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CASE REPORT

Identifying Dermatomyositis: A Case Report and Review

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ABSTRACT

Dermatomyositis represents an idiopathic inflammatory connective-tissue disease, characterized by inflammation of the muscles and the skin. For the definitive diagnosis of dermatomyositis (DM) requires a characteristic rash and other criteria, such as proximal muscle weakness and muscle enzyme level elevation. DM often overlaps with other connective tissue diseases. A photosensitive rash often is the initial manifestation. There is a high incidence of malignancy in patients with dermatomyositis. In this article, we offer a case report of a patient with DM along with the key points of diagnosis. A physical examination, laboratory tests, antibody tests and muscle biopsy were performed. Muscle biopsy is central in establishing the diagnosis.

Keywords: Dermatomyositis, photosensitive rash, muscle biopsy.

1. INTRODUCTION

Dermatomyositis is an idiopathic inflammatory connective-tissue disease related to polymyositis that is characterized by inflammation of the muscles and the skin (Nikolaos T et al., 2013). It is clinically characterized by progressive symmetrical proximal muscle weakness and a specific skin manifestation. It has extramuscular manifestations such as joint contractures, dysphagia, cardiac disturbance, pulmonary symptoms, and subcutaneous calcifications (Na SJ, Kim SM, Sunwoo IN, Choi YC., 2009). As this disorder shows multi system involvement and association with malignant conditions, early diagnosis and treatment are important to decrease the morbidity of systemic complications. In this article, we offer a case report of a patient with DM. Then we review the key points of diagnosis.

2. CASE REPORT

A 18-year-old female was admited with complaints of weakness in both upper and lower extremities more in proximal than distal muscle weakness, pain in the shoulders. The history also revealed that three months before these symptoms, the patient had a skin rash, mainly over the face, chest (upper front thoracic wall), back and upper extremities accompanied with fluctuating pruritus, The physical examination revealed a purple skin rash on the face and back, oedema of the right upper lid, nail disorders, and muscle weakness, concerning mostly the upper limbs (Figures 1, 2 and 3). Taking into account the above symptoms and signs, the clinical suspicion of dermatomyositis was confirmed by the laboratory findings. Diagnostic study results were as follows: normal electrolyte levels, complete blood cell count, and thyroid-stimulating hormone levels; SGOT 44 IU/L, SGPT 36 IU/L, CK-MB 36.33 IU/L, Serum LDH 1101 U/L, CRP 2.10 mg/L, RA Factor 8 IU/ml, ANA Abs 0.46, Anti ds DNA Ab- 0.89, ASO Titre was 10.36 IU/ml, Anti-Jo-1 antibody test was negative.

Muscle biopsy revealed perifascicular atrophy with muscle fibres showing internalization of nuclei and perivascular lymphoplasmacytic infiltration (Figures 4, 5 and 6). Biopsy from the involved skin revealed hyperkeratosis, thickened basement membrane, vacuolar degeneration of basal keratinocytes along with presence of necrotic keratinocyte i.e. apoptotic bodies (Figure 7) and mild infiltration of chronic inflammatory cells. X-RAY Chest revealed Bilateral Pneumonitis. Electromyography (EMG) was suggestive of primary muscle disease. From the clinical history, laboratory findings, skin and muscle biopsy the diagnosis of Dermatomyositis was given.

3. DISCUSSION

DM is characterized by subacute (over several weeks to months), progressive, and proximal muscle weakness. In our study, patients had subacute, progressive, and proximal muscle weakness. Dermatomyositis and polymyositis may present at any age, with a higher incidence in the age of 50s and 60s and a woman/man ratio of 2:1 (Nikolaos T et al., 2013). It is known that DM has a bimodal age distribution: one peak occurs in children between 5-14 yr of age and a second, larger peak occurs between 45-64 yr of age (Mastaglia FL, Ojeda VJ., 1985). The pathogenesis of dermatomyositis is definitely associated to autoimmune mechanisms. Anti-nuclear antibodies or antibodies against nuclear antigen (ENA) are often positive in patients with myositis but this is not specific as they are also positive in other diseases. However, a series of other auto-antibodies are almost exclusively found in myositis, such as anti-Jo-1, anti-PL-7, anti-PL-12, anti-EJ and anti-OJ. These antibodies target a series of enzymes that help in binding an amino acid to specific transfer RNA. In addition, auto-antibodies against various other intracellular proteins or elements (anti-SRP, anti-Mi-2) may be present in myositis (the majority of patients with anti-SRP have been reported to have polymyositis and anti-Mi-2 auto-antibodies are supposed to be very specific for dermatomyositis) (Bohan A, Peter JB., 1975). Histologically there is an inflammatory infiltration of the muscles by B-lymphocytes and the increased CD4+ / CD8+ T-lymphocyte ratio. This infiltration can be perivascular, around and inside the muscle bundles, and it may even involve solitary muscle cells (Bohan A, Peter JB., 1975). Exact cause not known, may be associated with infection, genetic, immunological factor, and malignancy like nasopharyngeal carcinoma, lung, breast, female genital tract, stomach (Holzmann H, Herz E., 1969). DM is characterized by immune complex deposition in the vessels and is considered to be in part a complement-mediated vasculopathy (Tony Burn, Stephan Breathnach., 2004). Skin manifestation are macular, symmetric, violaceous erythema in a shawl distribution, purplish red periorbital (heliotrope rash) erythema (Valia, R.G., 2008). Gottron papules and Gottron sign are characteristic feature (Sontheimer RD., 1999). Periungual erythema and mechanic hand are also seen.

