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CASE REPORT

Poikiloderma, a rare initial presentation of dermatomyositis

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ABSTRACT

Dermatomyositis (DM) is a clinically heterogeneous idiopathic inflammatory myopathy that can present diagnostic challenges. The classic form combines characteristic cutaneous signs with bilateral, symmetrical proximal muscle weakness. Poikilodermatomyositis is a specific variant characterized by the presence of poikiloderma and inflammatory myopathy. While poikiloderma is rarely the initial manifestation of DM—typically appearing in advanced photosensitive stages following erythroedema—only a few such cases have been reported. This case report discusses the clinical presentation and therapeutic management of this rare condition. A 27-year-old patient presented with progressive proximal muscle weakness primarily affecting the shoulder and pelvic girdles, accompanied by dysphagia, dysphonia, and a weight loss of 15 kg over three months. Clinical examination revealed generalized poikiloderma, leading to significant skin discoloration, and marked amyotrophy with a Manual Muscle Testing (MMT) score of 30/75. Laboratory investigations showed elevated inflammatory markers and muscle enzymes; however, autoimmune and myositis-specific antibodies were negative. Electromyography was consistent with inflammatory myositis. Upper airway endoscopy revealed vocal cord involvement. Extensive malignancy screening, including imaging, endoscopies, and tumor markers, was negative. The patient was treated with prednisone at an initial dose of 2 mg/kg/day for two months, followed by a gradual tapering. This regimen resulted in a significant improvement in both cutaneous lesions and muscle strength. This case highlights an atypical clinical presentation of dermatomyositis, where generalized poikiloderma was the prominent initial feature. Awareness of this clinical entity is essential, even in the absence of other classic dermatological signs, to prevent misdiagnosis and improve the patient's long-term prognosis.

Keywords: Dermatomyositis, Poikiloderma, Poikilodermatomyositis, Inflammatory myopathy, Gottron's sign, Heliotrope rash.

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1. INTRODUCTION

Dermatomyositis (DM) is an idiopathic inflammatory myopathy that is clinically heterogeneous and that can be difficult to diagnose (1). In its typical form, it combines the presence of characteristic skin signs and bilateral, symmetrical muscle weakness predominantly affecting the proximal muscles. Gottron's sign, heliotrope rash, periungual abnormalities, and flagellate erythema are distinct skin lesions and common dermatological features (2). More uncommon skin manifestations of dermatomyositis are described in the literature. Poikiloderma vasculare atrophicans refers to mottled hyperpigmentation and hypopigmentation of the skin (poikiloderma) with in-between telangiectases (vasculare) and areas of atrophy (atrophicans) (3). Poikiloderma is rarely the first sign of DM and is observed in advanced photosensitive forms, after erythroedema (1).

On reviewing the literature, we found that only a few cases are reported on poikiloderma in dermatomyositis. This observation enables us to explore this rare manifestation of DM. We discuss the clinical presentation and therapeutic approach.

2. CASE PRESENTATION

A 27-year-old patient developed myalgia with asthenia and progressive proximal muscle weakness predominantly in the shoulder and pelvic girdle muscles for several months. The patient reports stiffness in both shoulder joints, dysphagia with weight loss of 15 kg in three months, recent dysphonia, and a change in skin color, which he says has become darker. Clinical examination revealed heterogeneous speckled pigmentation with white and red macules and skin atrophy indicative of poikiloderma (figure 1). There was marked amyotrophy with muscle testing at 30/75. Otolaryngological examination revealed that the vocal cords were red and thick.

Figure 1. Poikyloderma of trunk and the legs.

Biology: microcytic hypochromic anemia; accelerated erythrocyte sedimentation rate ; inflammatory syndrome ; lactate dehydrogenase (LDH) levels three times higher than normal, while creatine phosphokinase (CPK) levels were twice normal; serology tests for hepatitis B and C and HIV were negative. Antibodies (anti-Mi2, anti-MDA5, anti-TIF1Y, anti-SAE1/2, anti-NXP2, anti-nuclear) and tumor markers (CA19/9, CA 125, ACE) were negative. Immunoglobulin electrophoresis was normal. Electroneuromyography (ENMG) revealed myogenic involvement. Other tests: upper digestive tract fibroscopy, electrocardiogram (ECG), Doppler echocardiography, and thoracoabdominal-pelvic CT scan—were normal.

The diagnosis of poikilodermatomyositis was made, and the patient was placed on prednisone at an initial dose of 2 mg/kg/day for 2 months, followed by a gradual reduction over 22 months, resulting in a marked improvement in muscle power and lightening of the skin (figure 2). Verbal informed consent was obtained from the patient forpublication of his case details.

