Acad Med J 2025;5(Suppl 1):S133-S138.
UDC: 616.5-003.829-085
https://www.doi.org/10.53582/AMJ2551133a
Case report

GENERALIZED MORPHEA: CASE REPORT AND TREATMENT APPROACH

Adjievska Nevenka, Zhaku Sevxhane, Pollozhani Nora, Rushiti Mehmeti Kujtime, Dimova Maja, Popovski Tomche, Zyberi Ramuka Rufije

University Clinic for Dermatology, Faculty of Medicine, Ss. Cyril and Methodius University in Skopje, Republic of North Macedonia
e-mail: nadzievska@yahoo.com

Abstract

Morphea, also known as localized scleroderma, is a rare autoimmune disorder characterized by excessive collagen deposition, leading to fibrosis of the skin. Unlike systemic scleroderma, morphea does not affect internal organs but can cause significant morbidity due to skin hardening and potential involvement of underlying tissues such as muscles and joints.

An 11-year-old female patient was hospitalized for the first time at the Dermatology Clinic due to hyperpigmented skin lesions. According to the heteroanamnesis provided by her father, the lesions initially appeared on her thighs approximately one year ago, with an estimated size of around 20 cm. She was treated on an outpatient basis by a dermatologist, though no details on the therapy were provided. One month prior to hospitalization, the lesions worsened, expanding to the lower legs, chest, and upper extremities, accompanied by itching. Dermatological assessment revealed erythematous-livid, slightly indurated patches with a shiny, smooth surface. The lesion demonstrated areas of coalescing hyperpigmentation and hypopigmentation, distributed across the chest, abdomen and extremities. A skin biopsy was performed and confirmed early inflammatory-stage morphea.

Phototherapy, particularly UVA is an effective treatment for early-stage and inflammatory morphea, helping to reduce fibrosis, restore skin flexibility, and improve pigmentation. When combined with immunosuppressive agents, it enhances clinical outcomes and slows disease progression. Early intervention, guided by a multidisciplinary team, leads to a favorable prognosis, though long-term follow-up is essential to monitor recurrence and optimize management.

Keywords: morphea, phototherapy, pediatric dermatology, inflammation

Introduction

Localized scleroderma (morphea) is a chronic autoimmune disorder characterized by excessive collagen deposition, leading to fibrosis of the skin and, in some cases, deeper tissues such as muscles and joints [1]. Unlike systemic scleroderma, morphea does not affect internal organs but can cause significant morbidity depending on its severity and extent [2]. The disease manifests in various clinical forms, classified into five major subtypes: plaque, generalized, bullous, linear, and deep morphea [3]. Among these, generalized morphea is considered one of the most severe subtypes due to its widespread distribution and potential for functional impairment [4].

The exact pathogenesis of morphea remains uncertain; however, multiple factors, including autoimmune mechanisms, genetic predisposition, and environmental triggers, are thought to contribute to its onset. Infectious agents such as *Borrelia burgdorferi*, trauma, and radiation exposure have been implicated as potential triggers [5]. Morphea primarily affects the

microvasculature and loose connective tissue, leading to inflammation, fibrosis, and skin hardening[6]. Although primarily cutaneous, severe cases may involve deeper structures, resulting in joint contractures and restricted mobility, particularly in pediatric patients [7].

Pediatric morphea, particularly the generalized subtype, is a rare but challenging condition due to its potential impact on growth, development, and quality of life [8]. Children with generalized morphea often present with widespread, indurated plaques that can cause skin tightness, pain, and limited joint mobility, leading to functional impairments if untreated [9]. Given its progressive nature, early diagnosis and intervention are crucial to managing disease progression and preventing complications.

Case report

An 11-year-old girl was hospitalized for the first time at the Dermatology Clinic due to hyperpigmented skin lesions. According to the heteroanamnesis provided by her father, the lesions initially appeared on her thighs approximately one year ago, with an estimated size of around 20 cm. She was treated on an outpatient basis by a dermatologist, though no details on the therapy were provided. One month prior to hospitalization, the lesions worsened, expanding to the lower legs, chest, and upper extremities, accompanied by itching. Dermatological assessment revealed erythematous-livid, slightly indurated patches with a shiny, smooth surface. The lesion demonstrated areas of coalescing hyperpigmentation and hypopigmentation, distributed across the chest, abdomen and extremities (Fig. 1). The patient often complained of itching. Prompt recognition and treatment at this stage are crucial to prevent disease progression and fibrosis. A skin biopsy was performed and confirmed early inflammatory-stage morphea (Fig. 2).

