Amyopathic dermatomyositis in an elderly patient. An unusual case

Dermatomiositis amiopática en el adulto mayor. Un caso inusual

Estefanía Elizabeth Abundis Márquez,[1] Sara Gabriela Yeverino Castro,[4] Ana Cecilia Muñoz Díaz,[1] Daniel Zacarías Villarreal Martínez,[2] Francisco Javier Torres Quintanilla,[3] Griselda Serna Peña,[3] Jorge Ocampo Candiani,[2] Ricardo Salinas Martínez[1] y Rocío Morales Delgado[1]

1 Geriatrics Service, Hospital Universitario Dr. José Eleuterio González, Monterrey, Nuevo León
2 Dermatology Service, Hospital Universitario Dr. José Eleuterio González, Monterrey, Nuevo León
3 Rheumatology Service, Hospital Universitario Dr. José Eleuterio González, Monterrey, Nuevo León
4 Christus Center of Excellence and Innovation, San Pedro Garza García, Nuevo León

ABSTRACT

Dermatomyositis (DM) is an idiopathic inflammatory disease that causes symmetrical proximal muscle weakness and skin involvement primarily observed in individuals aged forty to fifty. Within the disease's subtypes is amyopathic DM, which is a rare and complex syndrome that represents approximately 10% of DM cases.

We present an unusual case of amyopathic dermatomyositis subtype in a 77-year-old woman with multiple comorbidities and a history of dermatosis nine years earlier with the diagnostic criteria for this disease.

KEYWORDS: *dermatomyositis, amyopathic dermatomyositis, heliotrope rash, autoimmune disease, older adults.*

RESUMEN

La dermatomiositis (DM) es una enfermedad inflamatoria idiopática que causa debilidad muscular proximal y simétrica e involucramiento de la piel. Se observa principalmente en personas entre 40 a 50 años de edad. Entre los subtipos de la enfermedad se encuentra la variedad amiopática, que es un síndrome raro y complejo y representa 10% de los casos de DM.

Presentamos un caso inusual de DM del subtipo amiopática en una mujer de 77 años con múltiples comorbilidades y antecedente de nueve años de diagnóstico de DM.

PALABRAS CLAVE: *dermatomiositis, dermatomiositis amiopática, erupción cutánea en heliotropo, enfermedad autoinmune, adultos mayores.*

Introduction

Dermatomyositis (DM) is a rare autoimmune disease with myopathy, skin involvement, and an associated higher risk of lung disease and malignancy.[1] The amyopathic DM subtype (ADM), may exhibit classic dermatological features without muscular involvement and is often seen in individuals aged between 40 and 50**.** In older adults, ADM may present with an atypical clinical presentation, leading to delayed diagnosis and treatment.[2]

Age-related changes, such as decreased functional reserve and response capacity, contribute to an atypical presentation of diseases. In older adults, numerous pathologies are non-specific, often linked to multiple diseases and alterations inherent in the aging process.[3]

Case

We report a case of a 77-year-old woman with a history of hypertension, type 2 diabetes mellitus, heart failure, and gastroesophageal reflux disease, with no record of autoimmune disease or allergies.

Originally, the patient sought medical attention, at Dr. José Eleuterio González University Hospital in Monterrey, Mexico, due to disseminated pruritic dermatosis presented nine years earlier. It started with plaques on the forehead, progressing to the eyelids, anterior thorax, and upper extremities, accompanied by fatigue.

Upon physical examination, heliotrope rash, a violet discoloration in the upper eyelids accompanied by swelling, was observed (**figure 1a**). Over the anterior neck and chest, flat macular erythema known as the "v sign" (**figure 1b**) and a similar rash located in the upper and posteri-

CORRESPONDENCIA Dra. Rocío Morales-Delgado ■ drarociomoralesdelgado@gmail.com ■ Telephone: 81 8362 3296
Geriatrics Service, Hospital Universitario Dr. José Eleuterio González, UANL, Monterrey, Nuevo León, México

