

**Paper Title of Case Report: Fatal Skin Lesions**Clara F. Jorge<sup>1\*</sup>, Marlene Louro<sup>2</sup>, Sónia Gomes Coelho<sup>3</sup>, Artur Costa<sup>4</sup>, Margarida Ascensão<sup>5</sup>, Leopoldina Vicente<sup>6</sup><sup>1,2,4-6</sup>Internal Medicine Department Centro Hospitalar Universitário Cova da Beira, Covilhã, Portugal<sup>2</sup>Internal Medicine Department Centro Hospitalar Universitário Cova da Beira, Covilhã, Portugal<sup>3</sup>Internal Medicine Department Unidade Local de Saúde da Guarda, Guarda, PortugalDOI: [10.36347/sjams.2022.v10i01.021](https://doi.org/10.36347/sjams.2022.v10i01.021)

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**Abstract****Case Report**

We present a clinical case of a 75-year-old woman hospitalized for localized edema and erythema on the right hemiface, with progressive extension over 30 days. On objective examination, a heliotropic erythema, Gottron's papules and an ulcerated lesion on the right breast were founded. It's important to point out an increase of muscle enzymes in laboratory tests. According to classification criteria of the European League against Rheumatism, and taking into consideration the clinical suspicion of breast cancer, a paraneoplastic dermatomyositis was diagnosed. Highlight that dermatomyositis lesions are pathognomonic despite that they are not always recognized in the patient's first approach.

**Keywords:** Dermatomyositis, breast cancer, inflammatory myopathies, Gottron.

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**INTRODUCTION**

Idiopathic inflammatory myopathies (IIM) are essentially characterized by inflammation of the muscles and muscle weakness, as well by systemic manifestations. The most common subgroups of IIM in adults are dermatomyositis (DM), polymyositis (PM) and inclusion body myositis (IBM), and in children, juvenile DM (JDM) [1, 2].

The classification of Bohan and Peter are outdated and new clinical subgroups emerged and the classification criteria of the European League Against Rheumatism (EULAR) / American College of Rheumatology (ACR) - EULAR / ACR 2017 were developed. The objective of this classification was to elaborate criterias that would allow the diagnosis of IIM using a probability-score, as well as categorizing them in main subgroups (*Table 1*) [1, 2].

**Table-1: Components of the 2017 EULAR/ACR classification criteria for adult and juvenile idiopathic inflammatory myopathies [1, 2]**

When no better explanation for the symptoms and signs exists these classification criteria can be used		
Variable	Score	
	No muscle biopsy	No muscle biopsy
<b>Age of onset</b>		
Age of onset of first symptom assumed to be related to the disease $\geq 18$ years and	1.3	1.5
Age of onset of first symptom assumed to be related to the disease $\geq 40$ years	2.1	2.2
<b>Muscle weakness</b>		
Objective symmetric weakness, usually progressive, of the proximal upper extremities	0.7	0.7
Objective symmetric weakness, usually progressive, of the proximal lower extremities	0.8	0.5
Neck flexors are relatively weaker than neck extensors	1.9	1.6
In the legs, proximal muscles are relatively weaker than distal muscles	0.9	1.2
<b>Skin manifestations</b>		
Heliotrope rash	3.1	3.2
Gottron's papules	2.1	2.7
Gottron's sign	3.3	3.7

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<b>Other clinical manifestations</b>		
Dysphagia or oesophageal dysmotility	0.7	0.6
<b>Laboratory measurements</b>		
Anti-Jo-1 (anti-histidyl-tRNA synthetase) autoantibody present	3.9	3.8
Elevated serum levels of creatine kinase (CK)* or lactate dehydrogenase (LD)* or aspartate aminotransferase (ASAT/AST/SGOT)* or alanine aminotransferase (ALAT/ALT/SGPT)*	1.3	1.4
<b>Muscle biopsy features—presence of:</b>		
Endomysial infiltration of mononuclear cells surrounding, but not invading, myofibres		1.7
Perimysial and/or perivascular infiltration of mononuclear cells		1.2
Perifascicular atrophy		1.9
Rimmed vacuoles		3.1

Modified from [1]

\*Serum levels above the upper limit of normal.