A physical examination revealed no abnormalities in major organ systems. Dermatological assessment showed erythematous or violaceous patches with a characteristic lilac-colored border, indicating active inflammation. Mild induration may be present due to early collagen deposition.

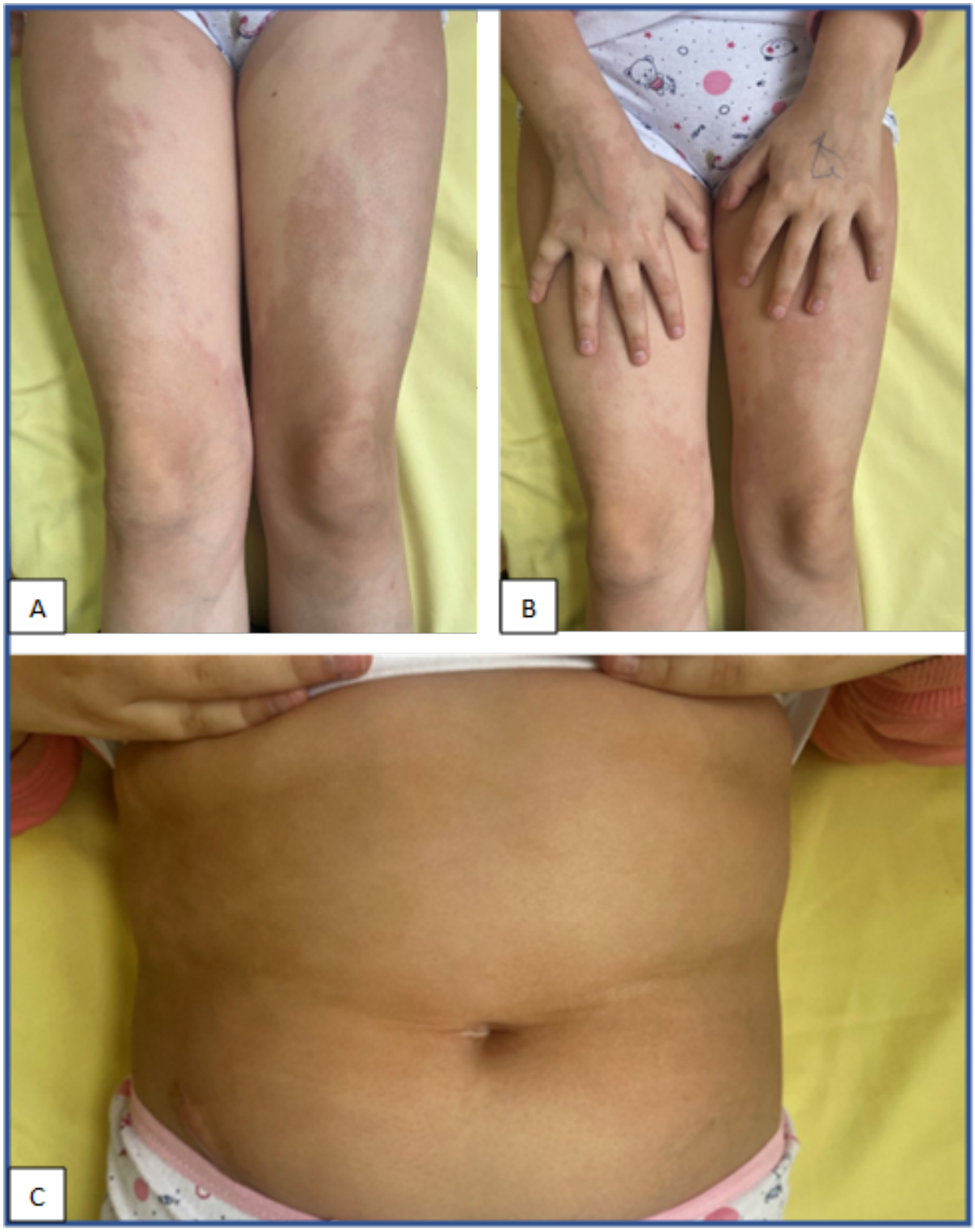

Fig. 1 Physical examination of the patient. **A**: Erythematous hyperpigmented patches in the legs. **B:** Erythematous hyperpigmented patches in the arms. **C:** Erythematous hyperpigmented patches in the chest.

Laboratory tests, including blood count, glucose, enzyme, protein, lipid, immunological, and hormonal status, were within normal ranges, except for an elevated ASO (389) and CRP (5.29). Morphea activity measure (MAM) SCORE is with moderate activity, which require topical therapy and phototherapy as a recommended treatment approach.

