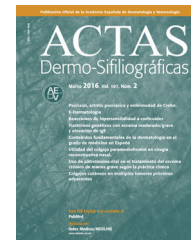




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CASE AND RESEARCH LETTERS

Edematous Dermatomyositis with Probable Evans Syndrome[☆]



Dermatomiositis edematosa asociada a probable síndrome de Evans

To the Editor:

Inflammatory diseases of the muscles and skin are rare and orphan conditions. Dermatomyositis is an idiopathic inflammatory disorder associated with typical muscle and/or skin manifestations. Presentation with edema, ie, edematous dermatomyositis, is an infrequent variant. We report a new case of edematous dermatomyositis and review the literature.

A 52-year-old woman with no clinical history of interest consulted with a pruriginous rash that first appeared 1 month previously. The rash took the form of violaceous erythema on the center of the face and around the eyes, erythematous macules on the dorsum of the metacarpophalangeal joints, and flagellate erythema on the back. The patient also experienced disabling muscle weakness that mainly affected proximal areas and intense dysphagia and dysphonia. The only findings of note in the laboratory workup were increased muscle and liver enzyme values, as follows: creatine phosphokinase, 4005 IU/L (> 140); aldolase, 44.4 IU/L (> 7.5); aspartate aminotransferase, 336 IU/L (> 31); alanine aminotransferase, 187 IU/L (> 40); γ -glutamyl transferase, 109 IU/L (> 30); and lactate dehydrogenase, 1022 IU/L (> 385). The results of antibody testing (antinuclear antibodies, anti-RNP, anti-Jo1, and anti-p155) were negative. Analysis of a skin biopsy specimen revealed vacuolar changes at the dermal-epidermal junction, solitary necrotic keratinocytes, and mucin deposits in the dermis. The electromyogram revealed signs of inflammatory myopathy, and occult underlying neoplasm was ruled out by tumor markers and positron emission tomography and computed tomography imaging. These findings confirmed the diagnosis of dermatomyositis, and treatment was started with intravenous methylprednisolone (1 mg/kg/d); after 5 days, 4 doses of immunoglobulin (1.5 g/kg/dose) were added, although there was little improvement. During the following months, the patient's condition progressed



Figure 1 Erythematous-violaceous rash affecting the face and neck. The rash is symmetrical and particularly noticeable on the eyelids.

with intense edema affecting the face, neck, and upper extremities (visible on the magnetic resonance image) and myositis (Figs. 1–3). Therefore, methotrexate was added to the treatment regimen at 2 months, hydroxychloroquine at 4 months, and, given the lack of improvement, rituximab (1 g) at 6 months in 2 doses separated by 2 weeks. The response was good, mainly in the skin. During follow-up, occasional thrombocytopenia and anemia (hemoglobin, 10.8 g/dL; platelets, 58 000/ μ L) were recorded, as were increased values for indirect bilirubin (1 mg/dL [> 0.7]) and lactate dehydrogenase (641 IU/L [> 385]). These findings were compatible with Evans syndrome with dermatomyositis occurring alongside worsening muscle enzyme values. However, as the patient was receiving treatment with corticosteroids, it was impossible to perform the Coombs test to confirm this associated autoimmune etiology. Similarly, it was not possible to rule out other etiologies.

Dermatomyositis is an autoimmune disease that mainly affects the skin and muscle. It has traditionally taken the form of mild periorbital edema accompanied by heliotrope rash. However, edematous dermatomyositis involves more extensive swelling. It is also a rare clinical variant of the condition, with only 23 cases reported in the literature.¹ The etiology and pathogenesis remain unclear, although

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Figure 2 Erythematous macular lesions on the metacarpophalangeal joints and the dorsum of the fingers, with edema (the arrow indicates the pressure exerted by the patient's ring).