Muscle Biopsy is the definitive test for establishing the diagnosis of inflammatory myopathy and excluding other neuromuscular diseases (Kasper. Harrison's principle of internal medicine, 2008). The hallmark histopathologic feature of DM is the strongly perifascicular distribution of atrophic, degenerating, and regenerating myofibers (Fig.6). This striking perifascicular pathology has been proposed to result from the destruction of capillaries populating this region. It is thought that a critical depletion of capillaries here could result in localized hypoxia and subsequent myofiber injury (Mammen AL., 2010). Without treatment, vasculitis, myocarditis , interstitial lung disease may develop. Hence, early diagnosis and treatment with corticosteroids, immunosuppressive drugs and high dose immunoglobulin should be given to avoid disability and life threatening complications.

4. CONCLUSION

Dermatomyositis is clinically characterized by progressive symmetrical proximal muscle weakness and a specific skin manifestation. The ability to recognize rash associated with DM is important. Muscle or skin biopsy or both play an important role in confirming the diagnosis.

DISCLOSURE STATEMENT

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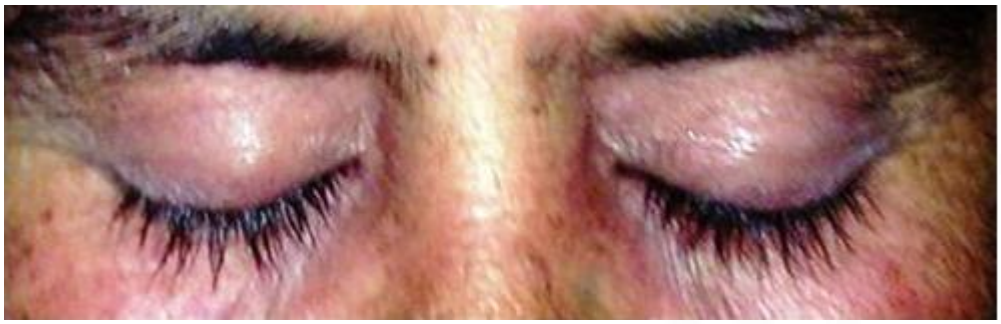

