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T. Díaz-Corpas,* A. Mateu-Puchades, R. Rojo-España, and A. Marquina-Vila

Servicio de Dermatología, Hospital Universitario Dr. Peset, Valencia, Spain

*Corresponding author.
E-mail address: ainatdc@hotmail.com
(T. Díaz-Corpas).

Erythema Elevatum Diutinum or Extrafacial Granuloma Faciale?

¿Eritema elevatum diutinum o granuloma facial extrafacial?

To the Editor:

Erythema elevatum diutinum (EED) and granuloma faciale (GF) are rare diseases. EED presents with multiple plaques or nodules typically located on extensor areas,[1,2] whereas GF is usually a single lesion in the facial region.[3] However, differential diagnosis is sometimes difficult due to clinical and pathological similarities.[4]

We report the case of a 57-year-old man who came to our department because of the appearance at least 15 years earlier of an asymptomatic brownish plaque of soft consistency on the anterior aspect of the left thigh; the plaque had grown slowly but progressively, becoming harder and darker (Figure 1). A further 3 brown, plaque-like lesions with an atrophic appearance had developed years later on the back of the right leg, on the left thigh, and over the left iliac crest; the patient said they were similar to the early appearance of the initial lesion. There were no systemic symptoms. The biopsy of the nodular lesion (Figures 2 and 3) showed fibrosis and perivascular interstitial infiltrates formed of lymphocytes, plasma cells, and neutrophils with karyorrhexis, and vascular proliferation with intraluminal fibrin thrombi. Laboratory tests revealed an immunoglobulin (Ig) A concentration of 384 mg/dL (normal range, 100-300 mg/dL) and IgM of 78.9 mg/dL (normal range, 80-250 mg/dL), as well as a ferritin of 402 ng/mL (normal range, 30-400 ng/mL), the high level of which led us to perform a study to exclude hemochromatosis. Protein electrophoresis, other laboratory values, electrocardiogram, and chest radiograph were normal. Serology for hepatotropic viruses and human immunodeficiency virus was negative. In agreement with the patient, no treatment was applied.

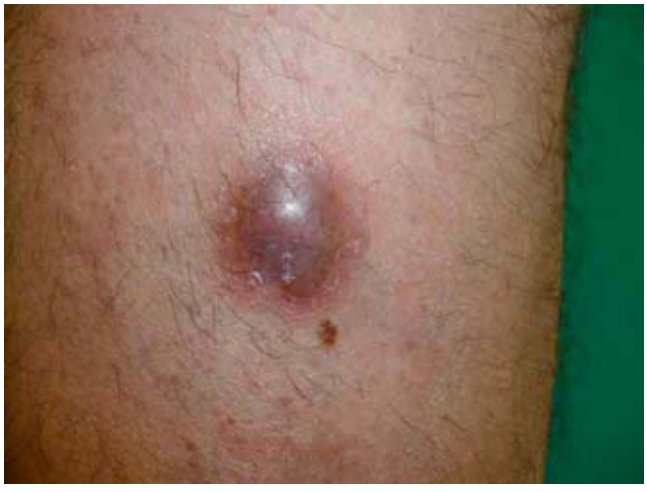

Figure 1 Well-defined brown nodule of 2 cm diameter on the anterior aspect of the left thigh, adherent to the superficial tissues.

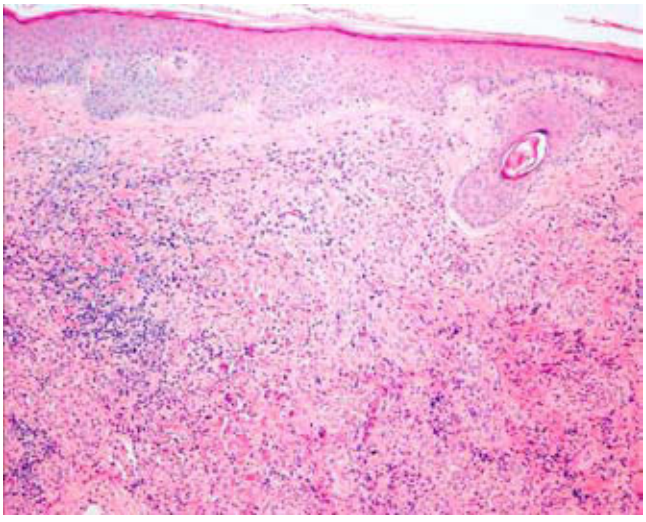

Figure 2 Mixed infiltrate with abundant neutrophils in the dermis, sparing the most superficial part of the papillary dermis (Grenz zone) (hematoxylin-eosin, original magnification ×100).

