.1) mg/dL
Sodium	136 mmol/L	(135–145) mmol/L
Potassium	3.50 mmol/L	(3.5–5.1) mmol/L
Liver function test		
Bilirubin, total	0.7 mg/dL	(0.2–1.3) mg/dL
Bilirubin, conjugated	0.1 mg/dL	(0.0–0.3) mg/dL
Alanine aminotransferase (ALT/SGPT)	370 IU/L	(5–45) IU/L
Aspartate aminotransferase (AST/SGOT)	616 IU/L	(5–45) IU/L
Total protein	6.30 g/dL	(6.3–8.2) g/dL
Albumin	3.50 g/dL	(3.5–5.0) g/dL
Alkaline phosphatase	71 IU/L	(38–126) IU/L
Lactate dehydrogenase	1657 IU/L	(25–250) IU/L
Creatinine kinase	17,160 IU/L	(26–192) IU/L
Magnesium	1.90 mg/dL	(1.6–2.3) mg/dL
Thyroid function test		
Free T ₃	3.12 pg/mL	(2.3–4.2) pg/mL
Free T ₄	1.74 ng/dL	(0.89–1.76) ng/dL
Thyroid stimulating hormone (TSH)	1.17 μ IU/mL	(0.35–5.5) μ IU/mL
Carbohydrate reactive protein (CRP)	25.3 mg/L	<3 mg/L

Note: Bold face indicates the higher level of biochemical parameter.

anti-Mi-2 antibody are highly specific to dermatomyositis. Based on the above findings, a diagnosis of dermatomyositis was made.

She was administered with 50 mg prednisolone with a significant improvement of muscular activities within a month; with a modest fall in serum creatinine kinase level as shown in Table 3 and 25 mg azathioprine once a day as immunosuppressive drugs. At present, she is orally administered 50 mg prednisolone therapy for maintenance therapy and still being followed up.

Table 3 CPK Level After Treatment

At Diagnosis	After 15 Days	After 30 Days	After 45 Days	After 60 Days
17,160 IU/L	10,430 IU/L	6316 IU/L	1108 IU/L	880 IU/L

Discussion

Dermatomyositis is classified into classical dermatomyositis, amyopathic dermatomyositis, juvenile dermatomyositis, hypomyopathic dermatomyositis, clinically amyopathic dermatomyositis, and clinical amyopathic evolving into classic.⁵ The patient showed cutaneous manifestation with evidence of myalgia occurring within 1 month after the onset of skin disease, which is classical dermatomyositis. In the older age group, the incidence of malignancy is higher in dermatomyositis. The common malignancies are ovarian cancer, colon cancer, melanoma, breast cancer, and non-Hodgkin's lymphoma, while in the Asian population nasopharyngeal cancer is common.⁶

The patient showed typical features for diagnosis of dermatomyositis such as progressive muscle weakness, elevated muscle enzyme, dysphagia, and erythematous rashes all over the body with positive anti-Mi-2 antibody, which is specific to dermatomyositis. Treatment is based on high-dose prednisolone until clinical and biochemical remission is obtained. Initially, CPK levels were higher; however, on treatment with steroid, the patient responded well and the CPK level reduced significantly,⁷ which is similar to our study represented in Table 3. Decrease in the level of CPK is a good prognostic factor in dermatomyositis patients.⁸

Mi-2 antigen, nuclear helicase protein, forms part of the nucleosome remodeling deacetylase (NuRD) complex involved in transcription regulation.⁹ Mi-2 is expressed highly in developing hair follicles and embryonic ectoderm, which plays a critical role in the development of epidermis.¹⁰ Mi-2 is also essential for differentiation and renewal of the basal epidermis.¹¹ Immune response to Mi-2 protein leads to the development of skin rashes and production of anti-mi-2 antibody.¹² Anti-Mi-2 is associated with typical skin manifestation where ultraviolet radiation intensity may have an important role in initiating the disease. Sunlight plays a significant role in anti-Mi-2 production through subcellular distribution, expression, and metabolism of the components of Mi-2 antigens.¹³ Mi-2 expression is regulated by UV radiation through protein translation and stability.¹⁴ A patient with anti-Mi-2 antibody presents cutaneous lesions and mild to moderate muscle involvement and responds well to glucocorticoid therapy. Our patient did not have any evidence of malignancy and showed better prognosis. However, recurrence of disease is rare but rashes on skin and retractable myositis can occur, and monitoring and follow-up are needed for the patients.¹⁵

This dermatomyositis patient showed typical clinical manifestation with increased muscle enzymes along with increased liver enzymes. Anti-Mi-2 antibody positive, the patient showed less evidence of malignancy. Proper management and therapy leads to better prognosis of the disease.

Consent

The patient had given written informed consent for the publication of her clinical details. Institutional approval is not required for this case study.

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Author Contributions

All authors contributed to data analysis, drafting and revising the article, gave final approval of the version to be published, and agree to be accountable for all aspects of the work.

Disclosure

The authors report no conflicts of interest in this work.

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