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Dermatology

Dermatomyositis with Positive Anti-TIF1 Antibodies: A Severe Case Presentation

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Abstract | **Case Report**

Dermatomyositis (DM) is a rare, primary inflammatory myopathy affecting all ethnicities. It's a chronic autoimmune disease characterized by progressive, symmetrical proximal myopathy and classic cutaneous manifestations. Its association with malignancy necessitates a thorough search for underlying neoplasia in all cases, especially in older patients.

Keywords: Dermatomyositis, Autoimmune disease, Inflammatory myopathy, Malignancy, Cutaneous manifestations.

INTRODUCTION

Dermatomyositis, the most common inflammatory myopathy, is a chronic autoimmune disease presenting with both cutaneous and muscular involvement. Its strong association with malignancies and potential for pulmonary involvement contribute to the severity of the disease [1]. We report the case of a patient with severe dermatomyositis and positive anti-TIF1 antibodies, without underlying malignancy, who experienced a fatal outcome.

CASE PRESENTATION

We present the case of a 60-year-old male with no significant past medical history, hospitalized for the management of dermatomyositis with significant muscle involvement. On examination, he presented with bilateral periorbital erythema and edema (Figure 1), a V-sign erythema on the décolletage, and erythema on the upper back and outer aspects of the arms, forming the classic Shawl sign (Figure 2). He also exhibited Gottron's papules and erythema, particularly over the metacarpophalangeal joints (Figure 3). The holster sign, characterized by erythema on the outer thighs, was positive (Figure 4). Mechanic's hands were also noted.

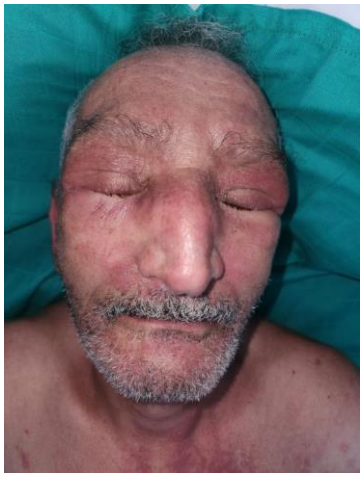

Figure 1: heliotrope rash

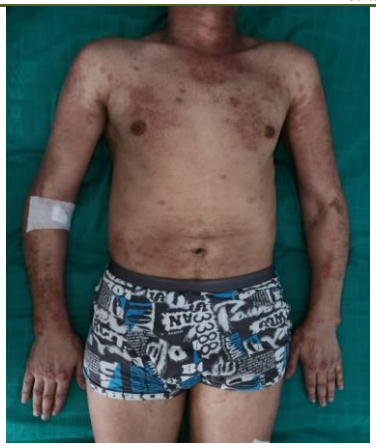

Figure 2: V-sign rash on the décolletage and erythema on the extensor surfaces of the arms (Shawl sign). Gottron's sign is also noted

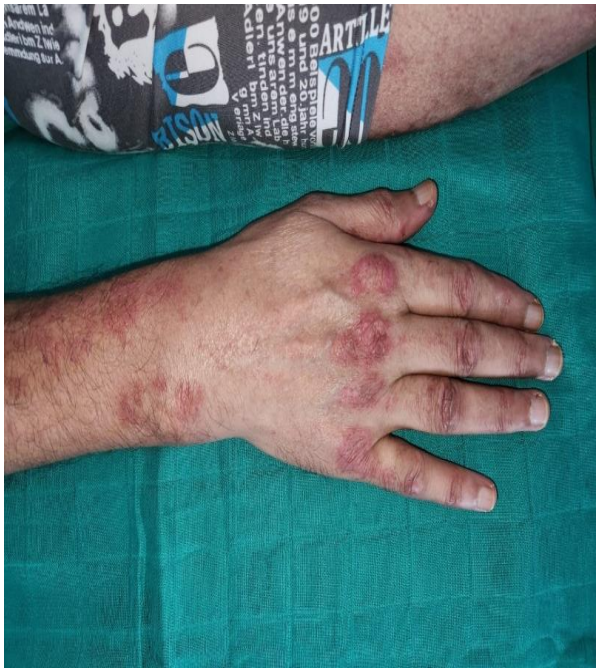

Figure 3: Gottron's sign: Metacarpophalangeal and proximal interphalangeal erythema associated with Gottron's papules

