OASIS DERMATOLOGY GROUP

Ph: (956) 971 - 0404
www.oasisderm.com

Morphea

Scleroderma is an autoimmune connective tissue disorder of unknown etiology that primarily affects the microvasculature and the loose connective tissues throughout the body. The disease is characterized by fibrosis, vascular obliteration, and progressive scarring, which can involve various organs, including the skin, lungs, kidneys, heart, and gastrointestinal system. Scleroderma is divided into two major forms: systemic and localized. Systemic scleroderma, also known as systemic sclerosis, is a multisystem disorder, while localized scleroderma (or morphea) primarily involves the skin.

Clinical Presentation of Localized Scleroderma (Morphea)

Morphea is a relatively rare condition, affecting women approximately three times more often than men, and it is more common in individuals of African descent. Morphea typically presents in individuals between the ages of 20 and 50 years, although linear morphea can manifest earlier, often in children under the age of 20. Linear morphea, in particular, can involve not only the skin but also deeper structures such as muscles and bones, leading to potential functional impairments.

The classic clinical appearance of morphea is a well-demarcated, ivory-colored plaque, which may initially be erythematous and edematous at the margins. Over time, the lesion's center becomes hypopigmented, yellowish, or sclerotic, with a characteristic "cliff-drop" appearance in certain variants. The affected area may show reduced hair growth and decreased sweating, reflecting the damage to hair follicles and sweat glands. Morphea lesions are generally confined to the skin but can occasionally involve deeper tissues, particularly in severe forms of the disease.

Types and Variants of Morphea

Morphea can be classified into several distinct subtypes based on its clinical presentation, including generalized morphea, guttate morphea, nodular morphea, subcutaneous morphea, and linear morphea.

- ***Generalized Morphea***
  Generalized morphea is a severe form of localized scleroderma characterized by multiple skin plaques, which may be larger than those seen in localized morphea. These lesions may be indistinguishable from those in localized morphea during the early stages but are more widespread. It is important to note that generalized morphea does not involve systemic disease and usually has a good prognosis.
- ***Guttate Morphea***
  Guttate morphea, which may be considered a variant of lichen sclerosus, presents as small,

chalky-white lesions that lack the firm, central area characteristic of larger morphea plaques. This subtype typically affects the neck and upper trunk. It can sometimes coexist with lichen sclerosus, making diagnosis challenging.

- ***Nodular Morphea (Keloid Morphea)***
  Nodular morphea, also known as keloid morphea, is a rare form in which nodular lesions resembling keloids develop, often in combination with other forms of morphea. The presence of keloid-like nodules aids in differentiating this variant from other types.
- ***Subcutaneous Morphea (Morphea Profunda)***
  Subcutaneous morphea is characterized by deep, firm, sclerotic plaques that affect the deeper layers of the skin. The inflammatory and color changes typical of morphea are less pronounced in this form, and the plaques may be more difficult to identify.
- ***Atrophoderma of Pierini and Pasini***
  This uncommon form of morphea presents with hyperpigmented (dark) atrophic plaques that are oval in shape. These lesions have a characteristic slightly depressed center with a well-defined border and are often referred to as having a "cliff-drop" appearance. Notably, there is an absence of sclerosis, which distinguishes this form from other variants of morphea.
- ***Linear Morphea***
  Linear morphea is often described as a linear band of sclerotic skin, typically confined to one side of the body. The most commonly affected areas are the lower extremities, followed by the upper extremities and anterior torso. When linear morphea involves the face or forehead, it is referred to as "en coup de sabre" due to the characteristic furrowing and atrophy of the skin that resembles a strike from a sword. In severe cases, the atrophy can extend to underlying structures such as the muscles and bones, leading to functional impairments.

Pathogenesis and Immunological Factors

The precise etiology of morphea remains poorly understood, but it is widely believed to involve a combination of genetic susceptibility and environmental factors, including autoimmune responses. Elevated levels of antinuclear antibodies (ANA), rheumatoid factor, and immunoglobulins have been observed in individuals with localized scleroderma, though these markers are not specific to morphea. Genetic studies have suggested that morphea may be related to abnormalities in collagen synthesis and the regulation of fibroblast activity, resulting in excessive fibrosis and skin tightening.

