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Case Report

Dermatomyositosis: a rare case report

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ABSTRACT

Dermatomyositis (DM) is a connective tissue disorder and is an idiopathic inflammatory myopathy characterized by skin manifestations. Diagnosis is based on progressive muscle weakness, skin rashes, elevated serum muscle enzymes, muscle biopsy, and abnormal electromyography. In this case study we report a case of classic DM without co-existing malignancy. Timely diagnosis and administration of corticosteroids and physiotherapy led to a better prognosis for the patient. Corticosteroids are the foundation of the treatment. However, the patient remained generally weak and treatment was advised to be continued on discharge.

Keywords: Dermatomyositis, Creatin phosphokinase, Inflammation, Myopathy

INTRODUCTION

Dermatomyositis (DM) is an idiopathic inflammatory myopathy (IIM) associated with proximal symmetric muscle weakness and skin rashes.[1] Dermatomyositis affects approximately 1 in 10,000 people. Women are more susceptible to this disorder than men.[2] Dermatomyositis is classified into classical dermatomyositis, amyopathic dermatomyositis, juvenile dermatomyositis, hypomyopathic dermatomyositis, and clinically amyopathic dermatomyositis.[3] Several substances may cause dermatomyositis, including anti-cancer medications (hydroxyurea, cyclophosphamide), anti-infectives (penicillin, sulfonamides, isoniazid), anti-inflammatory medications (diclofenac, phenylbutazone), statins, D-penicillamine, and vaccinations.[4] Dermatomyositis occurs as a result of the humoral attack on muscle capillaries and arteries.

Dermatomyositis is a condition occurring due to a humoral- mediated attack directed against the muscle capillaries and the endothelium of arterioles. The initiating event is the activation of completer factor-3 (C3), which forms C3b and C4b. This is followed by the formation of the neoantigen C3bNEO and the C5b-C9 membrane attack complex (MAC). The membrane attack complex deposits on vascular walls and causes inflammation. Hypoxic injury to the muscle fibers ensues, leading to atrophy of muscle fibers, particularly the fibers at the periphery that are the most remote and away from the vascular supply. Over time, the capillary density reduces, and muscle fibers start to undergo necrosis and degeneration.[5]

The symptoms of DM include progressive, symmetrical, proximal muscle weakness, as well as characteristic skin lesions like heliotrope rash, Gottron's sign, the V-sign, and the shawl sign (a widespread, flat, reddened area that appears on the upper back, shoulders, and back of the neck or erythema over the posterior aspect of the neck, upper back, and shoulders at times, extending to the upper arms). Additional cutaneous manifestations commonly include vasculopathy changes (i.e. telangiectasias and livedo reticularis), cuticular overgrowth (‘mechanic’s hands’), and poikiloderma.[6]

CASE REPORT

A 25-year male patient of Indian origin came to Gandhi Hospital and was admitted to ICU (intensive care unit) for complaints of low-grade fever, pain, and swelling in bilateral lower limbs, difficulty in lifting the hand above the head, difficulty in lifting the head from the pillow, difficulty in swallowing food, hoarseness of voice, rash on the neck and face and had weight loss since 15 days. He was on NSAID and vitamin B12 medication 15 days before hospital admission. A nasogastric tube was inserted for nutritional support. As the patient's oxygen level was 74% under room air the patient was given oxygen support through non-invasive ventilation mode. After 4 days in ICU, his oxygen levels had improved and he was moved to one of the general medicine wards. On examination, the patient was conscious and coherent. His pulse rate was 105 beats per minute and his blood pressure was 120/80 mm Hg. Neurological examination revealed that the muscle tone was normal in both the right and left upper and lower limbs. According to the medical research council (MRC) grade the patient's upper limb power was 5/5 and his lower limb power was 2/5. The bilateral plantar reflex was normal. In deep tendon reflex knee jerk (L3, L4) was hyperactive and ankle reflex (S1, S2) was absent. Dermatological examination confirmed certain clinical features such as multiple hyper-pigmented and hypo-pigmented maculopapular rashes seen on the nostrils, cheeks, and forehead. Telangiectasia, atrophy, scales, photo-exposed area, and the malar rash had been present on the neck. These clinical features can be seen in Figures 1 and 2.