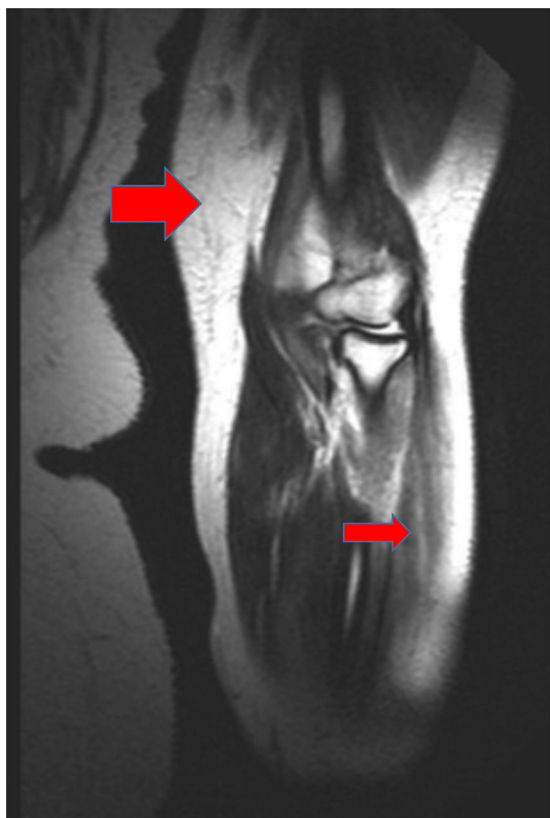


Figure 3 Magnetic resonance image of the left arm, where a hyperintense signal in several muscle groups is compatible with myositis. It is also possible to observe edema affecting the muscle and subcutaneous cellular tissue (the large red arrow points to an affected muscle group, and the small red arrow points to an area of edema in the subcutaneous cellular tissue).

some authors have pointed to intense inflammatory activity with activation and deposition of complement, leading to vascular disease with muscle microinfarcts that in turn increase vascular permeability.¹⁻⁵ This manifests clinically as edema affecting muscle and/or subcutaneous tissue, with or without pitting. Age at presentation is

variable,^{1,2,6} with most adult cases occurring in women.¹ Edema mainly affects the upper extremities, although it may be generalized^{4,6,7}; cases of local edema have also been reported.^{2,8,9} Edematous dermatomyositis usually progresses more rapidly than classic dermatomyositis.⁵ Edema usually develops after skin involvement, although it may also be the initial presentation⁶; there have even been reports of edematous dermatomyositis with no other cutaneous findings.⁵ Muscle involvement and dysphagia are frequently associated with edematous dermatomyositis,^{1,4} as observed in the case we report.

It remains unclear whether this clinical presentation of dermatomyositis implies a greater risk of neoplasm⁵; according to a recently published literature review, the risk of neoplasm was recorded in fewer than 30% of cases (6 of 23 cases reviewed).¹

Before a diagnosis of edematous dermatomyositis can be confirmed, it is important to rule out other, secondary causes of edema,¹ such as kidney, heart, and thyroid disease, as well as hypoproteinemia. Treatment of edematous dermatomyositis should be intensive and early, given the potentially severe nature of the symptoms^{1,4,5,9,10} and the probable poorer prognosis than with classic dermatomyositis.^{4,6,10} The combination of high doses of intravenous corticosteroids and immunosuppressants seems to be a good alternative; immunoglobulins can be added when there is no response,^{3,5,8} and rituximab can be administered in refractory cases.^{1,2}

Evans syndrome (anemia and/or autoimmune thrombocytopenia) rarely occurs with dermatomyositis. The first case was reported in 1990 in a woman with generalized edematous dermatomyositis.³ Although we were unable to confirm an autoimmune etiology in the case we report, the clinical and laboratory data were consistent with this diagnosis.