Figure 1
Heliotrope Rash

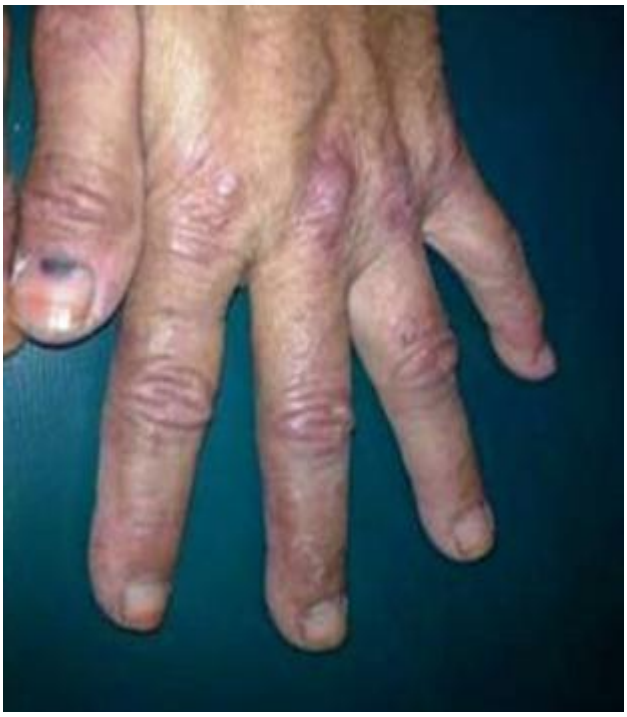

Figure 2
Gottron papules

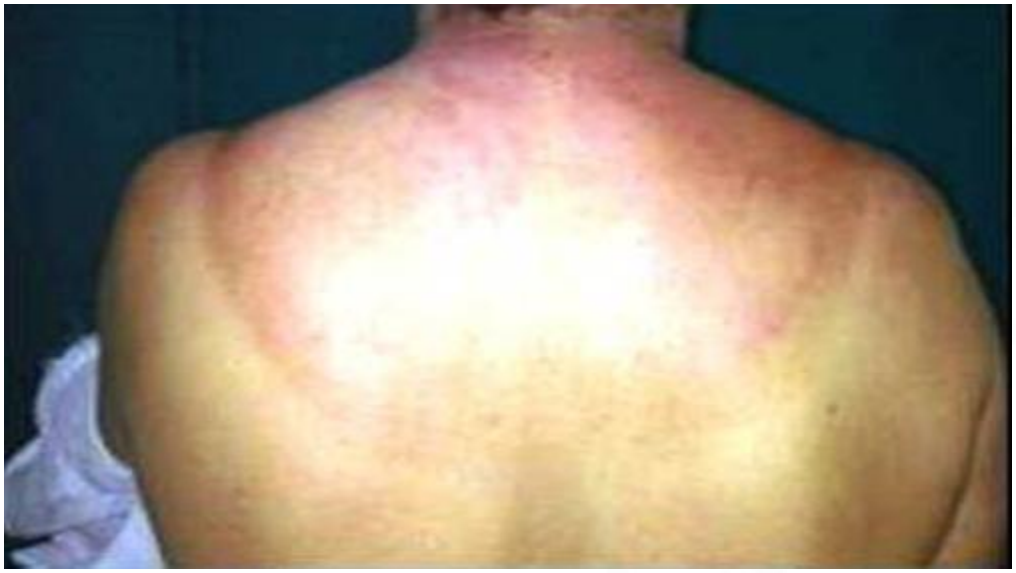

Figure 3
Shawl sign

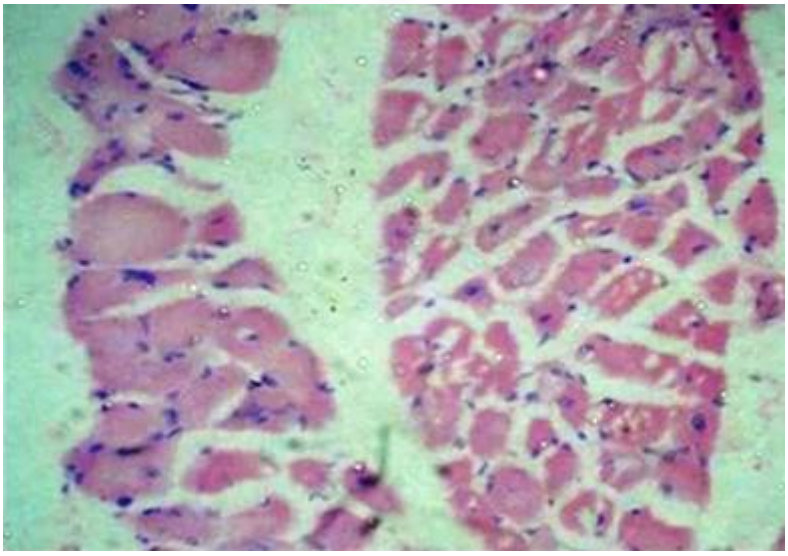

Figure 4
Microphotograph showing perifascicular atrophy (H&E10x)

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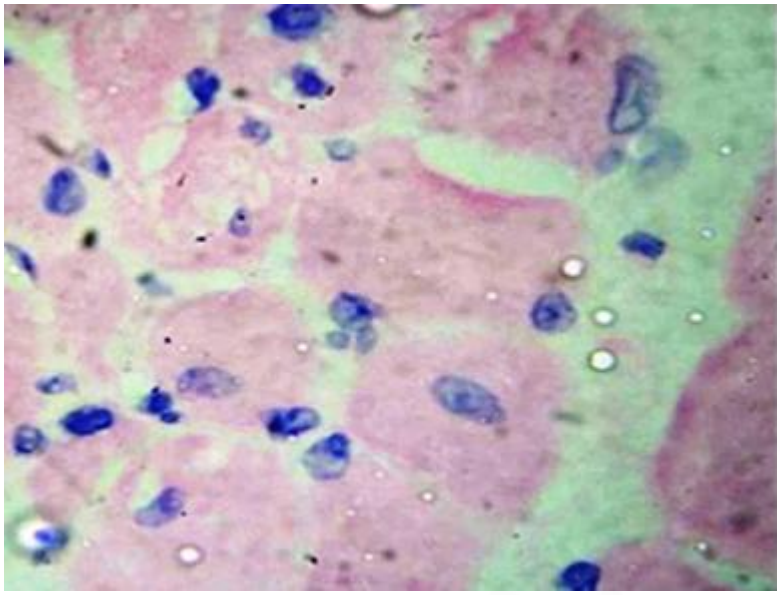

