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## CASE REPORT

Female patient, 50 years old, presenting asymptomatic erythematous lesions for two months, initially in the arms, which have spread to thighs, forearms and anterior chest. She complained of photosensitivity and difficulty in climbing stairs. A month ago, she received a diagnosis of breast cancer (invasive ductal carcinoma, stage-IIIb). On examination, it was observed: important periocular edema, reddish-wine-colored; erythematous-violaceous papules on the anterior thighs, arms, forearms, knees, elbows, metacarpophalangeal and interphalangeal joints; cervical poikiloderma in the upper chest and presence of periungual telangiectasia (Figures 1 and 2). The right breast was erythematous, hardened, with cutaneous infiltration and nipple retraction. She also presented right axillary lymphadenopathy and reduced muscle strength in all four limbs (++ / 4). Laboratory tests: creatine phosphokinase (CPK) 4092 UI/ml (normal: <110 UI/ml). Autoantibodies: negative. Electroneuromyography with myopathy pattern. Histopathology: hyperkeratosis, necrotic keratinocytes, intense edema and lymphohistiocytic infiltrate in the papillary dermis, with negative direct immunofluorescence.

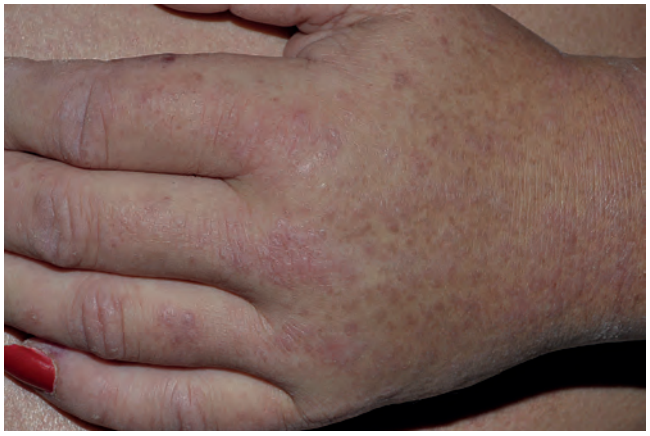

**FIGURE 1:** Erythematous-violaceous papules on the back of the hand with greater intensity on the back of the metacarpophalangeal and interphalangeal joints (Gottron's sign)

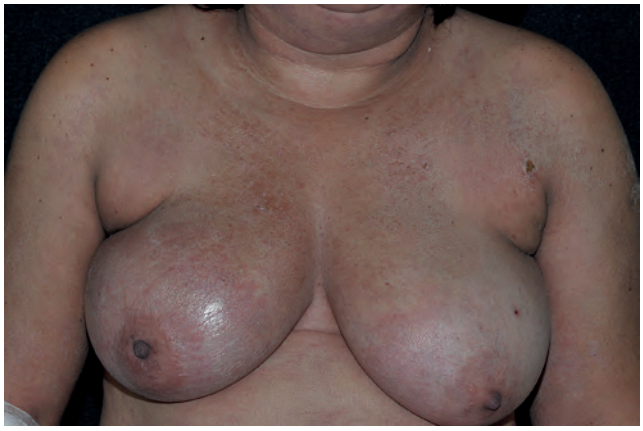

**FIGURE 2:** Poikiloderma of the upper chest, photosensitivity and neoplastic involvement of the right breast: erythema, skin infiltration and nipple retraction

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**Chart 1:** Diagnostic criteria of dermatomyositis. Final diagnosis: major criterion + 3 small criteria. Probable diagnosis: major criterion + 2 small criteria. Possible diagnosis: major criterion + 1 small criterion

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|---|---|
| **Major criterion (mandatory)** | Typical skin rash |
| **Small criteria** | Symmetrical proximal muscle weakness |
| | Muscle biopsy revealing myositis |
| | Increase of skeletal muscle enzymes |
| | Myopathy pattern on electromyography |

## DISCUSSION

Dermatomyositis (DM) is a rare inflammatory myopathy, autoimmune, which affects twice as many women as men. It presents two incidence peaks, 5-14 and 45-65 years, and can occur at any age.[1] The diagnosis depends on the identification of specific criteria (Chart 1).

The characteristic skin lesions are Gottron's papules: erythematous-violaceous lesions on metacarpophalangeal or interphalangeal joints, elbows or knees; shawl sign: poikiloderma in sun-exposed areas (neck, back, shoulders); heliotrope: erythematous-reddish stain and periorbital edema; periungual telangiectasia and "mechanic's hands": peeling, fissures, hyperkeratosis and symmetrical hyperpigmentation, non-pruritic, on the palms. Less common manifestations are facial edema, cuticular dystrophies, photosensitivity, calcinosis, erythroderma, cutaneous vasculitis and panniculitis. Patients present with progressive symmetric proximal weakness, often associated with myalgia.[1-3]

Laboratory tests show CPK levels directly related to the degree of muscle damage. Electromyography shows motor units of small amplitude, slight polyphase potential, bizarre repetitive discharges with high frequency, positive sharp waves and muscle fibrillation. Muscle biopsy is unusually performed by invasiveness, but allows the exclusion of other inflammatory myopathies. Histopathological examination of skin lesions results similar to lupus erythematosus, except for the absence of lupus band, assisting in the exclusion of other differential diagnosis.[1]

Up to 20-50% of cases of DM in adults are paraneoplastic manifestation, which usually presents a more abrupt onset, with early and exuberant clinical manifestations. Muscle alterations may be late, but rapidly progressive. Neoplasms more commonly related to DM are: ovary, breast, lung, colorectal and non-Hodgkin lymphoma; less frequently: pancreas, stomach and gallbladder. Amyopathic forms of DM have also been described as paraneoplastic manifestations.[4-6]

The diagnosis of neoplasia may precede (40%), be concurrent (26%) or follow (34%) the clinical manifestations of DM.[2,7,8]

Paraneoplastic syndromes are manifestations of tumors with greater local biological activity, invasiveness and metastasis.[9] Cases of paraneoplastic DM course with high mortality and morbidity, are less responsive to immune suppression and are likely to improve only after the cancer treatment.[8,10]

Our patient started topical and systemic corticosteroids (prednisone 0.5 mg/kg/d), hydroxychloroquine 400 mg/d, photoprotection and preoperative chemotherapy, with clinical control. Mastectomy and lymphadenectomy should lead to complete remission of symptoms.

In adults with DM, careful investigation of malignancies is mandatory.❑

**Abstract:** Dermatomyositis is a rare inflammatory disease, autoimmune, with proximal myopathy associated with characteristic dermatological manifestations. In adults, 20-50% of the cases are paraneoplastic manifestation, being mandatory the workup for malignancy Herein we report a case of a woman with classic dermatological presentation of dermatomyositis and newly diagnosed breast cancer. In general, the clinical presentation of paraneoplastic dermatomyositis is more exuberant and manifestations may precede, concur or succeed the diagnosis of neoplasia. The prognosis of cases associated with breast cancer is worse than the idiopathic form. Treatment is based mainly on the resolution of the underlying disease, beyond immunosuppressants.
**Keywords**: Carcinoma, ductal, breast; Dermatomyositis; Paraneoplastic syndromes

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