Necrotizing fasciitis secondary to the immunosuppressive treatment of eosinophilic fasciitis: radiological imaging, treatment modalities and outcome

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Title: Necrotizing fasciitis secondary to the immunosuppressive treatment of eosinophilic fasciitis: radiological imaging, treatment modalities and outcome

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Short title: Necrotizing fasciitis caused by eosinophilic fasciitis treatment.

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A 60-year-old Caucasian male in overall good medical state presented with a history of ankle edema and skin reddening, followed by severe, constant myalgia affecting lower extremities symmetrically. The symptoms escalated further, leading to a skin rash most prominent in the ankle area. On examination, range of motion in lower and upper limbs was decreased due to the developing hardening of the skin and fascia. Blood tests revealed mild hypoalbuminemia of 30.7 g/L and elevated C-reactive protein (CRP) level of 429 nmol/L. Differential blood count showed absolute and relative eosinophilia, with total leukocytes at $7.34 \times 10^9/L$ and 27% of eosinophils. Prior to diagnosis, the patient was reviewed by various consultants, including an infectious diseases specialist and a rheumatologist. Lyme disease was suspected and *Borrelia burgdorferi* testing was performed. Elispot tests indicated weak cellular activity against *B. burgdorferi*. The patient was treated for *Borrelia* infection using the Antibiotics Augmented Thermoeradication approach. Antinuclear antibodies (ANAs) titer was marginally increased (1:80), while Extractable Nuclear Antigen Antibodies (anti-ENA) and Antineutrophil Cytoplasmic Antibodies (ANCA) remained within normal limits. Myofascial biopsy revealed an inflammatory myopathy with mostly lymphocytic infiltrates in the perimysium, endomysium and fascia. Apart from lymphocytes and macrophages, occasional eosinophils were found. Based on these findings, 4 months after symptom onset, the diagnosis of eosinophilic fasciitis (EF) was made. Methylprednisolone 48mg daily (with dose reduction of 4mg weekly until 16mg) and methotrexate 15mg weekly were administered. 4 months post-treatment, the patient was admitted to the Department of Surgical Infections in Ljubljana (Slovenia) with necrotizing fasciitis (NF) type II of the left lower limb. On admission, patient was febrile, hypotensive, with altered mental status, CRP of 2410 nmol/L, computed tomography (CT) hallmarks (A, B) and a septic shock. Patient received antibiotic therapy – imipenem and clindamycin. Methotrexate was discontinued, and methylprednisolone substituted with hydrocortisone. Fasciectomy (C) and debridement were
performed. After a few days the patient stabilized. Post-fasciectomy wound therapy using negative pressure (D) and hyperbaric oxygen chamber (E, F) was applied.

EF is a rare disease, with less than 300 cases reported between 1977 and 2012. *B. burgdorferi* is the main postulated EF trigger, which agrees with our observations [1]. EF may mimic NF in magnetic resonance imaging and CT imaging, which adds to the complexity of its radiological interpretation. In both EF and NF, dermal thickening, increased soft tissue opacity and thickening or edema of myofascial structures can be observed. In this case, CT changes were accompanied by gas collection, which is characteristic of NF [2]. NF is a medical emergency. Out of the established NF risk factors such as diabetes mellitus and peripheral vascular diseases, only immunosuppression and advanced age were present in this case [3]. Since this is the first reported case of EF followed by NF, it is impossible to assess if the eosinophilic inflammation of the fascia was an additional local risk factor for necrosis development. Corticosteroid therapy can provoke NF relapse; therefore, alternative treatment should be considered. Treatment options include infliximab, which proved to be effective in several EF patients [4], as well as psoralen and ultraviolet A therapy [5]. Since the treatment of EF may cause NF, it should be carefully monitored.
References


Figure 1. Patient’s left lower limb presentation in different moments of necrotizing fasciitis.

A, B Computed tomography images of lower limbs on emergency admission, with two different coronal sections with visible fluid and gas collection.

C The left lower limb after the fasciectomy.

D Left lower limb during negative-pressure wound therapy.

E Left lower limb after 2 weeks of necrotizing fasciitis treatment with negative-pressure wound therapy. New granulation tissue is visible.

F Left lower limb after 12 weeks of necrotizing fasciitis treatment with negative-pressure wound therapy and hyperbaric oxygen chamber.