

Disabling pansclerotic morphea of childhood with skin ulceration and tendon retraction: A case report

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ABSTRACT

Disabling pansclerotic morphea of childhood (DPMC) is a rare, severe form of generalized morphea with significant diagnostic and therapeutic challenges. It is characterized by deep tissue involvement, extending to the muscles, tendons, and bones, leading to complications such as joint stiffness, deformities, and ulcerations. Herein, we report the case of a six-year-old girl with a five-month history of widespread sclerosis involving the lower limbs, acral areas, severe ulcerations, and tendon retractions. Histopathological evaluation confirmed DPMC, revealing a thickened dermis with eosinophilic collagen bundles and atrophic adnexal structures, without dermal homogenization. Treatment included monthly corticosteroid pulses, methotrexate, and physiotherapy. While partial improvement was observed, with reduced joint stiffness and improved mobility, significant sclerosis and depigmentation persisted, underscoring the disease's refractory nature. This case highlights the variability of DPMC, the importance of early recognition, and the need for aggressive multidisciplinary care. Further research is essential to optimize outcomes and establish standardized protocols for this condition.

Key words: Disabling pansclerotic morphea, Childhood, Ulceration, Tendon retraction, Multidisciplinary treatment

INTRODUCTION

Disabling pansclerotic morphea of childhood (DPMC) is a rare and severe form of localized scleroderma, primarily affecting children under the age of 14, with a prevalence of less than 1 per 10,000,000 [1]. It is characterized by systemic inflammation, extensive skin and soft tissues, and occasionally extends to the underlying muscles and bones.

The Juvenile Scleroderma Working Group of the Pediatric Rheumatology European Society identified linear morphea as the most common type (65%), followed by plaque-type morphea (26%), generalized morphea (7%), and deep morphea (2%) [2]. DPMC is recognized as a rare subtype of deep morphea [3].

DPMC may lead to complications such as muscle and skeletal atrophies, joint contractures, ankylosis, trophic

skin ulcers, nerve compressions, and an increased risk of skin cancer [4-6]. Although its etiology remains unclear, mechanisms such as endothelial cell injury, immune activation, and fibroblast hyperactivity, resulting in excessive collagen synthesis, are proposed. Genetic predisposition and environmental factors may also play a role.

Herein, we report the case of a six-year-old child with DPMC presenting with atrophic sclerosis over five months and significant cutaneous and articular complications. This case highlights the rarity and rapid progression of DPMC, contributing to the limited literature on this condition.

CASE REPORT

A six-year-old girl with no prior medical history presented with irregular, infiltrated plaques, beginning

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at the root of the left thigh and extending over the entire lower limb for five months. Additionally, the patient reported widespread joint pain, morning stiffness, fatigue, and weight loss. Developmental milestones were normal, and she was up-to-date with vaccinations. The family history was unremarkable.

On examination, skin sclerosis extended from the pubic region to the dorsum of the left foot, with sclerotic lesions noted over the toes of the right foot and along the lateral aspect of the right thigh. Furthermore, the patient exhibited tight, shiny skin on the proximal interphalangeal (PIP) and metatarsophalangeal (MTP) joints. Mild desquamation, hypopigmentation, and hyperpigmentation patches were noted (Figs. 1a – 1d). Two ulcerations were identified: one on the external right thigh (4 cm) and another on the internal left thigh, (7 cm), with irregular edges, fibrinous bases, and inflammatory halos on a scleroatrophic base (Fig. 2).

Functional assessment revealed a reduced range of motion in the left knee and ankle, attributed to hamstring tendon retraction and cutaneous sclerosis, causing walking difficulty.

Table 1 summarizes the initial laboratory investigations. X-rays of the left knee and ankle showed preserved joint spaces without structural abnormalities. Histopathological examination (Figs. 3a and 3b) revealed a thickened mid-dermis with eosinophilic collagen bundles surrounding the adnexal structures. There was no evidence of dermal tissue homogenization. The adnexal structures were atrophic, consistent with the features of DPMC. Lung function tests, capillaroscopy, as well as cardiac and ophthalmologic evaluations, revealed no abnormalities.

Treatment included monthly pulses of glucocorticosteroids (methylprednisolone 15 mg/kg/day for three days) for six months and methotrexate 15 mg/m²/week and physiotherapy. Ulcer care involved local wound management and debridement with antimicrobial dressings.

After three months, the patient showed partial improvement. Joint stiffness improved, and mobility increased, yet skin sclerosis and depigmentation persisted. This partial response highlighted the complexity of managing DPMC and the need for ongoing care.