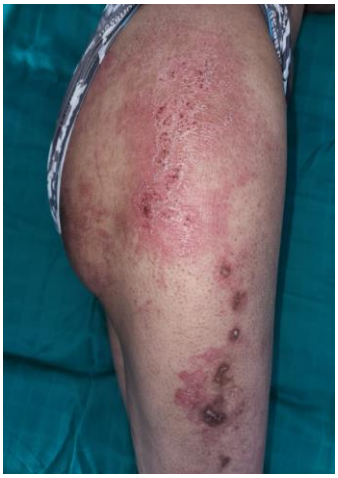

Figure 4: Holster sign: Erythema of the outer Thigh

Paraclinical investigations revealed an inflammatory syndrome with an erythrocyte sedimentation rate (ESR) of 50 mm/h and C-reactive protein (CRP) of 38 mg/L. Muscle enzyme levels were elevated, with creatine phosphokinase (CPK) at 362 IU/L (2x normal), lactate dehydrogenase (LDH) at 472 IU/L (2x normal), and alanine aminotransferase (ALT) at 68 IU/L. Anti-TIF1 antibodies were positive. Electromyography demonstrated active myopathic involvement in all four limbs, predominantly in the proximal muscles of the upper extremities. A muscle biopsy confirmed inflammatory myositis. Paraneoplastic workup, including a normal CT scan, was negative for any evolving malignant lesions.

The patient was started on corticosteroids at 1.5 mg/kg/day, along with adjuvant methotrexate 15 mg/week. Initially, he showed clinical improvement and normalization of muscle enzymes, with CPK decreasing to 53 IU/L. Corticosteroid dosage was gradually tapered. However, the patient's symptoms worsened after 5 months, leading to his demise.

DISCUSSION

Dermatomyositis (DM) is an autoimmune disease characterized by constant and typical cutaneous involvement, inconsistent muscle involvement predominantly affecting the girdles, and also extra-cutaneous and extra-muscular presentations (associated pulmonary or joint involvement). DM can pose diagnostic and nosological challenges with other inflammatory myopathies, especially when cutaneous involvement is atypical or muscle involvement is absent. It is often a severe disease due to its spontaneous evolution, functional impact, potential association with cancer, and treatment-induced complications [2].

DM is a rare condition, with 5-10 new cases per million inhabitants in adults and 1-3 cases per million children. DM affects all age groups, with a clear female predominance (sex ratio of 1.5-2:1). Several highly characteristic clinical aspects of the disease are present in 70% of cases and are sufficient to confirm the diagnosis, even without muscle involvement [3]. The cutaneous manifestations of DM are characterized by specific lesions such as Gottron's papules, Gottron's sign, heliotrope periorbital edema, V-sign erythema on the décolletage, Shawl sign (shoulders), and the holster sign (outer thighs and hips). Less common lesions include mechanic's hands, flagellate erythema, panniculitis, and non-specific lesions (Raynaud's phenomenon, pruritus, and photosensitivity). The histological appearance of cutaneous lesions is non-specific.

Muscle involvement can be the initial presentation of the disease, sometimes severely and abruptly, leading to major functional impairment of proximal muscles, predominantly and symmetrically affecting the shoulder and pelvic girdles. This results in difficulty or inability to raise the shoulders, comb hair, or rise from a chair without using the arms (stool sign, comb sign). Muscle enzymes may be normal or moderately elevated in amyopathic DM, with creatine phosphokinase (CPK) being the most specific indicator of striated muscle involvement.

Specific autoantibodies are of limited diagnostic value but are important prognostically. Each specific DM-associated autoantibody is linked to particular clinical (muscular, cutaneous, and/or systemic), biological, histological, and prognostic features [4]. In adults, the presence of anti-transcriptional intermediary factor 1 (anti-TIF1γ) antibodies is strongly associated with cancer [4]. Cancer-associated DM is more common in older patients, and the cancer may be discovered before, at the time of, or during the evolution of dermatomyositis. Age over 45 years, male sex, dysphagia, cutaneous necrosis, histological evidence of vasculitis, elevated ESR or CRP, severity of muscle involvement, and the presence of anti-TIF1γ antibodies [5] are criteria associated with an increased risk of cancer.

The choice of treatment in DM is linked to disease severity. Corticosteroids have revolutionized the prognosis, reducing mortality from 50% to 14%. The addition of an immunosuppressant, particularly methotrexate, should be considered early to reduce the toxicity of prolonged high-dose corticosteroid therapy in children or adults [6].

CONCLUSION

Dermatomyositis can precede neoplasia in 60-80% of cases, accompany it, or be revealed by it. Predictive factors for cancer association include advanced age, male sex, presence of cutaneous necrosis, erythroderma or intense pruritus, severity of muscle involvement, and positive anti-TIF1 antibodies. The presence of anti-TIF1 antibodies in a patient with DM warrants comprehensive systematic cancer screening, which should be repeated if initial investigations are negative. Advanced age, malignancy, dysphagia, and muscle involvement are associated with an increased risk of mortality.

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