

CLINICAL VIGNETTE

Morphea: Evaluation, Management and Medical Implications of a Common Benign Skin Disorder

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Abstract

Morphea is a condition of localized inflammation and deposition of collagen occurring in the dermis, subcutaneous tissues or both, appearing grossly as thickened skin carrying potential functional and cosmetic defects. A lack of standardized scientific literature regarding its evaluation, treatment and impact has caused physicians to frequently misdiagnose and improperly treat this not uncommon dermatologic condition. Herein we present a case of morphea with an accompanying review of the literature and a discussion of its evaluation, management and implications for patients and providers.

Patient Presentation

A 50-year-old woman in overall good health presented to dermatology with a six-month history of slowly progressive, asymptomatic lesions on her trunk (inframammary and lateral abdomen, in areas of some friction from clothing). On physical exam, the lesions consisted of non-tender, 3-6 cm oval, whitish, waxy, sclerotic thin plaques, with subtle yet visible violaceous borders. An examination of her face and hands revealed no sclerodermoid changes and no capillary nail fold changes. A clinical

diagnosis of plaque type morphea was made and the patient agreed to a skin biopsy

Histopathology

A 4-mm punch biopsy of an active border of a lesion revealed thickened collagen bundles and lymphocytic perivascular infiltrate with rare plasma cells and eosinophils, consistent with morphea.

Work-up, Management

Limited autoantibody screening tests (ANA and RF) were negative, and the patient opted for topical tacrolimus therapy BID for two months resulting in a significant clinical softening of the lesions.

Discussion

Although often referred to as localized scleroderma, morphea must be differentiated from scleroderma (systemic sclerosis), as this nomenclature is a common source of confusion. Morphea is characterized as a fibrosing disorder of the skin and deeper tissues. Importantly, morphea lacks the classical findings of sclerodactyly, capillary nail fold changes, Raynaud's phenomenon and internal organ

involvement seen in scleroderma. The disease's cutaneous distribution and morphology may vary, but lesions progress from an early inflammatory stage to sclerosis and eventual atrophy of involved tissue. Excessive production of collagen by fibroblasts is the underlying pathophysiology in all forms of morphea although the specific etiology of the disease is unknown. Environmental, autoimmune, genetic and vascular factors have been hypothesized. Many report the occurrence of morphea after radiation, especially for breast cancer. The incidence of morphea is estimated to be approximately 0.4 to 3 per 100,000 people^{1,2}. Morphea presents in all races, but most frequently Caucasians³. Women are nearly three times more likely to develop morphea. Of note, half of all morphea cases present in childhood.

Morphea initially presents as an erythematous, soft or eventually indurated patch or plaque, often in areas of friction, trauma, or body folds, such as beneath the breasts or near the groin. The plaques eventually develop a central region of sclerosis, appearing smooth and hypopigmented, surrounded by an active erythematous or violaceous border. The active lesions may be accompanied by localized pruritus, pain or numbness. As the inflammation resolves, the remaining lesion may become shiny and sclerotic, with hyper- or hypopigmentation and secondary alopecia. These lesions can become atrophic and extend as deeply as the subcutis, fascia, muscle and bone, producing joint contractures, limb discrepancies and functional impairments, especially in the pediatric population. This is the major cause of morbidity associated with morphea.

The distribution, depth and morphology of the lesions define the subtypes. These major subtypes, in order of prevalence, are: circumscribed, generalized, linear or mixed⁴. Children predominantly present with linear morphea, which can be cosmetically disfiguring and physically debilitating. Circumscribed morphea, also known as plaque morphea, is the most common form in adults with peak incidence in the third and fourth decade and is associated with lower morbidity³. The most frequent complications of morphea include arthralgias, uveitis, and joint contractures, especially in the linear and deep forms¹. Linear morphea, which usually follows Blaschko's lines, can present on the face and produce Parry-Romberg syndrome, or hemiatrophy of the skin, soft tissues, muscles and bones of the face⁵. The course of the disease is generally slow and chronically active over a period of years, however, reactivation may follow remission in approximately twenty-percent of cases³. Patients may also

experience multiple lesions at varying stages of development⁶.

Some of the diseases commonly included on the differential diagnosis of morphea include: scleroderma, lipodermatosclerosis, eosinophilic fasciitis, chronic graft-versus-host disease, lichen sclerosis and porphyria cutanea tarda. The diagnosis of morphea is made clinically based on history and physical examination, as histopathology is usually unnecessary. Its heterogeneous and sometimes changing appearance over time may complicate the diagnosis, making a clear history of lesion evolution useful. Recent studies have reported a significant delay in the accurate diagnosis of morphea, with widely variable treatments depending upon the specialty of the physician⁷. Some experts suggest that early, active lesions of morphea are more responsive to therapy, thus emphasizing the importance of swift and proper evaluation. Atypical presentations of suspected morphea may warrant biopsy to aid diagnosis and treatment. Histologically, active morphea is characterized by lymphocytic perivascular infiltrate of the dermis, and may include evidence of inflammation in the subcutis. In late sclerotic lesions, thickened collagen bundles are seen in the reticular dermis, with a loss of blood vessels and dermal appendages. The pattern and quantity of excess collagen deposition seen in the dermis on histology have been reported to correlate with the clinical subtypes and their severity, suggesting the potential use of histopathology to more accurately guide therapy (i.e. superficial versus deep)⁸. More recent studies have incorporated the use of ultrasonography as an alternative evaluation of morphea based on subcutaneous tissue echogenicity, cutaneous blood flow and dermal thickening⁹. Magnetic resonance imaging is also used in cases of suspected deep tissue involvement.

Due to the relatively low prevalence and lack of standard outcome measures, there is a relative lack of evidence-based treatments for morphea. In studies monitoring disease activity, results have shown that morphea can produce significant permanent morbidity and that treatment of early, active disease is more efficacious, supporting the initiation of treatment as early as possible¹⁰. The most well supported treatment options include phototherapy, methotrexate with or without systemic steroids and ultraviolet (UV) phototherapy, topical tacrolimus, and topical calcipotriene. The choice of therapy is governed by the relative activity of the lesions, depth of involvement, distribution, and duration. Phototherapy with narrowband UVB and UVA1 is

supported for diffuse superficial lesions¹¹. Treatment with topical tacrolimus or calcipotriene have shown efficacy when used for superficial, inflammatory plaques¹². Systemic treatment using methotrexate in combination with corticosteroids and phototherapy is indicated in cases of severe morphea possessing the features of widespread distribution, rapidly progressive disease, and subcutaneous involvement, especially when leading to functional or cosmetic defects¹⁰. Important to note, many patients diagnosed initially with circumscribed morphea will later progress to more extensive linear or generalized morphea. Thus, patients who present with only a few plaques should receive regular follow-up to ensure that the treatment is adequate for the trajectory of the disease.

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