DISCUSSION

This case highlighted the rarity and complexity of DPMC, a severe and progressive form of generalized morphea classified as an autoimmune disease. It deeply affects the subcutaneous tissue, muscles, tendons, and bones, leading to complications such as joint stiffness, deformity, ulcerations, and calcifications. Lesions typically involve the extensor surfaces of the limbs and trunk [7], sparing acral areas; however, in our case, acral areas were notably affected. Additionally, ulcerations and tendon retractions, rarely reported in pediatric cases, were prominent features.

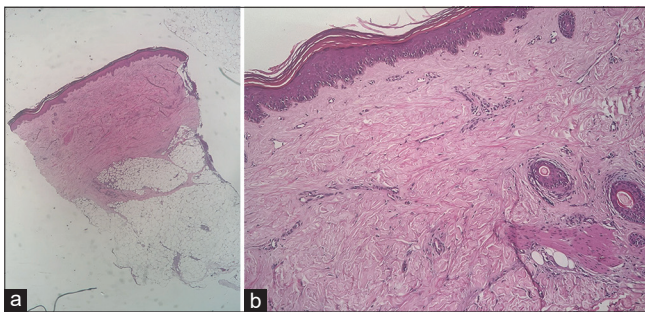
Diagnosing DPMC is particularly challenging in its early stages, as it mimics other localized scleroderma subtypes such as plaque-type morphea or linear morphea [7]. Definitive diagnosis is often delayed until complications such as ankylosis or muscle atrophy become evident. Extensive clinical and histopathological evaluations were crucial in differentiating this case from other connective tissue diseases.



Figure 1: (a-d) Clinical photos showing extensive sclerotic lesions with areas of hypo- and hyperpigmentation.

Table 1: Baseline laboratory findings in a patient diagnosed with disabling pansclerotic morphea of childhood

Category	Parameter	Results	Units	Reference Range
1. Inflammatory markers	C-reactive protein (CRP)	10	mg/L	<8
	Erythrocyte sedimentation rate (ESR)	12	mm/h	0–6
2. Immunity and autoimmunity	Antinuclear antibodies (ANA)	1:160 (speckled pattern)	-	<1:80
	Specific autoantibodies (SSA, SSB, etc.)	Negative	-	-
3. Eosinophilia	Eosinophils	11.8	%	<5
4. Coagulation and inflammation	Platelets (thrombocytosis)	593	×10 ⁹ /L	120–340
5. Muscle enzymes	Creatine kinase (CK)	141	IU/L	40–240
	Lactate dehydrogenase (LDH)	517	U/L	313–618

**Figure 2:** Clinical photo showing ulceration with irregular edges and erythematous inflammatory halos resting on a scleroatrophic base.**Figure 3:** (a) Histopathologic examination at low magnification (x10) showing a compact and thickened dermal layer with dense collagen bundles extending into the deeper layers of the skin. (b) Histopathologic examination showing thickened collagen bundles in the mid-dermis, without collagen homogenization, and a sparse perivascular lymphohistiocytic infiltrate (H&E stain; x100).

To date, only 39 cases of DPMC have been reported in the literature, with even fewer involving ulcerations or

squamous cell carcinoma (6.7% incidence), as noted by Wollina et al. [8].

Managing DPMC remains challenging due to limited evidence. In a cross-sectional study [9], methotrexate (MTX) has been identified as the second most commonly initiated treatment after systemic corticosteroids. MTX disrupts inflammatory cascades mediated by cytokines such as IL-1, IL-2, IL-4, IL-6, and TNF, which are implicated in the disease. It has been used in combination with corticosteroids (both intravenous and oral) with varying degrees of success.

Emerging therapies, including mycophenolate mofetil, biologics, tyrosine kinase inhibitors (TKIs), and Janus kinase inhibitors (JAKIs), have shown promise in conditions such as systemic sclerosis, supporting their trial use in DPMC. Alternative treatments, including sildenafil for ulcers, colchicine, cyclosporine, and tacrolimus, have shown benefits in individual cases.

Most children with DPMC require high-dose immunosuppressive therapy, with MTX and corticosteroids forming the cornerstone of care. The growing number of case reports and advances in biologics offer hope for improved treatments. Collaborative research and clinical studies are essential to optimize outcomes and standardize protocols.

CONCLUSION

DPMC is a rare and severe form of generalized morphea with significant diagnostic and therapeutic challenges. This case highlighted the disease's variability, including unique acral involvement, ulcerations, and tendon retractions. Early diagnosis, multidisciplinary care, and ongoing research are essential for improving outcomes.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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