EOSINOPHILIC FASCIITIS: The Rare and Unusual Cause of His Edema

Karina Monica A. Borromeo MD\(^1\), Lia Jamian MD\(^2\)

\(^1\)University of Connecticut Health Center Primary Care Internal Medicine, \(^2\)Hartford Healthcare Medical Group Rheumatology

**INTRODUCTION**

- **Eosinophilic fascitis** is a rare fibrosing disease of the subcutaneous fascia with unknown etiology.
- In its early phase it can present as an acute symmetric edema of the extremities later progressing to induration of the skin.

**CASE REPORT**

- 46-year-old male presented with myalgias and swelling of his lower extremities which progressed over 2 months to involve his upper extremities.
- Endorsed heavy manual labor several weeks prior to presentation.
- Pertinent medical history: ulcerative colitis - not on therapy, quiescent
- Social history: former smoker 1ppd ~ quit 25 years ago, drinks 4 beers a week, occasionally smokes marijuana

**IMAGING**

- MRI of lower extremities with contrast revealing subcutaneous and fascial edema (yellow arrows) on T2-weighted image.
- Radiology: MRI lower extremities with contrast (Figure 1): Fascial edema involving multiple muscles of both lower legs, predominantly the posterior muscles, and most pronounced around the medial heads of the gastrocnemius muscles bilaterally.

**DISCUSSION**

- **EOSINOPHILIC FASCIITIS (EF)** is a rare connective tissue disease of uncertain etiology and pathogenesis.
- **Incidence and prevalence**: unknown
- **Three stages of presentation**
  - **Stage I**: symmetric pitting and edema of extremities sparing fingers and toes
  - **Stage II**: development of severe induration of skin and subcutaneous tissues of affected sites
  - **Stage III**: may result in carpal tunnel syndrome, flexion contractures, muscle atrophy
  - **Laboratory work-up**: associated with peripheral eosinophilia, elevated ESR and CRP, hypergammaglobulinemia
  - **Diagnosis**: clinical presentation, MRI with contrast revealing evidence of fascial inflammation, full thickness skin-to-muscle biopsy
  - **Possible Triggers**: physical exertion, Lyme disease, medications such as statins, autoimmune diseases, and both benign and malignant hematologic diseases
  - **Treatment**: corticosteroids - either PO or IV
  - Patients may experience residual joint contractures which can greatly impact quality of life.

**Further Work-up and Management**

- **Serology and Chemistry:**
  - ANA, anti-centromere negative
  - CK level normal
  - ESR \(\uparrow\) 33 mm/hr (Ref <15 mm/hr)
  - CRP \(\uparrow\) 28.5 mg/L (Ref <8 mg/L)
  - Aldolase \(\uparrow\) 9.7 U/L (Ref <8.1 U/L)
- **Radiology:**
  - MRI lower extremities with contrast: Fascial edema involving multiple muscles of both lower legs, predominantly the posterior muscles, and most pronounced around the medial heads of the gastrocnemius muscles bilaterally.
- **Clinical presentation and MRI consistent with diagnosis of:**
  - **EOSINOPHILIC FASCIITIS**
- **Treatment:**
  - Patient was started on prednisone and physical therapy.
- **Follow-up on Month 8:**
  - Edema of extremities resolved however with persistent skin induration and achilles tendon contractures (Figure 2)
- **Figure 1:** residual skin induration demarcated from normal skin above (white arrow); with contractures of achilles tendons (black arrows)

**References:**