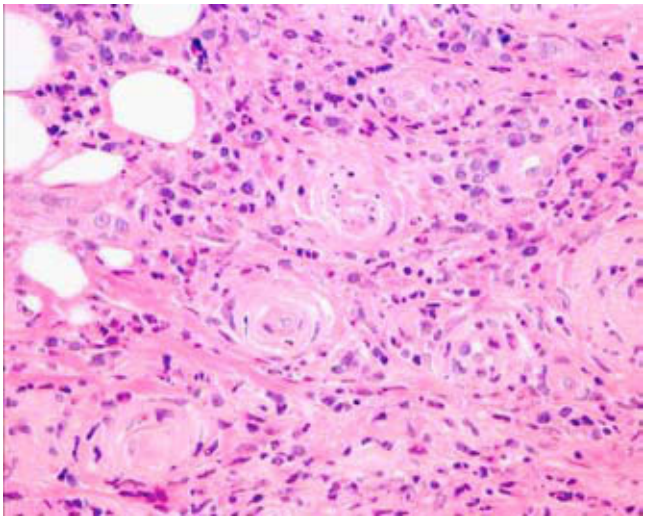

Figure 3 Concentric lamellar perivascular fibrosis in the dermis (hematoxylin-eosin, original magnification ×400).

The chronic nature and the histopathology of the lesions suggest that this is what Carlson and LeBoit[4] described as chronic fibrosing vasculitis, a pattern that can be found in diseases such as EED or GF.

The typical presentation of EED is with multiple brownish papules or nodules symmetrically distributed on extensor surfaces of the limbs. Very few cases with atypical morphologies and locations have been reported in the literature.[5]

EED has been associated with various systemic diseases, especially hematologic ones, of which the most common is IgA paraproteinemia.[1,2]

This gammopathy is not found as such in the majority of patients, who instead show elevated levels of IgA,[1] as occurred in our case. By contrast, GF is characterized by a solitary nodule in the facial region. However, it is sometimes accompanied by extrafacial lesions, which may exceptionally be the only clinical manifestation,[6,7] in which case the distribution is usually asymmetric.

Histologically, the early lesions of EED are characterized by leukocytoclastic vasculitis and a neutrophil-rich infiltrate, which declines in the longer-standing lesions to be replaced by dermal fibrosis and capillary proliferation.[4]

In both facial and extrafacial GF the infiltrate contains abundant eosinophils and spares the upper part of the dermis. Vasculitis—and in more advanced lesions fibrosis—can also be found.[3,4]

In conclusion, both disorders are characterized by leukocytoclastic vasculitis in the early stages and by perivascular fibrosis—with or without the persistence of vasculitis—in the later stages. Our patient demonstrates the difficulty of reaching a diagnosis in cases of chronic fibrosing leukocytoclastic vasculitis that do not correspond clinically to the most typical forms of these 2 dermatoses. On the basis of the hypothesis proposed by Ortonne et al,[7] we believe that EED and GF with extrafacial involvement, in addition to other diseases such as localized chronic fibrosing vasculitis, belong to the same spectrum of diseases. However, despite a possible histologic overlap, the classical clinical form of GF could be considered as a defined disease that is independent of EED.

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R. Navarro,[a,*] D. de Argila,[a] J. Fraga,[b] and A. García-Diez[a]

[a]*Departamento de Dermatología, Hospital Universitario La Princesa, Madrid, Spain*
[b]*Anatomía Patológica, Hospital Universitario La Princesa, Madrid, Spain*

*Corresponding author.
E-mail address: nat_eraa@hotmail.com (R. Navarro).