En Coup de Sabre

Marie S. Lewars DO
Lehigh Valley Health Network, Marie_S.Lewars@lvhn.org

Gwyn Frambach DO

Charles Grooper MD

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Marie Lewars, DO; Gwyn Frambach, DO; Charles Grooper, MD
Lehigh Valley Health Network, Allentown, Pennsylvania

Case Presentation:

CC: I have developed a scar on my forehead
HPI: 45 y.o. AA female with a 2 month h/o an enlarging scar on her forehead, which has become lighter, depressed and extremely pruritic
PMH: 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 into the hairline
Differential Diagnosis: Discoid lupus, linear morphea, cicatraix, Parry-Romberg syndrome, systemic sclerosis, scleredema, morphea profunda, chemical/toxin 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

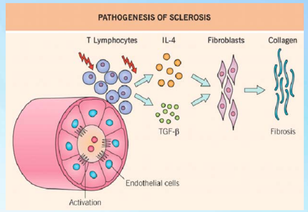
MORPHEA

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

	Systemic Sclerosis	Morphea (Localized scleroderma)
Major clinical variants	• Limited • Diffuse	• Plaque-type morphea • Generalized morphea • Linear morphea
Raynaud's phenomenon	++	-
Symmetric induration	++	-
Sclerodactyly	++	-
Facial involvement	+	- plaque-type and generalized +linear
Systemic involvement	++	-
Antinuclear antibodies	++	+ generalized and linear-plaque- type
Anticentromere antibodies	+ limited	-
Antitopoisomerase I	+ diffuse	-
Spontaneous remission	-	++ plaque-type + generalized ± linear

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

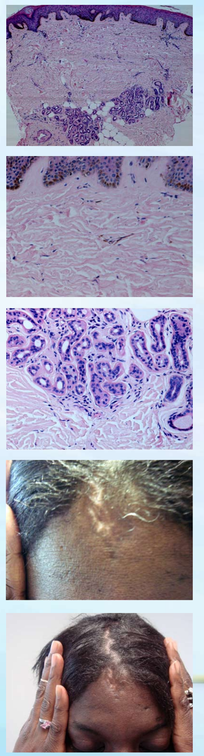
- Clinical
  - Most frequent clinical presentation
  - Elevated erythematous or violaceous, expanding plaque
  - Central part of lesion becomes sclerotic and ivory-colored
- Course
  - Variable 3-5 years
  - Post inflammatory hyperpigmentation

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 atraaophay
  - 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 progressive



PARRY ROMBERG SYNDROME

- Severe variant linear morphea or separate entity
  - Hemifacial skin, tissue
  - Atrophy of subcutaneous fat
  - Muscles and bones of face
  - Little or no sclerosis
  - Pathogenesis unknown
  - 10% epilepsy or neurological abnormatlities
- 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
  - Acitretin 10-50mg/day localized scleroderma, response seen after months
  - Retinoids - inhibit TGF-B
  - Calcitriol – antiinflammatory modulates fibroblast growth TGF-B

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