Clinically, patient exhibit proximal muscle weakness and typical skin lesions including: edema and periorbital heliotrope erythema (heliotropic rash), an erythematous scaly plaques located on the dorsal surface of the extensors of various joints (Gottron's papules), considered a pathognomonic sign in DM. Other less specific lesions may be observed: ill-defined erythematous macules involving the anterior aspect of the neck and the upper chest (V sign), erythema over the upper back (Shawl sign), cutaneous calcinosis, poikiloderma involving the lateral aspects of the thighs (Holster sign) and periungual hyperemia [3].

As DM is a systemic disease, it can affect the lung, in form of interstitial pneumonia in 10% of cases, as well as the heart inducing valvular disease, heart failure or myocarditis [4]. Oropharyngeal and upper esophageal muscle involvement may result to dysphagia or aspiration, and weakness of the diaphragm and chest wall muscles usually lead in respiratory failure.

The main laboratory findings are the increase of muscle enzymes (alanine aminotransferase, aspartate aminotransferase, creatine phosphokinase, lactate dehydrogenase and aldolase), which have a good relationship with disease activity [4]. An electromyoneurography (EMNG), a muscle biopsy and magnetic resonance imaging are complementary exams that also help in the diagnosis.

## CASE REPORT

We present a female patient with 75 years old, with mild congenital mental deficiency, independent for activities of daily living, was brought by her daughter to the Emergency Department (ED) due to worsening of photodistributed edema and erythema with 1 month of evolution.

The patient already came several times to the EM for the same reasons, and performed blood tests that didn't reveal any changes. Initially, she presented an edema and erythema on the right hemiface, progressing to all face in a week. She denied fever and airway involvement. It was initially diagnosed a facial

cellulitis and was empirically prescribed amoxicillin and clavulanic acid (875 mg + 125 mg every 12 hours) associated to a topical corticosteroid, demonstrating a slight improvement. In another emergency admission, she was treat with intramuscular injection of penicillin associated with deflazacort (30 mg per day). Almost a month later, she was hospitalized for deterioration of the clinical state with mixed dysphagia and hoarseness.

At this time, the patient denied any other type of cardiac-respiratory, genitourinary or gastro-intestinal symptoms. She denied relevant history of clinical disease and usual treatment, and knownd drug or food allergies.

On physical examination, stand out a generalized skin rash with heliotropic rash of the face and the "V sign" (*Image 1*). She also presented an edema with accentuated godet in the limbs with impetiginized lesions. A thickening of the cuticles and telangiectasias of the nail folds were seen, as well as Gottron papules in knees and elbows (*Image 2 and 3*) and Holster sign (*Image 4*).

Upon performing the gynecological examination, an ulcerated, adherent, infiltrative and painless lesion on palpation was found, in the upper external quadrant of the right breast, as well as axillary adenopathies on the same side (*Image 3*). In the rheumatologic examination, a notorious bilateral proximal muscle deficit was found in the lower limbs.

Analytically, was identified an increase of muscles enzymes: creatine kinase (6347 U/L), lactate dehydrogenase (1559 U/L), aspartate aminotransferase (424 UI/L), alanine aminotransferase (161 UI/L) aldolase (34.2 UI/L) and myoglobin (> 1200 ng/ml). The chest radiograph and the electrocardiogram didn't show any significant modifications.

The EMNG demonstrated signs of muscle fiber injury with signs of muscle fiber irritability.



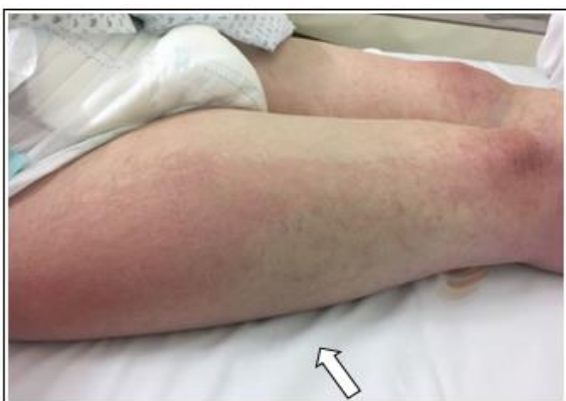
**Image-1: Triangular erythema in the neckline (V sign) with impetiginized vesicular-bullous lesions**