Laboratory Findings: Biochemical parameters measured reported an elevated level of creatine phosphokinase 79080 IU/l (normal 26–192 IU/l) and elevated creatine kinase 181 IU/l (normal 24-145 IU/l). His liver enzymes were elevated reported were alanine aminotransferase (ALT).

478 IU/l (normal 5–45 IU/l) and aspartate aminotransferase (AST) 833 IU/l (normal 5–45 IU/l). Auto-antibody tests such as ANA-anti nuclear antibody and anti-Jo-1 were negative. Chest X-ray, ECG, and CT chest were normal. A muscle biopsy showed focal peri fascicular atrophy and perivascular lymphocytic infiltration in the perimysium and endomysium. Hence, the diagnosis of dermatomyositis was made based on the aforementioned clinical features and laboratory findings. The treatment given to the patient for 30 days was listed in (Table 1).

Figure 1: Hyperpigmented maculopapular rash

Figure 2: Diffuse scaling on scalp

Table 1. Treatment given to the patient

Brand name	Generic name	Dose	Route	Frequency	Duration (days)
Prednisolone	Prednisolone	40 mg	PO (per oral)	OD	10
Mucopain gel	Benzocaine gel	15 gm	LA (local application)	TID	30
Breezon sunscreen	Spf 30+	50 gm	LA (local application)	TID	30
Liquid paraffin	Liquid paraffin	100 ml	LA (local application)	BD	7
Amoxicillin and potassium clavunate	Amoxicillin and potassium clavunate	1.2 gm	IV (Intravenous)	BD	7

Prednisolone was prescribed to improve the body's immune system by reducing inflammatory attacks on muscles, skin, and other body systems. Further, the dose was gradually reduced to 40 mg, once a day as the patient showed improvement in his body organ functioning. An antibiotic was prescribed to treat any bacterial infections as the patient was admitted with a fever. Physiotherapy was also started to improve his muscle coordination and

strengthening. Mucopain gel (local anesthetic) was given to the patient to suppress the throat pain and irritation as a result of sore throat. A topical application of sunscreen was given to prevent the DM-associated rash from worsening due to UV radiations from sun exposure. Liquid paraffin was advised to treat the dry, itchy skin due to rashes and to prevent any dehydration by keeping the area moist and hydrated. On using the prescribed medications during his hospital stay considerable improvement was observed in the patient. The patient was able to lift his hand above the head and lift the head from the pillow, and pain and swelling were reduced in the lower limbs. The clinical features observed initially due to DM were also seen to be subsided to an extent. The CPK level had improved as shown in (Table 2). As the patient's well-being was seen improving he was discharged from the hospital. Physiotherapy was advised to be continued for his upper and lower extremities' normal functioning. Prednisolone, 10mg once a day was asked to continue on discharge. A protein-rich diet was advised along with regular follow-ups to the hospital.

Table 2: Improved CPK levels on treatment.

At Diagnosis (IU/l)	After 5 days (IU/l)	After 10 days (IU/l)	After 20 days (IU/l)
79080	69470	44470	181

DISCUSSION

Dermatomyositis can be classified into many types as mentioned previously. However, in this case, it is associated with maculopapular rashes and diffuse scaling of the skin regions.

The patient showed typical features for diagnosis of dermatomyositis such as progressive muscle weakness, elevated muscle enzyme, dysphagia, and erythematous rashes in a few areas over the body. Though the anti-Jo-1 test was negative the muscle biopsy revealed perifascicular atrophy which is specific to dermatomyositis.

