

Eosinophilic Fasciitis and Common Variable Immunodeficiency: An Unusual Association and Literature Review



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Eosinophilic fasciitis (EF), first described in 1974 by Lawrence E. Shulman, is a rare condition of unknown etiology characterized by fascial inflammation with eosinophilic infiltrates. It typically presents as a scleroderma-like syndrome, with limbs and trunk fasciitis with a mild hypereosinophilia.¹

A 61-year-old man with suspected atypical myositis was referred to our institution. He suffered from essential hypertension. He did not report recurrent infections. The patient reported gradually worsening myalgias in the legs and upper limbs for 4 years.

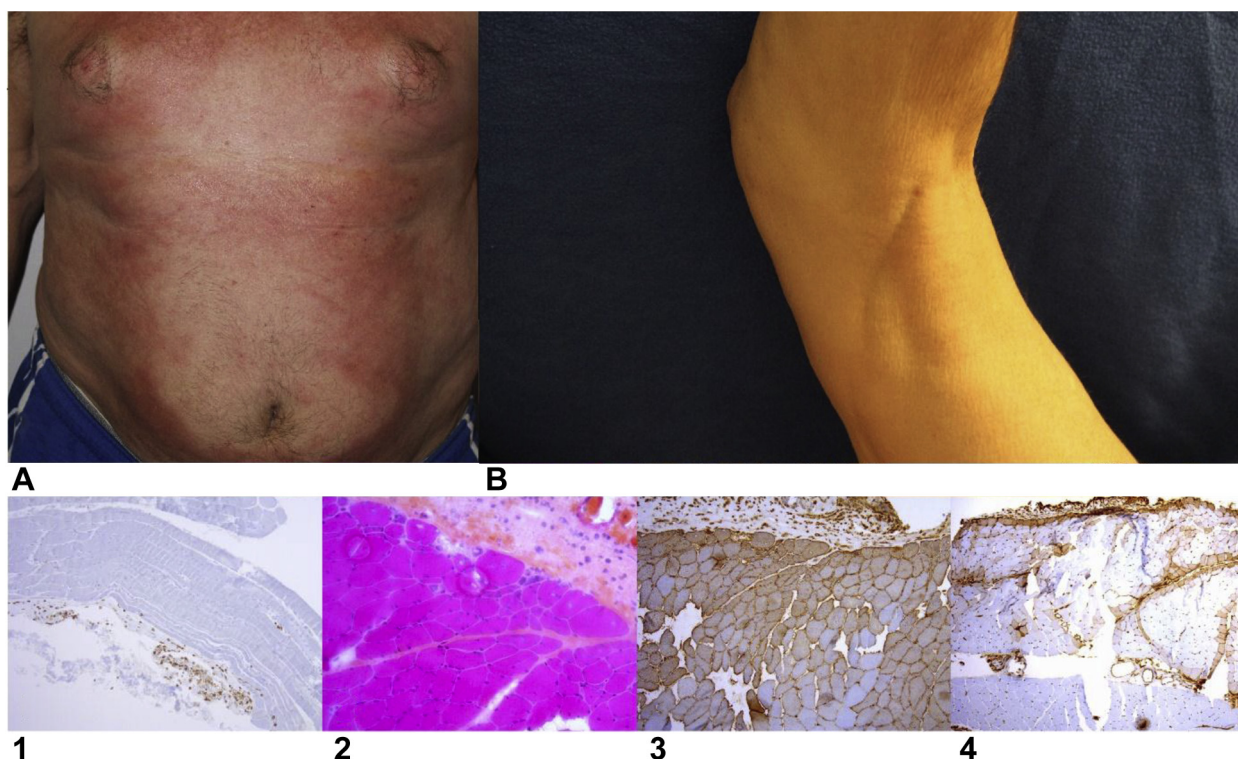


FIGURE 1. (A) Trunk morphea. (B) "Groove sign." Groove sign is a skin depression that is seen along a vein path. Fascial inflammation and perifascicular atrophy are seen on histopathology (1 [CD45 staining] + 2), and HLA-DR and HLA-I are expressed by all perifascicular fibers (3 + 4) on muscle biopsy.

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Conflicts of interest: M. Gerfaud-Valentin is associated with Sobi and Novartis. P.

Petiot is associated with LFB, Teva, and Novartis. P. Sève is on the advisory boards for AbbVie and Novartis and lectures for LFB, Roche, Sobi, AbbVie, and

Pfizer. The rest of the authors declare that they have no relevant conflicts of interest.

