Morpheea in the Pediatric Patient

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Disclosures

I have no disclosures for this presentation
For today’s lecture:

- Definition, epidemiology, pathogenesis, classifications
- Clinical presentations of plaque and linear variants, diagnosis, complications, & treatment
- Clinical Case
What is morphea?

Also known as localized scleroderma

Fibrosing disorder of skin and subcutaneous tissues

Inflammatory disorder characterized by skin hardening
  ◦ Caused by increased collagen density from a combination of immune, genetic, and environmental factors

Systemic sclerosis-severe internal organ involvement, sclerodactyly, Raynaud phenomenon, and nail-fold capillary changes
Epidemiology

Incidence of morphea is 0.4 to 2.7% per 100,000 population

More common in Caucasians and females

Linear morphea has equal sex distribution

No lab markers are indicative of active disease but may correlate with disease extent

90% of children present between the ages of 2 and 14

Linear morphea is the most common form in children, but can have other types

- Often have a fhx of other autoimmune diseases
Pathogenesis

Both autoimmune and environmental factors (infection/trauma) lead to inflammation→ increase in collagen production and deposition into the skin

Mononuclear cells infiltrate the skin and surrounding blood vessels in early morphea→ functional and structural changes to the microvascular system

Long stretches of disease remission are common, but most patients with morphea develop new lesions over time
Clinical Presentation

Initial inflammatory stage: erythematous or violaceous patches or plaques
- Central region becomes white and sclerotic from local edema and increased collagen deposition, while the active borders remain red
- Color changes caused by increase in local temperatures

Later stages: active stage subsides and leaving behind sclerotic plaques that are white or hyperpigmented, skin becomes indurated and bound down

Overabundant collagen deposition destroys adnexal structures and hair follicles
Five subgroups of morphea-Mayo Clinic Classification

Circumscribed (plaque) morphea

Generalized morphea

Bullous morphea

Linear morphea
  ◦ En coup de sabre
  ◦ Progressive hemifacial atrophy
  ◦ Linear Limb

Deep morphea
  ◦ Subcutaneous morphea, morphea profunda, disabling pansclerotic morphea, and eosinophilic fasciitis
Plaque Morphea

< 3 discrete plaques, usually presents on the trunk in a well-circumscribed, oval shaped plaque

Superficial or Deep
  ◦ Superficial-limited to the epidermis and dermis
  ◦ Deep-dermis and SC tissue, +/- fascia and muscle

Often affects pressure areas (hips, waists, bra line, and proximal extremities)

NO systemic symptoms

Linear Morphea Variant

*En coup de sabre* - 1st described in 1854

- Usually paramedian forehead, frontoparietal dermis (muscle, bone, and CNS)
- Systemic symptoms: Headaches, CNS or vision changes
- Scalp: alopecia atrophic plaque of parietal scalp, usually shiny, smooth and often pigmented that may extend to cheeks, nose, and lip
  - Deforms bone

Linear Morphea Variant

**Progressive hemifacial atrophy or Parry-Romberg syndrome**
- Minimal cutaneous change, significant atrophy of adipose tissue, muscle and bone resulting in severe facial asymmetry
- Can overlap with en coup de sabre
- Mean onset age: 11
- Systemic symptoms: seizures
- Higher predominance in females
- CNS involvement in 20%
- Ophthalmic involvement in 15%

Linear Morphea Variant

Linear Limb:
- Unilateral arms or legs +/- muscle atrophy
- usually following Blaschko lines
- Systemic symptom: limb-length discrepancy, joint contracture
- Dermis and Subcutaneous tissue

- Most likely to have extracutaneous manifestations with linear morphea

Diagnosis

History and physical examination

Establish extent of skin involvement and signs of disease activity

Look for extracutaneous involvement

Skin biopsy

- Histology shows perivascular infiltrate of lymphocytes, admixed with rare eosinophils and plasma cells in the reticular dermis
- Thickened collagen bundles
- Later stages will have absent inflammatory cells, thickened collagen with atrophic eccrine glands, diminished number of blood vessels, trapped fat in dermis
Complications

In linear morphea: neurologic manifestations such as seizures, headaches, peripheral neuropathy, vascular malformations, brain calcifications, and CNS vasculitis
  ◦ Ocular complications with en coup de sabre (uveitis or episcleritis)
  ◦ Require screening even in the absence of symptoms

Depression and anxiety are more common

Associated autoimmune diseases reported in 2-5% of children with morphea
Treatment

Most lesions tend to regress spontaneously over 3-5 years leaving residual pigmentary or atrophic changes

- Aim to treat active areas of involvement

Physical therapy to prevent contractures

Ocular examination

Few true evidence-based studies regarding treatment, few small retrospective or prospective studies involving MTX, systemic steroids, cyclosporine, and mycophenolate mofetil
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<th>Table 3</th>
<th>Morphea treatment algorithm</th>
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<tr>
<td></td>
<td>Plaque</td>
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<tr>
<td>Active</td>
<td>Topical corticosteroids(^a)</td>
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<td></td>
<td>Topical tacrolimus</td>
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<td>Topical calcipotriene +/- betamethasone</td>
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<td>Residual sclerosis</td>
<td>Topical imiquimod</td>
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<td>Burnt-out</td>
<td>Cosmetic camouflage</td>
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\(^a\) Mid- to high-potency steroids are more effective; discontinue use if no signs of improvement in 2-3 months.
\(^b\) Corticosteroids could be administered as oral prednisone (1-2 mg/kg/d) or intravenous methylprednisolone (30 mg/kg/d for 3 days, monthly for 3 months).
\(^c\) Narrow band ultraviolet radiation could be used; ultraviolet A1 phototherapy has also been reported.
\(^d\) Fat injection, bone paste, fat transplant, maxillofacial reconstruction, tendon lengthening.

Consensus shows to treat for at least 2-3 years allowing for minimum 1 year of disease inactivity before discontinuing. 15-28% will still have recurrence especially with linear limb and older age of onset

Case 1

**HPI:** 10 year old Caucasian female presented, with her mother, with a 2 month history of two round red itchy rashes on the right and left abdomen. No color changes since it first appeared

**PE:** 9cm x 6cm round plaque with erythematous borders and slightly hypopigmented center on the right abdomen with a similar but smaller lesion on the left side

**In the office:** 2mm punch biopsy was done on the right abdomen

**DDx:** Morphea, Lichen sclerosis, MF, vitiligo, trauma
Case 1

Pathology: Superficial and deep perivascular and focal predominate lymphocytic dermatitis. No sclerosis. Compatible with inflammatory stage of morphea

- DDx: erythema chronicum migrans and less likely deep gyrate erythema or tumid lupus erythematosus. If clinically suspicious, suggest LYME and LE serology labs
Case 1

**Treatment:** Patient was placed on clobetasol ointment BID with calcipotriene ointment 0.005% BID and noted improvement

**Patient Course:** After 2 months, clobetasol was weaned off but patient had a flare

**Final recommendation:** calcipotriene BID with clobetasol 1-2 times per week and patient has since done well with no new flares
References


Thank you!