Treatment is based on high-dose prednisolone until clinical and biochemical remission is obtained. Initially, CPK levels were found to be high but on treatment with steroids, the patient responded well and the CPK level reduced significantly. A decrease in the level of CPK was a good prognostic factor in dermatomyositis patients. This was similar to a study done by Rita Volochayev et al 2012 which demonstrated the same parameters for the diagnosis of DM.[7]

The importance of calcium and Vitamin D3 was of almost importance to this patient. A study done by Kanis et al in 2004 showed that the long-term use of corticosteroids was conferred with an increased risk of fracture.[8] Hence, this patient was prescribed calcium and vitamin D3 supplements on discharge.

De Oliveira et al conducted a study that showed that physical training was an effective method for increasing muscle strength, improving functional impairment, and enhancing the quality of life of patients with systemic autoimmune myopathies.[9] This research was identical to our case where the patient had undergone physiotherapy during his hospital stay and showed improvement in his muscle strength and coordination activities.

It has also been observed that dermatomyositis is associated with a 6-fold higher risk of malignancy in comparison with the general population.[10] However, our patient did not have any evidence of malignancy and showed a better prognosis. As his health was seen improving the patient was discharged from the hospital. Regular monitoring and follow-up were advised to the patient.

CONCLUSION

In this case, we have emphasized the clinical presentation and evaluation of dermatomyositis (DM). It is important to recognize that DM is a unique disease comprising idiopathic inflammatory myositis with a characteristic feature of skin involvement. Some recent studies have shown that these patients can develop lung diseases and malignancies. However, in this patient pulmonary issues and malignancies were not present but due to the risk of development in the later stages of life, he was asked to come for routine check-ups at the hospital. Physical therapy and occupational therapy are also advised to be started at diagnosis. As DM involves muscle inflammation the patient requires monitoring on a long-term basis. Therefore, a multifaceted approach is necessary to treat DM depending on skin, muscle, lung involvement, and as well as any other organs.

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REFERENCES

1. Marvi U, Chung L, Fiorentino DF. Clinical presentation and evaluation of dermatomyositis. Indian J Dermatol. 2012;57(5):375–81.
2. Kalyan M, Kanitkar S, Gaikwad A, Kumar H. Dermatomyositis: a case report. J Mahatma Gandhi Inst Med Sci. 2016;21(1):53–5.
3. Gerami P, Schope JM, McDonald L, Walling HW, Sontheimer RD. A systematic review of adult-onset clinically amyopathic dermatomyositis (dermatomyositis sine myositis): a missing link within the spectrum of the idiopathic inflammatory

myopathies. J Am Acad Dermatol. 2006;54(4):597–613.
4. Dourmishev AL, Dourmishev LA. Dermatomyositis and drugs. Adv Exp Med Biol. 1999;455:187-91.
5. Krathen MS, Fiorentino D, Werth VP. Dermatomyositis. Curr Dir Autoimmun 2008;10:313-32.
6. Garcia FCL, Collado CR, Hernandez RJ, Alejandro EAG, Andrea VM, Guillermo VS, et al. Dermatomyositis and Breast Cancer: A Case Report and Review. Med Case Rep J. 2018;1:101.
7. Volochayev R, Csako G, Wesley R, Rider LG, Miller FW. Laboratory Test Abnormalities are Common in Polymyositis and Dermatomyositis and Differ Among Clinical and Demographic Groups. Open Rheumatol J. 2012;6:54-63.
8. John KA, Helena J, Anders O, Olof J, Chris De L, Melton J, et al. A meta-analysis of prior corticosteroid use and fracture risk. Journal of bone and mineral research 2004;19:893-9.
9. De Oliveira DS, Giovani MR, Rodrigues LF, Katsuyuki SS. Physical exercise among patients with systemic autoimmune myopathies. Adv Rheumatol. 2018;58(5).
10. Olazagasti JM, Baez PJ, Wetter DA, Ernste FC. Cancer risk in dermatomyositis: a meta-analysis of cohort studies. Am J Clin Dermatol. 2015;16(2):89-98.

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