**Image-2: Gottron papules in the knees**



**Image-3: Gottron papules in the elbow (arrow 1) and ulcerated lesion in the upper right quadrant of the breast (arrow 2)**



**Image-4: Holster sign**

The patient underwent a biopsy of the breast lesion for anatomopathological study. Due to the highly suspicious nature of breast cancer, the patient was promptly staged with computed tomography scanner (CT scan) that revealed a nodular formation in the right breast, 28 mm in diameter, apparently invading the skin at the transition with the axillary region, and another nodular formation with 19.5 mm in diameter. Smaller adenopathies were also visible in the right axillary region. The bone scintigraphy was normal.

With the start of corticosteroid therapy with methylprednisolone (60 mg/day), the patient showed a slight improvement of the skin lesions, as well as a decrease of muscle and cardiac enzymology markers. However, the anatomopathological result showed an invasive carcinoma, and a palliative hormone therapy was decided based on multidisciplinary consultation meeting.

## DISCUSSION

Dermatomyositis is an idiopathic inflammatory myopathy, with an estimated prevalence rate of approximately 1 per 100000 in the general population [5]. The different forms of DM are more frequent in women than in men, in a ratio 2: 1 [6,7]. Typically it presents with progressive, symmetrical, proximal muscle weakness and characteristic skin lesions such as heliotrope rash, Gottron's papules, Gottron's sign, the V-sign and Shawl sign [8].

All patients newly diagnosed with DM should be evaluated for the possibility of an underlying malignancy. Cancer can be diagnosed before, simultaneously with, or after the diagnosis of inflammatory myopathy [9]. It is essential to carry out a neoplastic screening, especially in patients with newly diagnosed DM and aged over 50 years. The relationship between neoplasia and persistent inflammatory myopathy is not understood [10], however it is estimated that 15-30% of patients with DM have an underlying neoplastic disease [11]. This type of DM represents about 50% of IIM after 65 years old and less than 10% before 65. Clinical factors associated with an increased risk of malignancy include older age at disease onset, dysphagia, evidence of capillary damage on muscle biopsy, cutaneous necrosis, cutaneous leukocytoclastic vasculitis [12]. Adenocarcinomas of the cervix, lung, ovary, pancreas, bladder and stomach are responsible for the most frequent neoplasms associated with inflammatory myopathies [13, 14].

Autoimmunity plays an important role in the pathogenesis of inflammatory myopathies, being identified autoantibodies in more than 50% of patients. The following autoantibodies have been found to confer a positive risk on the development of neoplasia: anti-transcriptional intermediary factor-1 $\gamma$  (TIF-1 $\gamma$ ) and anti-nuclear matrix protein 2 (anti-NPX-2) [15-17].

According to the criteria of Troyanov and Targoff, the diagnosis of dermatomyositis appears to be probable or even definitive, as the ENMG report was not properly detailed [5, 18].

Conforming with the classification criteria of inflammatory myopathies proposed by EULAR/ACR 2017 (Table I), the calculated probability of IIM was 100%, with the diagnosis of DM (without associating the muscle biopsy result). This classification has a diagnostic sensitivity and specificity for IIM of 87 and 82%, respectively, when not associated with muscle biopsy results, and reaches 93 and 88% when correlated with biopsy [1].

Treatment of the underlying cancer is generally insufficient and DM needs to be treated. This treatment involves use of corticosteroids in first-line, however in this type of IIM, patients often do not respond to corticosteroid therapy, as happened in this patient. The evolution was fulminating and in 15 days, she died after starting the systemic treatment.

## CONCLUSION

Despite the presence of pathognomonic clinical signs of DM, these lesions were not recognized in various medical evaluations, which demonstrates that clinicians are unfamiliar with this disease. Some differential diagnoses were made, like facial cellulitis and drug toxicity, perhaps due to the benignity of the analytical control *ab initio*. The importance of an exhaustive objective examination in order to unravel the lesions of DM and breast neoplasm, would have allowed a quicker diagnosis, and thus avoided such a quick and fatal outcome.

## Conflicts of Interest

The authors have no conflicts of interest or financial relationships to disclose.

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