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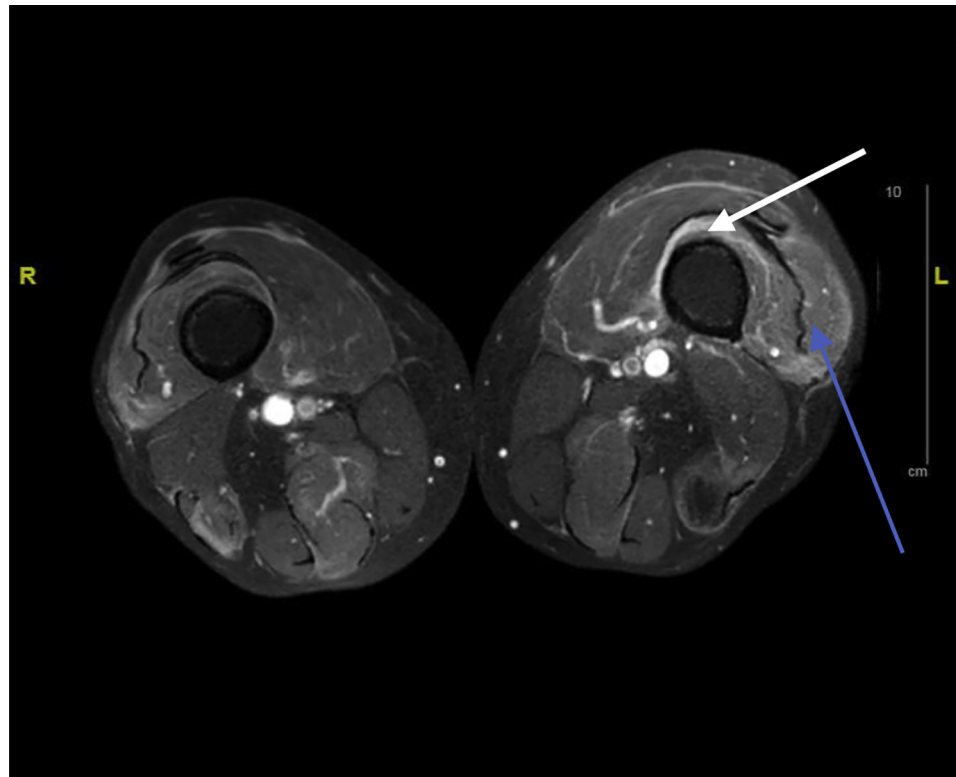


FIGURE 2. T2 short tau inversion recovery magnetic resonance imaging: myositis (blue arrow) and fascial thickening (white arrow) with postgadolinium enhancement.

On admission, the clinical examination revealed an extensive trunk morphea (Figure 1, A) and a “groove sign” (Figure 1, B) on the arms. Proximal muscle strength was rated 4/5 and there was no distal defect.

Laboratory tests showed mild eosinophilia ($900/\text{mm}^3$) and a normal creatinine phosphokinase level. The C-reactive protein level was 20 mg/L. The protein electrophoresis showed marked hypogammaglobulinemia (total immunoglobulins = 1.5 g/L; IgA = 0 g/L; IgM = 0 g/L; and IgG = 1 g/L). There was neither proteinuria nor digestive protein loss. The lymphocyte phenotyping was consistent with the diagnosis of common variable immunodeficiency (CVID) (low switched memory B cells: $7/\mu\text{L}$ [normal, 20-90/ μL], 2.4% of the total B-cell count; paradoxically increased transitional B cells: $27/\mu\text{L}$, 9.24% of the total B-cell count). Neither vaccinal responses nor plasma IL-5 levels were studied. The bone marrow biopsy did not reveal any lymphomatous infiltrate, and the whole body computed tomography and fluorine-18 fluoro-2-deoxy-D-glucose positron emission tomography-computed tomography did not show any lymphadenopathy or thymoma.

The electrophysiologic examination of the inferior limbs showed muscle and neurogenic patterns during contraction in proximal muscles with spontaneous activity. The muscle magnetic resonance imaging showed T1 gadolinium and T2 hyper-signals in the right quadriceps femoris, consistent with muscle fatty degeneration with fascial gadolinium uptake (Figure 2). A skin biopsy was performed. On pathological examination, a morphea pattern was described. The histopathologic examination of a quadriceps femoris muscle sample revealed a fasciitis with perivascular lymphocytic infiltrates along with nonspecific

myositis (Figure 1, 1-4) without eosinophils. These results led to the diagnosis of EF associated with CVID. The patient was started on high-dose intravenous immunoglobulin along with corticosteroids (pulsed intravenous methylprednisolone, then 1 mg/kg/d prednisone). The patient did not show any improvement and was started on methotrexate (0.3 mg/kg/wk). Then, intravenous immunoglobulin as a substitutive therapy and adalimumab (40 mg/2 wk) were prescribed. After 7 months, the clinical examination showed a slight thinning of the skin in affected areas, the electrophysiologic examination showed an improvement in neurogenic and myogenic patterns, and the magnetic resonance imaging showed a decreasing inflammatory signal. The disease then went into remission without any severe infection occurring.

Our case report is original because hypogammaglobulinemia is rarely associated with EF. Three similar cases and also “morphea-hypogammaglobulinemia” associations have already been reported.

In CVID, experts usually recommend maintaining immunoglobulin substitution while patients are treated with immunosuppressants, even in the absence of recurrent infections² such as in the present case. Given those elements, we consider that hypogammaglobulinemia should be ruled out in EF mostly to avoid severe infections that may complicate immunosuppressive therapy.

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