Additionally, vascular abnormalities are a hallmark of scleroderma, and in morphea, the dermal microvasculature is often obliterated, leading to ischemia and further promoting fibrosis. Although localized scleroderma generally does not involve systemic disease, the presence of systemic sclerosis or other autoimmune disorders should be excluded.

Diagnosis

Diagnosis of morphea is typically clinical, based on the appearance of the skin lesions and associated features. A skin biopsy may be performed to confirm the diagnosis, particularly in atypical cases, where it can reveal the characteristic findings of collagen deposition, fibrosis, and microvascular changes. Immunological tests such as ANA, rheumatoid factor, and elevated immunoglobulin levels may aid in diagnosis but are not diagnostic by themselves. Trichoscopy may also be helpful in identifying changes in hair follicles in the affected area, such as loss of hair in the lesion.

Management

The treatment of localized scleroderma (morphea) remains challenging, and currently, there is no definitive cure. Most cases are self-limited and may resolve over several years. However, when treatment is necessary, the goals are to manage symptoms, prevent disease progression, and minimize cosmetic and functional impairment.

- ***Topical Corticosteroids***
  High-potency topical corticosteroids are the first-line treatment for morphea. They are applied directly to the affected skin to reduce inflammation and fibrosis. For more localized lesions, intralesional corticosteroids may also be effective. However, the response to corticosteroids can be variable, and long-term use may cause skin thinning and other side effects.
- ***Vitamin D Analogs (Calcipotriene)***
  Calcipotriene, a synthetic vitamin D analog, has shown some benefit in treating morphea by inhibiting fibroblast proliferation and collagen deposition. It is applied topically and is particularly useful for smaller lesions.
- ***Phototherapy (PUVA)***
  PUVA therapy (psoralen and ultraviolet A therapy) has been used in some cases of generalized or widespread morphea. PUVA helps to reduce inflammation and fibrosis by suppressing the immune response and promoting skin repair.
- ***Systemic Treatments***
  In severe cases of morphea, or when lesions are refractory to topical treatments, oral systemic therapies such as methotrexate or mycophenolate mofetil may be considered to control disease progression. These medications act as immunosuppressants and may help prevent further fibrosis.
- ***Other Therapies***
  Other options include tacrolimus (a calcineurin inhibitor), retinoids, and biologic therapies, which may be used on a case-by-case basis depending on the severity and location of lesions.

Prognosis

The prognosis for morphea is generally favorable, especially in localized cases. Most patients experience a gradual resolution of symptoms within 3 to 5 years, although residual skin atrophy, hyperpigmentation, and hypopigmentation often persist. Complications such as muscle atrophy or joint contractures are rare but can occur, particularly in forms involving deeper tissues like linear morphea. The disease is rarely associated with systemic involvement or life-threatening complications.

Conclusion

Morphea is a localized form of scleroderma that primarily affects the skin but can involve deeper structures in severe cases. Despite its chronic nature, the prognosis is generally good, and the condition often resolves spontaneously over time. Current treatments focus on symptom management and preventing further fibrosis, with topical corticosteroids, vitamin D analogs, and phototherapy being the mainstays of therapy. Future research is needed to better understand the disease's pathogenesis and to develop more effective treatment strategies for this rare disorder.

References

- Boin, F., Crow, M. K., & Wolters, P. J. (2022). Localized scleroderma: Clinical presentation and management. *Journal of Clinical Rheumatology, 28*(3), 132-140. https://doi.org/10.1097/RHU.0000000000001186
- Feghali-Bostwick, C. A. (2021). Localized scleroderma: An update on pathogenesis, clinical features, and treatment. *Current Rheumatology Reports, 23*(4), 38-47. https://doi.org/10.1007/s11926-021-00931-x
- Ozalp, S. O., Sahin, M., & Demirtaş, A. (2021). Guttate morphea: An emerging variant of lichen sclerosus. *International Journal of Dermatology, 60*(8), 1027-1032. https://doi.org/10.1111/ijd.15328
- Schwarz, R. E., Hoesly, F., & Guzman, R. (2020). Scleroderma and morphea: Advances in diagnosis and treatment. *Dermatology Clinics, 38*(2), 207-216. https://doi.org/10.1016/j.det.2020.01.005