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CASE REPORT

Optimal management strategies for paediatric morphea – insights from a case series

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ABSTRACT

Juvenile localized scleroderma, or morphea, is a rare chronic inflammatory disease primarily affecting the skin and subcutaneous tissue. Treatment options for paediatric cases are diverse, with a lack of standardised guidelines. This case series investigates the efficacy of systemic therapy combining methotrexate (MTX) and oral prednisone in children diagnosed with linear scleroderma. Seven paediatric patients diagnosed with linear scleroderma received systemic treatment consisting of MTX alongside oral prednisone. Clinical evaluation focused on monitoring disease progression and assessing improvements in skin manifestations. All patients demonstrated effective suppression of disease progression and gradual enhancement in skin condition following treatment initiation with MTX e and oral prednisone. This combined systemic therapy shows promising efficacy in halting disease advancement and improving skin manifestations in children with linear scleroderma. These findings underscore the potential benefit of this treatment approach in managing juvenile localised scleroderma; however, further research is necessary to establish comprehensive therapeutic guidelines.

KEY WORDS:

morphea, juvenile localised scleroderma, methotrexate, children, treatment.

INTRODUCTION

Juvenile localised scleroderma (morphea) refers to a number of different conditions characterised by skin thickening with increased collagen deposition. The prevalence of all types of morphea ranges from 0.4 to 2.7 *per* 100,000 people, and it is more common in Whites and females [1]. Several types of localised scleroderma are distinguished: plaque, generalised, bullous, linear, and deep morphea [2]. Linear scleroderma, with a prevalence of 65%, seems to be the most common type in children [3].

The spectrum of skin lesions in morphea is wide – from single plaques, through linear streaks of sclerosis, to serious cases such as facial hemiatrophy in Parry-Romberg syndrome [2]. The aetiology of the condition remains unclear, but different factors, such as trauma, vascular abnormalities, genetic factors, autoimmune processes, and infection, are taken into consideration [2]. Even though many treatment options are available, there is a lack of united guidelines regarding recommended therapies in children.

CASE REPORT

Seven children were hospitalised at the Dermatology Department because of linear morphea. All of them were

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FIGURE 1. Patient with morphea en coup de sabre. **A**) Clinical condition before treatment. **B**) Clinical condition after 6 months of treatment

girls aged 5–12 years. In 2 of them, lesions were located on the forehead, while others presented lesions mainly on their limbs. All were treated with a combination of methotrexate (MTX) and systemic corticosteroid – in 2 cases the corticosteroid therapy preceded MTX administration. In all cases, the doses of prednisone were gradually decreased after MTX administration and improvement of clinical condition (inactive stage of the disease); any side effects or increase in morphea activity were reported. In addition, all patients used topical agents on the affected area – in the first phase local glucocorticoids were administered, which were later substituted by tacrolimus. In each patient we achieved suppression of progression of disease activity and gradual improvement of skin condition (Figures 1, 2).

Each patient had regular controls of blood parameters and clinical condition. We observed improvement of skin lesions in all patients. The first effects were seen already on the first control appointments, which took place 1–2 months after the beginning of therapy. First reported effects were mainly ceasing the development and enlargement of the present lesions and lack of new ones. Later, the reduction of erythema, increased mobility, and reduced thickness of the skin appeared. We use the modified localised scleroderma severity index to monitor the effectiveness of the used therapy.

Only one patient reported side effects – stretch marks and nausea. When laboratory parameters are taken into consideration, we observed intermittently slightly increased levels of serum creatinine (up to 0.93, N 0.39–0.73 mg/dl) and total bilirubin (up to 1.55 mg/dl, N 0.20–1.00 mg/dl) in only one patient, but they were present before MTX administration and did not worsen during the therapy. Other patients presented no significant deviations in morphology, creatinine level, and liver function tests before and after introducing the treatment.

In one case MTX therapy was discontinued because of lack of compliance. Consequently, we observed a significant progression of disease activity and skin lesions. After reintroducing MTX to the therapy, the condition of the skin improved again. The therapy is being continued without any side effects or alterations in laboratory tests.

All information about our patients is presented in Table 1.

DISCUSSION

Morphea, or localised scleroderma, is a rare chronic inflammatory disease affecting the skin and subcutaneous tissue [4]. It manifests equally in adults and children, with incidence peaks typically occurring the ages of 7–11 years in children and 40–50 years in adults [5, 6]. Approximately 15% of cases occur in children under 10 years old [7]. Our case report series, consistent with literature findings, included only female patients, highlighting a female predominance in morphea [4].

