A TIGHT SITUATION: A CASE OF PROGRESSIVE SKIN INDURATION WITH ATYPICAL DISTRIBUTION

A CLINICAL VIGNETTE

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DISCLOSURES

- No financial disclosures or relationships with commercial entities

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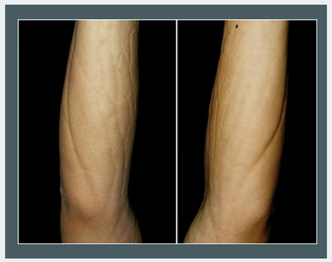
CASE HISTORY

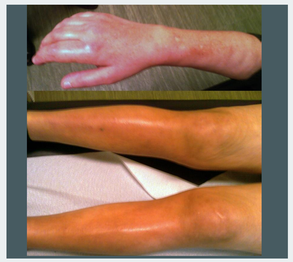
- 61-year-old male with a 6-month history of progressive, painful swelling and induration of his lower limbs as well as pruritus with sparing of his hands, feet, and face
- Associated difficulty sleeping and 30-lbs unintentional weight loss
- No sicca, Raynaud's, arthralgias, gastrointestinal, or neurologic symptoms
- Past medical history significant for hypertension and nephrolithiasis
- Medications included Hydrochlorothiazide, pantoprazole, gabapentin, naproxen and Tylenol as needed
- No current or prior substance use
- No contributory family history of malignancies, autoimmune or rheumatologic diseases

EXAMINATION FINDINGS

1. Symmetric, non-pitting induration, tightening, and skin thickening of the distal legs, thighs, and forearms, with areas of non-purulent exudate
2. Proximal muscle weakness (4/5) in upper and lower extremities
3. No joint contractures, p'eau d'orange, or signs of systemic sclerosis
4. Vital signs normal



https://www.mja.com.au/journal/2023/219/1/groove-sign-eosinophilic-fasciitis



https://www.journalmc.org/index.php/JMC/article/view/118/100

INVESTIGATIONS & DIAGNOSIS

- CRP 34 mg/L, fibrinogen 5.54 g/L, and haptoglobin 3.38 g/L. Iron studies showed low serum iron (5 µmol/L) and transferrin saturation (9%). Free kappa light chains were elevated (28.23 mg/L) but with a normal ratio and no monoclonality. CBC revealed a slightly elevated platelet count (441 × 10^9/L) with normal eosinophils
- CT thorax, abdomen, pelvis negative for malignancy
- A deep skin-to-muscle fascial biopsy of the right thigh showed fascial thickening, sclerosis, and lymphocytic inflammation extending into the muscle. Plasma cells were present, and eosinophils were rare – this made the diagnosis of Eosinophilic Fasciitis

DISCUSSION

Eosinophilic Fasciitis (EF) is a rare, scleroderma-like disorders characterized by inflammation and fibrosis of the fascia, typically affecting the limbs but sparing the face and fingers. EF remains underrecognized especially when classic features such as peripheral eosinophilia or p'eau d'orange skin are absent.

Clinical Presentation

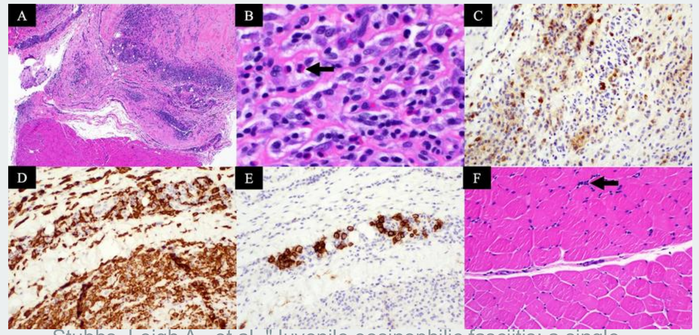
- Most commonly presents with painful swelling, induration, and tightness of the extremities
- In contrast to systemic sclerosis, EF usually spares the digits, face and internal organs although joint contractures and restricted range of motion may develop overtime
- The skin may have a woody texture or exhibit the "groove sign" which are liner depressions along veins due to fascial retraction (refer to first image on the left panel)
- Peripheral eosinophilia is a hallmark in many cases; however, it is not universally present
- Given overlap with many other disorders, it is important to entertain a broad differential

Box 1:Differential Diagnosis of Eosinophilic Fasciitis

- Localized scleroderma (morphea)
- Systemic sclerosis
- Nephrogenic systemic fibrosis
- Eosinophilia-myalgia syndrome
- Scleromyxedema
- Paraneoplastic fasciitis

Pathophysiology

- EF is thought to be an immune-mediated disease, possibly triggered by trauma, medications, or infections
- It may also occur as a paraneoplastic syndrome, particularly with hematologic malignancies such as myelodysplastic syndrome and lymphomas
- Histologically, EF shows fascial thickening, sclerosis, and a mixed inflammatory infiltrate, often with eosinophils, lymphocytes, and plasma cells
- Eosinophils may be sparse, but the fascial involvement with extension into adjacent muscle and preservation of the dermis are characteristic
- Superficial skin biopsies often miss the pathologic changes; thus, deep biopsies that include the fascia and muscle are essential of diagnosis



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Management

- First line treatment is high-dose corticosteroids. Early initiation is associated with better outcomes
- Immunosuppressive agents such as methotrexate, mycophenalate mofetil, and azathioprine are often added as steroid-sparing agents or in refractory cases
- Methotrexate remains a widely used first-line adjunct especially in patients with inadequate steroid response or contraindications to long-term steroid use
- Physical therapy is critical for maintaining mobility and preventing contractures
- Regular monitoring for complications such as joint restriction, secondary infection or hematologic abnormalities is essential

KEY POINTS

- Eosinophilic fasciitis (EF) may present without peripheral eosinophilia or classic skin findings.
- Early diagnosis and treatment with corticosteroids and immunosuppressants can improve outcomes.
- Deep fascial biopsy is crucial for diagnosis; superficial samples may miss pathologic changes.
- EF can mimic other scleroderma-like conditions and requires high clinical suspicion.

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