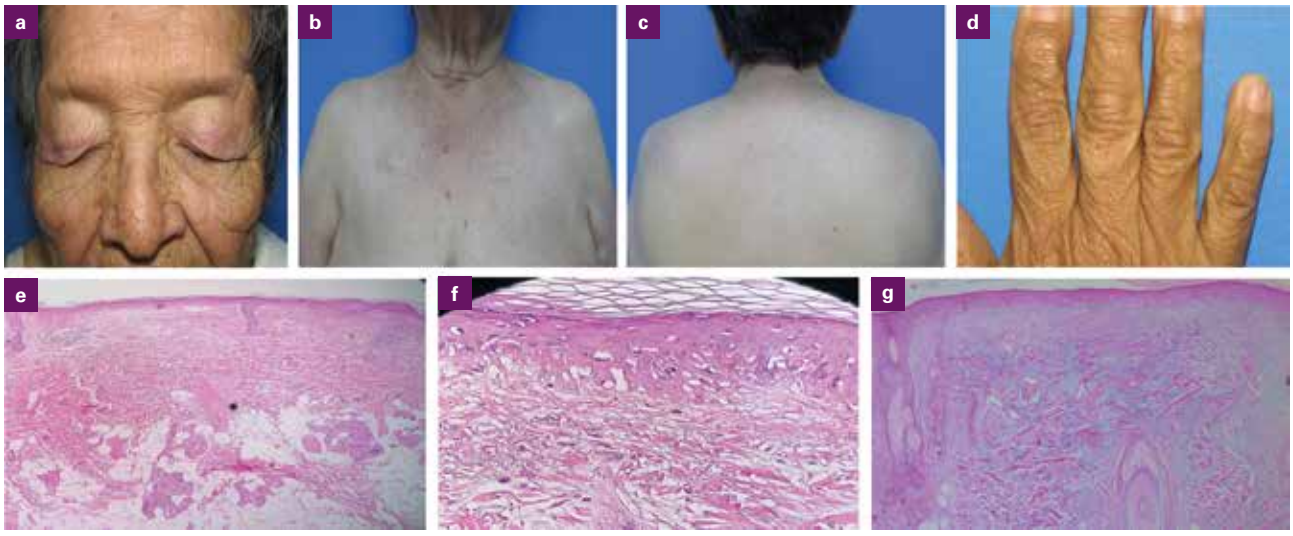

Figure 1. **a)** Heliotrope Rash. **b)** v sign. **c)** Shawl sign. **d)** Gottron's papules. **e)** Skin biopsy (panoramic view). **f)** Skin biopsy (interface dermatitis). **g)** Skin biopsy (colloidal iron histochemical staining).

or neck and shoulders, known as the "Shawl sign", both characteristic of DM, were observed (figure 1c). Gottron's papules were observed over the metacarpophalangeal joints; erythematous-violaceous papules, slightly raised, commonly scaly, that cover the bony prominences (figure 1d). Panoramic view with marked edema at the level of the papillary dermis (figure 1e). Interface dermatitis with vascular degeneration at the level of the dermo-epidermal junction and slight thickening of the basal lamina which was demonstrated with histochemical periodic acid-Schiff stain (PAS) (figure 1f). With colloidal iron staining a diffuse and moderate acidic mucin deposition at the dermis level, including eccrine glands and cutaneous appendages, was observed (figure 1g). To assess the strength and performance of the muscles, the *Manual Muscle Testing* 8 was performed with no clinical evidence of muscle involvement.

Laboratory tests were normal, including blood count and liver and kidney function. Muscle enzymes such as creatine kinase (CPK), lactate dehydrogenase (LDH), and aldolase were also normal, and antinuclear antibodies (ANA) were negative.

An autoimmune myopathy panel was conducted, yielding a positive result for transcription intermediary factor 1-gamma (TIF1-γ) and a borderline result for anti-MDA5. The colloidal iron histochemical staining of the biopsy was positive for interstitial mucin, highly suggestive of DM (figure 1f).

The comprehensive geriatric assessment revealed mild cognitive impairment, depression ranging from moderate to severe, and a notable degree of self-sufficiency; although dependence on instrumental activities of daily living and mild frailty were both observed (clinical frailty scale score of 5).

The final diagnosis was ADM; management included topical high-potency corticosteroids, systemic oral corticosteroids (prednisone), immunosuppressants (methotrexate), hydroxychloroquine, folic acid, and comorbidities and geriatric syndromes assessment.