A skin biopsy confirmed early inflammatory-stage morphea. Microscopically, the tissue samples showed a section of skin composed of the epidermis and dermis with skin adnexa. The epidermis exhibited flattened dermo-epidermal papillae, slight atrophy, and orthokeratosis. In the papillary and reticular dermis, proliferated collagenous connective tissue was observed, with visible thick eosinophilic collagen bundles, some trapped skin adnexa, and a discrete lymphocytic inflammatory infiltrate (Fig. 2).

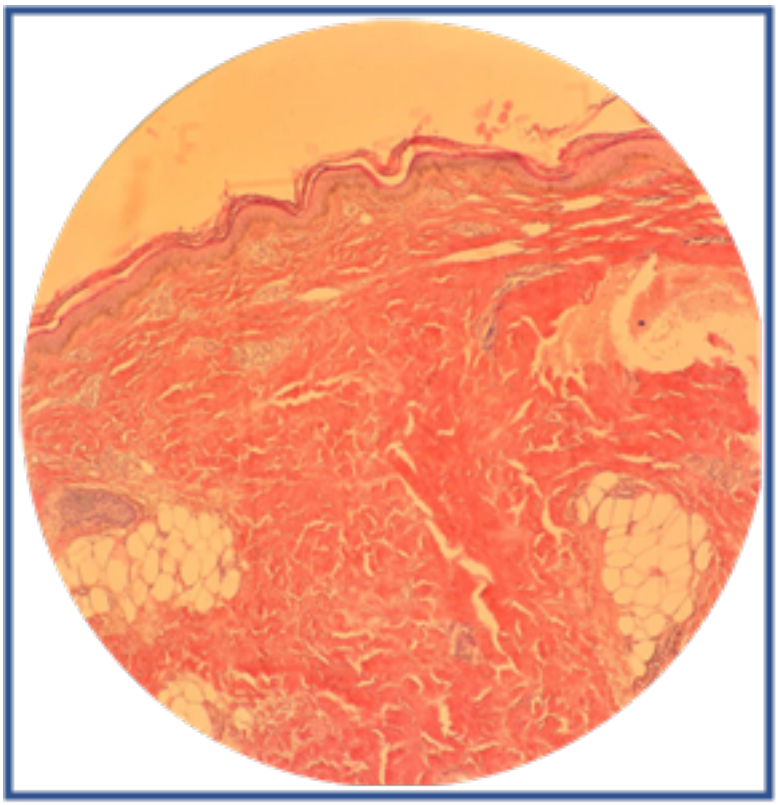

Fig. 2 Histopathological findings

The patient's clinical history, laboratory analysis and histopathologic examination were consistent with generalized morphea, a rare subtype of localized scleroderma.

The patient was treated with 30 UVA phototherapy sessions, along with topical corticosteroids and calcineurin inhibitors. The combination of phototherapy and immunosuppressive topical agents was administered to manage the disease progression. There was a significant improvement in the skin changes.

Discussion

Localized scleroderma (morphea) presents significant challenges in both diagnosis and management due to its variable clinical manifestations and unpredictable progression. While morphea is distinct from systemic scleroderma in its lack of internal organ involvement, its potential to cause substantial morbidity especially in generalized and pediatric cases necessitates a comprehensive approach to treatment and monitoring. The broad spectrum of

clinical subtypes further complicates disease characterization and underscores the need for individualized therapeutic strategies.

The pathogenesis of morphea remains incompletely understood, though emerging evidence supports a multifactorial origin involving autoimmune mechanisms, genetic susceptibility, stress and environmental triggers [10]. The proposed association with *Borrelia burgdorferi*, though debated, highlights the potential role of infectious agents in initiating or exacerbating disease onset. Additionally, factors such as trauma and radiation exposure emphasize the importance of external influences in disease pathogenesis [11]. Further research into these mechanisms is essential for developing targeted therapies that address both the inflammatory and fibrotic components of the disease.

Pediatric morphea, particularly the generalized subtype, poses unique challenges due to its impact on growth and development. The risk of joint contractures and functional impairment in children underscores the need for early and aggressive intervention. Delayed diagnosis can lead to irreversible complications, highlighting the critical role of heightened clinical awareness among pediatricians and dermatologists. Current treatment options, including systemic immunosuppressive therapies and physical rehabilitation, aim to mitigate disease progression and improve quality of life [12]. However, long-term studies are needed to establish standardized treatment protocols and optimize patient outcomes. Phototherapy, particularly UVA1 and narrowband UVB (nbUVB), has proven to be an effective and well-tolerated treatment modality for early-stage and inflammatory morphea, facilitating fibrosis reduction, skin flexibility restoration, and pigmentation improvement. UVA phototherapy is the most effective treatment, penetrating deeply into the skin to reduce inflammation and fibrosis. nbUVB is useful for superficial lesions but is less effective than UVA. When combined with immunosuppressive agents, phototherapy enhances clinical outcomes and mitigates disease progression. Overall, phototherapy has shown promising results, particularly when used alongside topical or systemic immunosuppressive therapy in active disease. Topical corticosteroids work well in the early inflammatory stages by reducing inflammation, but long-term use can lead to skin atrophy. Calcineurin inhibitors offer a safer alternative for long-term maintenance, helping to suppress inflammation without causing skin thinning.

This combination therapy is effective for localized and mild forms of morphea, helping to control disease progression. These treatments are valuable for mild and localized morphea, especially in early stages. However, in more severe or rapidly progressing cases, systemic therapy is often needed. Combined approaches, such as phototherapy with topical immunosuppressants, enhance treatment efficacy while minimizing side effects.

Conclusion

Given the chronic and potentially debilitating nature of morphea, a multidisciplinary approach is essential for effectively managing affected individuals. Future research should focus on elucidating disease mechanisms, identifying reliable biomarkers for early detection, and exploring novel therapeutic options. Advancements in understanding the pathophysiology of morphea will not only improve patient care but also contribute to the broader field of autoimmune and fibrotic disorders. Early intervention, supported by a multidisciplinary team, can result in a favorable prognosis. However, long-term follow-up remains crucial to monitor for recurrence and to optimize ongoing management strategies.

Conflict of interest statement. None declared.

References:

1. Dimitrova V, Yordanova I, Gospodinov DK. Generalized morphea: a case report. *J IMAB*. 2007;13(1):68-70. doi: 10.5272/jimab.2007131.64.
2. Cunningham BB, Landells ID, Langman C, Sailer DE, Paller AS. Topical calcipotriene for morphea/linear scleroderma. *J Am Acad Dermatol*. 1998 Aug;39(2 Pt 1):211-5. doi: 10.1016/s0190-9622(98)70077-5.
3. Falanga V, Medsger TA Jr, Reichlin M, Rodnan GP. Linear scleroderma. Clinical spectrum, prognosis, and laboratory abnormalities. *Ann Intern Med*. 1986 Jun;104(6):849-57. doi: 10.7326/0003-4819-104-6-849.
4. Hunzelmann N, Anders S, Fierlbeck G, Hein R, Herrmann K, Albrecht M, et al. Double-blind, placebo-controlled study of intralesional interferon gamma for the treatment of localized scleroderma. *J Am Acad Dermatol*. 1997 Mar;36(3 Pt 1):433-5. doi: 10.1016/s0190-9622(97)80221-6.
5. Ihn H, Sato S, Fujimoto M, Kikuchi K, Takehara K. Clinical significance of serum levels of soluble interleukin-2 receptor in patients with localized scleroderma. *Br J Dermatol*. 1996 May;134(5):843-7.
6. Leroy EC. Connective tissue synthesis by scleroderma skin fibroblasts in cell culture. *J Exp Med*. 1972 Jun;135(6):1351-62. doi: 10.1084/jem.135.6.1351.
7. Kucharz EJ, Sierakowski SJ. Metabolizm tkanki Lacznej w twardzinie uogólnionej [Metabolism of the connective tissue in generalized scleroderma]. *Postepy Hig Med Dosw* (Online). 1989;43(1):93-102. Polish.
8. Sink JR, Chiu YE. Pediatric morphea state-of-the-art literature review: Reframing morphea as a systemic disease. *Pediatr Dermatol*. 2021 Sep;38(5):1020-31. doi: 10.1111/pde.14688.
9. Li SC. Scleroderma in Children and Adolescents: Localized Scleroderma and Systemic Sclerosis. *Pediatr Clin North Am*. 2018;65(4):757-81. doi: 10.1016/j.pcl.2018.04.002.
10. Khan Mohammad Beigi P. The Immunogenetics of Morphea and Lichen Sclerosus. *Adv Exp Med Biol*. 2022;1367:155-72. doi: 10.1007/978-3-030-92616-8_7.
11. Spalek M, Jonska-Gmyrek J, Gałecki J. Radiation-induced morphea - a literature review. *J Eur Acad Dermatol Venereol*. 2015;29(2):197-202. doi: 10.1111/jdv.12704.
12. McGaugh S, Kallis P, De Benedetto A, Thomas RM. Janus kinase inhibitors for treatment of morphea and systemic sclerosis: A literature review. *Dermatol Ther*. 2022 Nov;35(6):e15437. doi: 10.1111/dth.15437.