



Ivory Coloured, Slightly Indurated Atrophic Plaque

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This 12-year-old child presented with a 6 x 8 cm ivory coloured, mildly erythematous to hyperpigmented, slightly indurated, atrophic plaque on her left medial ankle, of nine months duration (Figure 1). When the lesion initially developed, it had an erythematous color with a violaceous border, but the erythema progressively faded resulting in an ivory plaque with central brown hyperpigmentation and loss of hair follicles. There was no history of trauma, associated systemic symptoms or other skin lesions. Past medical history and family history are unremarkable, with no history of autoimmune diseases.



Figure 1. Plaque on left medial ankle

What is your diagnosis?

- a. Lichen sclerosus et atrophicus
- b. Morphea (localized scleroderma)
- c. Nephrogenic sclerosing fibrosis dermopathy
- d. Amyloidosis
- e. Eosinophilic fasciitis

Answer: Morphea

About Morphea

Based on clinical and histologic findings, the patient was diagnosed with plaque-type morphea. Morphea is a hardening of the skin and superficial tissues limited to the skin, with

possible extension into subcutaneous fascia, muscle and bone. Morphea has been classified into five basic forms, each discernible by the skin lesion distribution and other findings, as described in Table 1.

Plaque-type morphea appears as well-circumscribed, indurated, erythematous to violaceous plaques on the skin. The violaceous inflammatory border (*i.e.*, “lilac ring”) is a characteristic feature of early morphea. Over the course of weeks to months, the center of the lesion evolves to an ivory colour and loses appendageal structures (*i.e.*, hair follicles). Most morphea lesions remain stable in size. With time, usually between three to five years for the most common plaque morphea lesions, the sclerotic plaques will soften, leaving behind a hyper or hypopigmented atrophic plaque or patch, often with overlying telangiectasias. Plaques can occur anywhere, however, plaque morphea frequently appears on the trunk.

Morphea usually occurs in isolation, but approximately 25% of patients demonstrate features of a systemic syndrome, such as arthralgia, dysphagia (from esophageal dysmotility), ocular symptoms (e.g., uveitis, episcleritis and keratitis), shortness of breath (from pulmonary fibrosis), fever, lymphadenopathy, and neurological involvement, *etc.*¹

The pathogenesis of morphea is unclear, but has been hypothesized to be due to abnormal fibroblast function, increased cytokine production and autoimmune dysfunction. A number of etiologic factors are associated, including microvascular injury, drugs (e.g., bleomycin, D-penicillamine), radiation therapy (e.g., for breast cancer

and other malignancies), localized trauma, infection by *Borrelia burgdorferi*, and genetics, although none have been confirmed as definitive causes.


Definitive diagnosis of morphea requires a skin biopsy to the level of subcutaneous fat or fascia (for morphea profunda) of either the active inflammatory border, or fibrotic center. Active lesions will demonstrate dense collagen bundles within a thickened dermis, extending to the level of subcutaneous fat or fascia.

Treatment for plaque-type morphea includes superpotent topical corticosteroids (e.g., clobetasol propionate 0.05% cream), vitamin D analogues

Table 1

Categories of Morphea

Type of Localized Scleroderma	Cutaneous Manifestations	Additional Notes
Plaque-type	<ul style="list-style-type: none"> Ivory to erythematous and/or violaceous (with lilac ring) indurated plaques involving few anatomic regions Plaque diameter ranges from 1 cm to over 20 cm 	<ul style="list-style-type: none"> Most benign form Often only a few lesions Least likely to have systemic involvement
Generalized	<ul style="list-style-type: none"> Numerous confluent plaques 	<ul style="list-style-type: none"> May lead to joint contractures and muscle atrophy More likely to have systemic involvement
Bullous	<ul style="list-style-type: none"> Tense subepidermal bullae on sclerotic plaques 	<ul style="list-style-type: none"> Secondary to scleratodermatous involvement Associated with previous trauma or lymphatic obstruction
Linear Scleroderma	<ul style="list-style-type: none"> Most common type in children, plaques are elongated and may involve dermis, subcutaneous tissue, muscle, and bone 	<ul style="list-style-type: none"> Important to investigate for systemic involvement En coup de sabre (frontoparietal linear morphea) may be associated with scarring alopecia and brain abnormalities May require extensive physiotherapy to prevent limb contractures
Deep (Profunda)	<ul style="list-style-type: none"> Poorly-defined, bound-down, sclerotic hyperpigmented plaques May involve the dermis, subcutaneous tissue, fascia, and superficial muscle 	<ul style="list-style-type: none"> Least common Can be very debilitating

(e.g., calcipotriene 0.005% ointment, calcitriol), combination calcipotriol-betamethasone dipropionate ointment, intralesional corticosteroids (e.g., triamcinolone acetonide), calcineurin inhibitors (e.g., tacrolimus ointment) and imiquimod 5% cream.²⁻⁴ Phototherapy may be an option for more diffuse cutaneous involvement.⁵ Of note, limited plaque-type morphea has been known to resolve spontaneously in three to five years without any treatment at all. 

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