En Coup de Sabre

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En Coup de Sabre

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Case Presentation:
CC: I have developed a scar on my forehead
HPI: 45 y.o. AA female with a 2 month history of an enlarging scar on her forehead, which has become thicker, depressed and extremely pruritic.
PMT: HTN, GERD, DM type 2, Eczema
Medications: noncontributory
Allergies: NKDA
Family History: noncontributory
Social History: works as a direct care provider in a group home, no recent illnesses, smoker, denies alcohol or recreational drug use

Physical Exam: 1cm x 3.5cm hypo and hyperpigmented, atrophic linear patch on central forehead extending to the hairline.

Differential Diagnosis: Discoid lupus, linear morphea, caitaxia, Parry-Romberg syndrome, systemic sclerosis, scleroderm, morphea profunda, chemical/toxic exposure, lichen sclerosis, linear melorheostosis

Diagnostic procedure: 3mm punch biopsy central forehead and hairline

Pathology: Dermal Sclerosis compatible with morphea

Laboratory:
- CBC: mild normocytic anemia
- BMP: normal
- ANA: negative
- SCL-70: <1.0 (normal)

Diagnosis: En Coup de Sabre

MORPHEA
- Inflammatory disease of the dermis and subcutaneous tissue characterized by dermal sclerosis
- Represents a local form of scleroderma lacking internal organ involvement
- Sporadic; familial cases reported
- Genetic susceptibility - environmental triggers
- No clear HLA association
- Morphes
  - Generalized
  - Plaque-like
  - Linear
  - Morpheaform

PATHOGENESIS
- Vascular changes
- Microvascular injury
- Adhesion molecules expressed
- Endothelial swelling
- Thickening of basement membrane, intimal hyperplasia
- Activated T-cells stimulate connective tissue production by fibroblasts
- Pathologically enhanced collagen production by T-cell derived cytokines, IL-4 and TGF-B

PLAQUE-LIKE MORPHEA
- Clinical
  - Most frequent clinical presentation
  - Elevated erythematous or violaceous, expanding plaque
  - Central part of lesion becomes sclerotic and ivory-colored
- Course
  - Variable 3-5 years
  - Post inflammatory hyperpigmentation

LINEAR MORPHEA
- Clinical
  - Linear, erythematous
  - Band-like or circular
  - Lower and upper extremities, frontal head, thorax
- Involves fascia, muscle, tendon
- Course
  - Longer +/- progressive

EN Coup de Sabre
- Clinical
  - Linear band of atrophy
  - Median or paramedian
  - Resembles a sabre cut
  - Multiple lesions rare
  - Facial atrophy may occur
  - May involve muscles, bones, meninges and brain
- Course
  - Longer and occasionally progresive

PARRY-ROMBERG SYNDROME
- Varient linear morphea or separate entity
  - Hemifacial skin, tissue
  - Atrophy of subcutaneous fat
  - Muscular atrophy of face
  - Little or no sclerosis
  - Systemic involvement unknown
  - 10% epilepsy or neurological abnormalities

Prognosis
- Although 10x more prevalent than systemic sclerosis, most cases resolve within 3 years
- 10% develop functional limitations or disfigurement due to extent or location of disease

TREATMENT OF MORPHEA
- Topical and/or intralesional steroids
- Systemic corticosteroids – inflammatory stages of morphea with rapidly progressive linear or disabling morphea
- Do not improve established sclerosis
- Methotrexate 15-20mg/wk – in acute phase
- PCN 30x10IU/dy 3-4 weeks helpful (5%)
- Penicillamine – similarly effective, not used side effects
- Vitamin Derivatives
  - Actretin 10-50mg/day localized scleroderma, response seen after months
  - Calcitriol – antiinflammatory modulates fibroblast growth TGF-B

References: