Diagnosis and Management of Morphea and Lichen Sclerosus and Atrophicus in Children

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KEYWORDS

• Morphea • Localized scleroderma • En coup de sabre • Parry-Romberg syndrome
• Lichen sclerosus et atrophicus • Atrophoderma of Pasini and Pierini

KEY POINTS

- Early recognition may facilitate early intervention and decrease chances of residual cosmetic and functional problems.
- A team approach is beneficial in management of patients with morphea.
- Treatment of morphea depends on the extent, level of activity, and potential for cosmetic and functional disability.
- Treatment of lichen sclerosus et atrophicus alleviates symptoms.

INTRODUCTION

Overview

Morphea or localized scleroderma is a rare fibrosing disorder of the skin and underlying tissues. It is an inflammatory disorder characterized by skin hardening caused by increased collagen density resulting from a complex interplay of immune, genetic, and environmental factors. Morphea should be differentiated from systemic sclerosis (SSc) based on the appearance and distribution of the cutaneous manifestations and absence of severe internal organ involvement. Lichen sclerosus et atrophicus is a rare chronic inflammatory dermatosis that tends to affect primarily prepubertal girls. Both genital and extragenital involvement may occur. It can be coexistent with morphea, hence the inclusion of the entity in the morphea classification, or on its own with no evidence of other skin findings.

Disclosure: None to declare.
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Pediatr Clin N Am 61 (2014) 309–319
http://dx.doi.org/10.1016/j.pcl.2013.11.006 **pediatric.theclinics.com**
0031-3955/14/$ – see front matter

Epidemiology

Epidemiologic studies have reported incidence rates for morphea of up to 2.7 cases per 100, 000 population.[1] A female predominance of 2.4:1 has been noted.[2,3] Although morphea affects all races, it is more prevalent in Caucasians.[1–3] It usually starts in childhood, especially the linear subtypes, with nearly 90% of children presenting at a mean age of 7 years. There is another peak between 40 and 50 years of age for circumscribed or plaque morphea.[4]

The exact incidence of lichen sclerosus et atrophicus in childhood is not known, as most data on its frequency come from the specialized vulvar clinics that follow both adults and children. A prevalence of 1 in 900 cases has been reported.[5] Anogenital lichen sclerosus et atrophicus is primarily recognized in prepubertal girls.

Pathophysiology

The etiology and pathogenesis of morphea are not completely understood. A complex interplay of autoimmunity and environmental factors including possibly infection and/or trauma leads to local inflammation and ultimately increased collagen synthesis and deposition in the skin. This process is believed to occur due to increased fibroblast proliferation and extracellular matrix deposition through activation and release of transforming growth factor (TGF) α and β; platelet-derived growth factor (PDGF); connective tissue growth factor (CTGF); and interleukin (IL) 4, 6 and 8, among others; and a decrease in the collagen degradation through a decrease in the matrix metalloproteinases (MMPs).[6] A family history of autoimmune conditions is present in 12% to 24% of cases or morphea.[2,7] Moreover, 5% of children have other autoimmune diseases, and up to 69% of them have antinuclear antibodies.[7,8] Among infectious agents, *Borrelia* species organisms have been extensively studied, but their pathogenic role, particularly outside Europe, remains unclear.[6,9] Lichen sclerosus et atrophicus has a strong autoimmune association; 65% of a cohort of pediatric patients were homozygous for HLA-DQ7, as opposed to 5% of controls.[10]

HISTORY

Lesions of morphea occur insidiously. As they are largely asymptomatic, and the early stages of the disease are nonspecific, seeking medical attention is often delayed. Moreover, lack of familiarity with this condition further adds to delay in diagnosis and treatment. Significant delay has been described ranging from 6 months up to years.[11–13] Anogenital lichen sclerosus et atrophicus seems to be diagnosed within 1 year from onset, likely due to its symptomatic nature.[5] Extragenital lichen sclerosus et atrophicus is likely underdiagnosed, as it is generally asymptomatic.

CLINICAL FEATURES

The clinical picture of morphea depends on the stage (**Fig. 1**).[2,3,14,15] The inflammatory phase presents with local redness or violaceous discoloration. When the discoloration is at the periphery of the lesion, it creates the classical lilac ring appearance. It is often mistaken for other entities (see differential diagnosis) or overlooked. Color changes are accompanied by increased local temperature. Local edema and increased collagen deposition will ensue, particularly in the middle part of the lesion, often with waxy, shiny, white discoloration (porcelain-like). As the disease progresses toward more collagen synthesis, the skin becomes indurated and bound down. Burnt-out lesions are characterized by dyspigmentation (often hyperpigmentation), epidermal atrophy (shiny skin with visible venous pattern), dermal atrophy (cliff drop appearance), and