Figure 5
Microphotograph showing internalizationof nuclei (H&E40X)

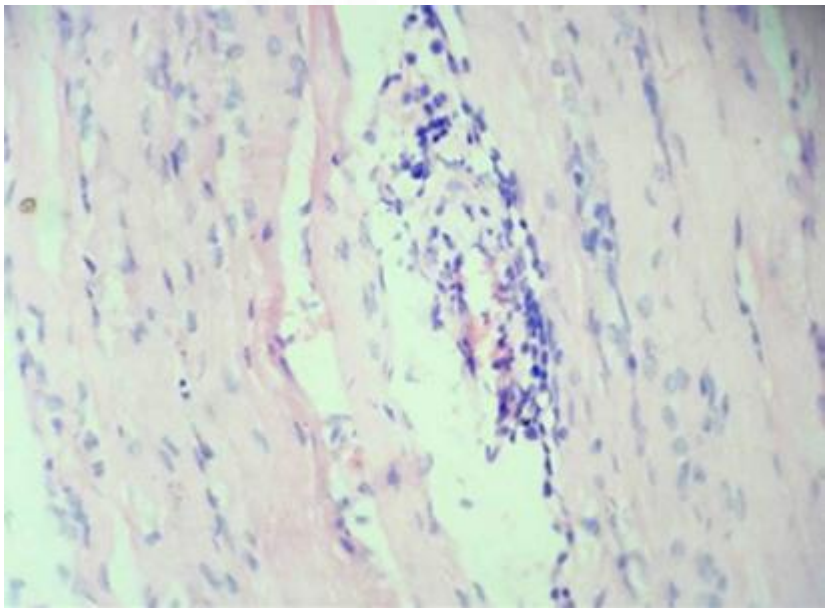

Figure 6
Microphotograph showing perivascular lymphoplasmacytic infiltration (H&E10X)

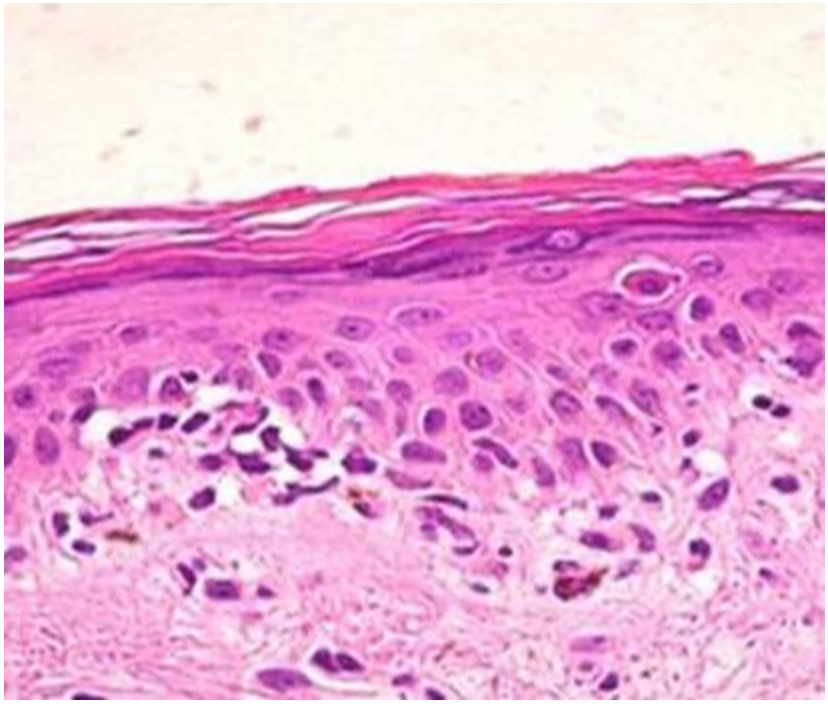

Figure 7
Skin biopsy showing vacuolar alteration of basal cell layer, Necrotic kerationcytes (H&E10X) (apoptotic bodies)