We report a new case of severe edematous dermatomyositis that proved refractory to several systemic treatments but responded well to rituximab. The condition probably occurred with Evans syndrome.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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Reticulate Acropigmentation of Kitamura and Nevus of Ito[☆]



Acropigmentación reticulada de Kitamura y nevus de Ito

To the Editor:

Reticulate acropigmentation of Kitamura (RAPK) is an uncommon genodermatosis that is classed as a congenital reticulate pigmentary disorder. It is characterized by acral lentiginous pigmentation and mainly affects Japanese individuals. Nevus of Ito is a congenital dermal melanocytosis that is congenital and affects the deltoid and acromioclavicular region, more commonly in Asian women.

We report the case of a 47-year-old woman (phototype III) with no clinical history of interest who consulted for progressively slow-growing asymptomatic darkening of the skin on the dorsum of the hands and on the face that began during adolescence. She reported that her mother had similar pigmentation on the hands, although other family members did not (2 maternal aunts, 2 siblings, 3 children). She also had a stable congenital lesion on her back and right hemithorax. Examination revealed symmetrical lentiginous pigmentation on the dorsum of the hands, the anterior aspect of the wrists, the eyelids, and the area around the lips, together with palmoplantar pits and interrupted palmar creases (Fig. 1). A blue-grayish plaque was observed on her back and right hemithorax and arm (Fig. 2). Analysis of a biopsy specimen from the hand revealed lentiginous epidermal hyperplasia with hyperpigmented bulbous projections of the epidermal rete ridges; in the case of the dorsum, it revealed dendritic melanocytes in the dermis (Fig. 3). These findings led us to diagnose the patient's condition as RAPK and nevus of Ito.

Reticulate pigmentary disorders comprise a group of congenital and acquired conditions with reticulate or freckle-type hyperpigmentation and occasional

hypopigmentation. There is some confusion in the literature with respect to definitions and terminology. Hereditary forms are unusual and have variable inheritance patterns, although they are generally autosomal dominant, with possible associated abnormalities. They include dyskeratosis congenita, Dowling-Degos disease, RAPK, Haber syndrome, reticulate acropigmentation of Dohi, Naegeli-Franceschetti-Jadassohn syndrome, X-linked reticulate pigmentary disorder, and dyschromatosis universalis hereditaria. In the acquired forms, the lesions are usually larger and include confluent and reticulate papillomatosis (Gougerot-Carteaud), prurigo pigmentosa, lichen planus pigmentosus, Riehl melanosis, erythema ab igne, cutis marmorata, livedo reticularis, and postinflammatory pigmentation.¹

RAPK is an uncommon disorder, with approximately 130 reported cases.^{2–7} While more common in Japan, it can be found throughout the world. The inheritance pattern is autosomal dominant, although nonhereditary cases occur sporadically. It first appears during the first or second decade of life, with reticulate or freckle-like hyperpigmented macules that measure 1 to 5 mm. The lesions are initially atrophic and are found on the dorsum of the hands and feet and can extend proximally with progressive darkening aggravated by exposure to sunlight. They rarely affect the face and are accompanied by palmoplantar pits and interrupted palmar creases, and there have been isolated reports of localized alopecia and absence of the distal phalanges of the toes. Dermoscopy shows the pigmentation to have a fine network of nonspecific reticulate brown pigment and the palmoplantar pits to have brown spots.² Histopathology reveals atrophy of the epidermis, elongation, and increased melanin on the interpapillary ridges, with scarce perivascular lymphocytic infiltrate, and an increased number of DOPA-positive basal melanocytes.⁴

There is some debate over whether RAPK, Dowling-Degos disease, Haber syndrome, and reticulate acropigmentation of Dohi are variants of the same disease, since their clinical and histologic characteristics overlap to some extent, mainly in Dowling-Degos disease and RAPK. The exception is Galli-Galli disease, in which suprabasal acantholysis can be observed.^{3,4} Dowling-Degos disease is autosomal dominant, more commonly affects adult women, and manifests with reticulate pigmentation and brown hyperkeratotic papules mainly on flexures and the trunk. It may be accompanied by

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