The aetiopathogenesis of morphea remains unclear, with genetic predisposition, autoimmune dysregulation, and environmental factors implicated [4]. Linear localised scleroderma, the most common clinical type in the paediatric population, affects 51–65% of children with morphea, with peak morbidity observed around ages 7–8 years [3, 7]. Two of our patients exhibited forehead lesions, a manifestation of rare subtypes within linear scleroderma such as *en coup de sabre* and progressive facial hemiatrophy (PFH). *En coup de sabre* typically begins at the upper eyebrow ridge and extends to the scalp, potentially leading to cicatricial alopecia, while PFH involves severe atrophy affecting soft and osseous tissues, oral structures, and teeth, often accompanied by craniofacial deformities and neurological complications [6, 8]. Due to these potential complications, it is recommended that head magnetic res-

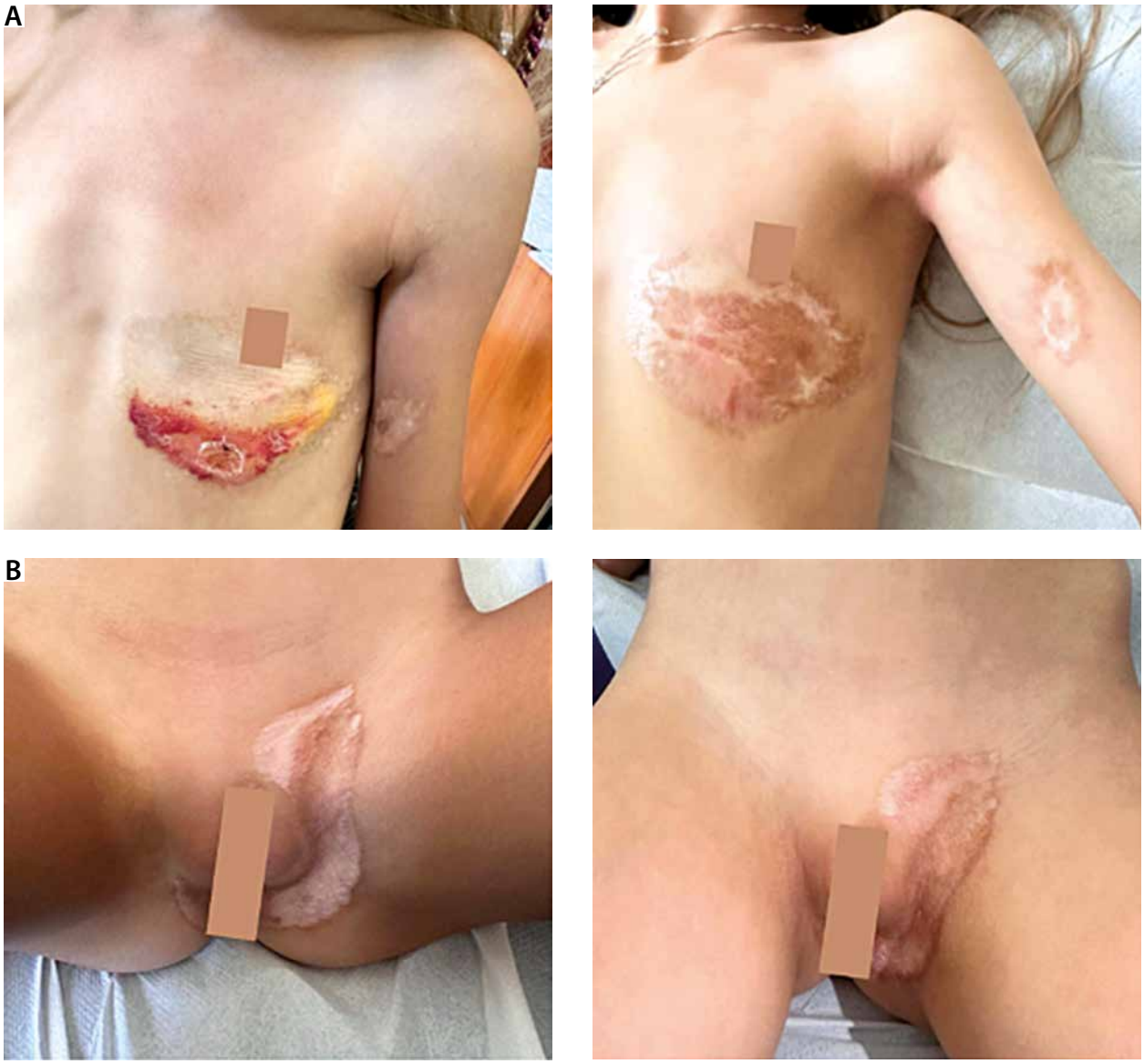

FIGURE 2. Patient with linear morphea. **A**) Clinical condition before treatment. **B**) Clinical condition after 2 months of treatment

onance imaging (MRI) and ophthalmological evaluations in affected patients be conducted.