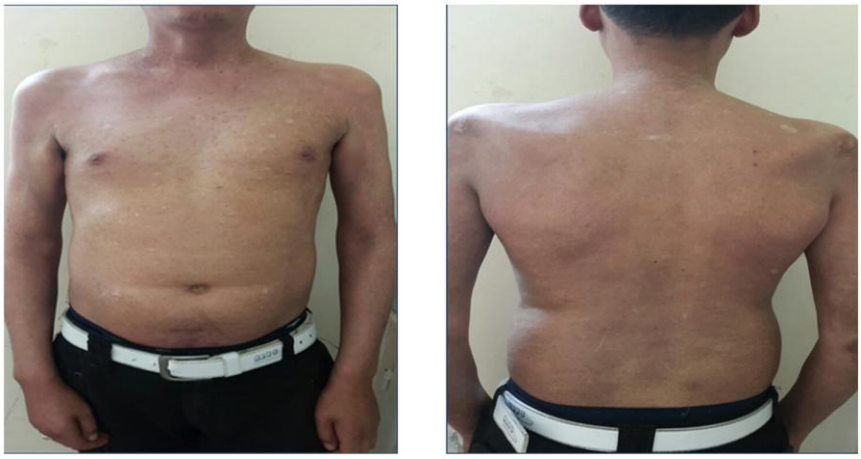

Figure 2. Evolution under treatment.

3. DISCUSSION

Dermatomyositis is a rare disease that may affect the skeletal muscles and the skin (4). In the absence of characteristic dermatologic findings or myopathy, DM can be difficult to diagnose. In our patient, the diagnosis was based on Bohan and Peter criteria, formulated in 1975 (5) and EULAR/ACR (2017) (6). He had a cutaneous rash (poikyloderma), proximal muscle weakness, elevated serum muscle enzymes (CPK) and myopathic changes on electromyography.

Specific rashes of dermatomyositis include heliotrope rash, Gottron papules, nailfold changes, and shawl sign (7). However, the full spectrum includes rashes throughout the body. Acquired poikiloderma is an uncommon sign of dermatomyositis that combines erythema, telangiectasia, pigmentary disorders such as hypo- or hyperpigmentation, and atrophy. They are mainly located on the upper part of the trunk (shoulders, upper back, neckline) and buttocks. Most often they result from a chronic evolution of purplish macules, either spontaneously or following treatment, or constitute advanced photosensitive forms (8). Rarely, they may be the main clinical presentation of the disease, thus defining poikilodermatomyositis, as in our case. This rare condition was first described in 1906 by G. Petges and Clejat under the name « atrophic sclerosis of the skin with generalized myositis » (9) in a patient with myositis, cutaneous atrophy, and poikiloderma.

Since the first reported case, there have been a few publications of poikiloderma associated with other skin manifestations, such as Raynaud phenomenon, scleroderma-like skin, muscle weakness, and joint stiffness (10). Perales-Martinez et al. described a female patient who initially had poikiloderma and later developed heliotrope erythema, periorbital edema, Gottron's papules, proximal muscle weakness, loss of weight, and interstitial lung disease (11).

In other cases, poikilodermatomyositis was associated with subcutaneous calcification (12) and erythroderma (13). This condition is characterized by widespread erythema and scaling affecting over 80% of the skin's surface area (14) and may be life-threatening. The diagnosis is discussed with radiodermatitis, epidermotropic cutaneous T-cell lymphoma, and, more rarely, lupus erythematosus or hereditary diseases (congenital poikiloderma, Zinser-Cole-Engman syndrome).

Others uncommon skin manifestations of dermatomyositis described in the literature are ichthyosis, panniculitis, lichen planus-like lesions, porcelain white atrophic scars, vesicle and bullae formation, follicular hyperkeratosis, malakoplakia, and papular mucinosis (15). Dermatomyositis in this case is often discovered late, mainly in the absence of myositis or other specific cutaneous signs, making the prognosis severe.

The risk of developing malignancy is highest within a year of diagnosis and remains elevated for ≤ 5 years (16). Malignancy risk is also increased for males and those > 45 years of age at the time of diagnosis (17). Screening studies for occult malignancies have been negative, and the patient made a good recovery with steroid treatment.

4. CONCLUSION

This case illustrates a rare clinical presentation of dermatomyositis initially presenting as poikiloderma. Lack of awareness of this clinical entity, in the absence of other specific signs of dermatosis, can avoid misdiagnosis and jeopardize the patient's prognosis.

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