Given the association between DM and certain cancers, the patient was evaluated for underlying cancer, and the results from the evaluation were negative.[2]

Discussion

ADM is a rare clinical condition, particularly in older adults. The average age of onset is between 40 and 50 years, with a higher prevalence in women. In our case, an older woman presented with a classic ADM, which is unusual for this age group. ADM presents with cutaneous involvement but no myopathy; a > 6-month typical dermatosis was found in this patient, and laboratory studies excluded muscle involvement.[4] A recent case series report by Rodríguez-Tejero and collaborators in Spain, described three other cases of clinical ADM, in two males and one female, aged 63, 76 and 70, respectively.[5] The first and the last of mentioned cases also presented neckline v rash and shawl sign. Only the eldest of the patients in the case series had positive anti-extractable nuclear antigen antibodies. In addition, the youngest had a bronchogenic cancer diagnosis while the other (aged 70) had a locoregional endometrial cancer recurrence, which coincided with the fact that both presented constitutional

syndrome. By 2004, only one biopsy confirmed drug-induced case of clinical ADM in a 53-year-old-female was reported in Mexico.[6,7] The patient presented with diffuse pruritic violaceous erythematous lesions on the neck and Gottron papules on her knuckles, with no malignancy development.[7] However, unlike our patient, despite treatment (steroids, antimalarials, and methotrexate) the patient persisted with dermatologic lesions.

After nine years, the patient received a final diagnosis of ADM, for which she started treatment with steroids, antimalarials, and methotrexate, with a successful response. However, in a retrospective study by Tang and collaborators that included 38 ADM patients in China, a combined therapy was the most prevalent treatment, while in the whole cohort the use of a single antimalarial had a 68.8% response rate.[8] In this same study, when compared to the typical ADM group, patients with hypomyopathic dermatomyositis received combined treatments with steroids and immunosuppressants more frequently. Moreover, in a systematic review where only observational studies were included, 60% of patients had received more than one treatment due to side-effects or lack of efficacy. In this same study, antimalarials were the most associated with lack of improvement.[9] More research is needed to reach a consensus on ADM classification and treatment efficacy.[4]

We performed a differential diagnosis regarding the skin condition. Seborrheic dermatitis, rosacea, and contact dermatitis were excluded. We must consider that our patient had a 9-year history of clinical symptoms and that her diagnosis was made in the seventh decade of life. A study by Da Silva and collaborators reported a DM diagnostic delay from presentation to final diagnosis that ranged from 2.2 weeks to 267 months, and a median for ADM of 17.1 months compared to 12.2 in classic DM.[10] Authors in the latter study argued that patients without muscle involvement are even at a greater risk of a misdiagnosis. More than half of DM confirmed cases had previously received a different diagnosis (e.g. cutaneous or systemic lupus erythematosus, undifferentiated connective tissue disease, dermatitis, rosacea, eczema, psoriasis, among others.[10] In older adults, ADM diagnosis represents a significant challenge, primarily linked to the presence of comorbidities and the overlap of ADM symptoms with other clinical conditions, especially frail patients with fatigue, anorexia and weight loss.[11,12]

ADM is associated with lung disease and malignancy; however, a detailed evaluation was performed, and it was further ruled out.[13] Still, factors such as diagnostic delay are relevant since, although malignancy risk is highest in the first two years after symptom onset, a six-time greater risk of malignancy associated to DM has been reported.[14] Hence, these patients must be monitored closely, especially those with history of smoking, the shawl sign, skin necrosis, constitutional syndrome, or those in which high risk antibodies have been detected (e.g. TIF1 and anti-nuclear matrix protein 2 [NXP2]). Up to 5-10 years after DM onset, an annual age-appropriate malignancy screening is recommended.[5] Considering that our patient had borderline anti-MDA5 antibodies and that its presence is associated with the possible onset of rapidly progressive interstitial lung disease, a pulmonary screening should also be considered.[4]

Conclusion

ADM is a rare disease, especially in the older adult. Individual case reports that can contribute to the disease's knowledge and clinical experience in this age group is fundamental. Since diagnostic delays and misdiagnosis are prevalent, describing age-related variations influencing the clinical manifestation of ADM is crucial, as it may facilitate earlier diagnosis and reduce treatment delay in older adults.

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