In children, linear scleroderma tends to present a more severe clinical course compared to adults, often resulting in significant atrophy of skin, fat tissue, fascia, and muscle, leading to functional impairment and disability [9]. Systemic therapy introduced early during the active stage of juvenile linear scleroderma is crucial to halt disease progression, prevent new lesion formation, and improve existing ones [10]. Although various topical and systemic therapies are available (Table 2), including MTX monotherapy and combinations with glucocorticosteroids like methylprednisolone or prednisone, evidence supporting their efficacy in children remains limited [11]. Our case series employed the combination of MTX and oral prednisone, demonstrating satisfactory disease control and gradual regression of skin lesions [5, 6, 10, 12–14]. Notably, side effects were minimal, with only one patient experiencing stretch marks and nausea.

Assessment of treatment efficacy typically reveals reduction in skin sclerosis within 8–12 weeks post-therapy initiation [5]. Further research is needed to establish optimal treatment protocols tailored to the specific needs of paediatric patients with morphea.

CONCLUSIONS

Juvenile localised scleroderma represents a rare and inflammatory condition necessitating tailored therapeutic strategies. In cases where sclerotic lesions involve the face and head, conducting comprehensive evaluations such as head MRI and ophthalmological assessment is imperative to exclude associated underlying pathologies. Our study underscores the efficacy of combining MTX with oral prednisone as a viable treatment modality for linear localised scleroderma in paediatric patients. Nevertheless, there remains a critical need for further rigorous cross-sectional studies to generate evidence of high cer-

TABLE 1. Detailed characteristics of presented children with morphea

Parameters	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7
Age of disease onset	12	9	6	12	10	10,5	5
Age of MTX therapy onset	13	10	7	13	10	11	6
Dose of MTX *per* week (15 mg/m^2/week)	15 mg s.c.	15 mg p.o.	10 mg p.o.	15 mg s.c.	15 mg s.c.	10 mg s.c.	7.5 mg p.o., reduction to 2.5 mg p.o. and discontinuation; after disease relapse 15 mg s.c.
Duration of MTX treatment	5 months, treatment continued	1 year 5 months, treatment continued	MTX therapy was started at the last appointment	10 months, treatment continued	9 months, treatment continued	2 months, treatment continued	3.5 years of therapy, discontinuation for 2 months, reintroducing MTX and treatment for 7 months; treatment continued
Additional corticosteroid therapy (daily dose)	Prednisone 20 mg; after 3 months reduction to 10 mg	Prednisone 15 mg; after 3 months reduction to 10 mg	Prednisone 15 mg	Prednisone 30 mg; after 3 months reduction to 20 mg; after 1 month reduction to 10 mg; after 5 months reduction to 5 mg	Prednisone 20 mg; after 1 month reduction to 15 mg; after 4 months reduction to 10 mg; after 1 month reduction to 5 mg	Prednisone 15 mg	Prednisone 15 mg, after 4 months reduction to 10 mg After disease relapse: 20 mg, after 1 month reduction to 10 mg, after 6 months reduction to 5 mg every second day
Duration of corticosteroid therapy	6 months, treatment continued	1 year 5 months, treatment continued	1 month, treatment continued	10 months, treatment continued	9 months, treatment continued	2 months, treatment continued	3 years 9 months of therapy, discontinuation for 2 months, reintroducing prednisone and treatment for 7 months; treatment continued
Localization of skin lesions	Right thigh lateral	Forehead	Lower limb from ankle to groin, sclerosed skin lesions on left upper limb	Left thigh	Forehead	Left shoulder and left breast	Left thigh and groin
Reported side effects	None	None	None	Stretch marksnausea	None	None	None
mLoSSI before treatment							
Erythema	2	1	2	2	1	2	2
Skin thickness	2	2	2	2	1	2	2
New lesion/lesion extension	3	0	3	3	3	3	3
mLoSSI after 6 months of treatment							
Erythema	0	0	–	0	0	–	1
Skin thickness	1	1		0	1		1
New lesion/lesion extension	0	0		0	0		0
Family history	Psoriasis in grandfather	Suspicion of a skin sarcoidosis in mother	Negative	Negative	Negative	Negative	Negative

mLoSSI – modified localized scleroderma skin severity index, MTX – methotrexate

TABLE 2. The recommended dosage and duration of therapy with methotrexate, intravenous methylprednisolone, and oral prednisone in treatment of juvenile localised scleroderma

Name of the drug	Recommended dosage	Recommended duration of therapy	Authors	Possible side effects
Methotrexate	15 mg/m BSA/week (max 25 mg/week)	At least 12 months	Kreuter *et al.* [8]	Hair loss, headache, nausea, tiredness and liver damage [2]
	15 mg/m2/week MTX as single oral or subcutaneous dose	First 3 months with corticosteroids (preferably prednisone) – "bridge therapy", later more than 12 months after achieving clinical remission on medication.	Zulian *et al.* [3]	
	1 mg/kg/week s.c., (max 25 mg)	12 months	Li *et al.* [11]	
Intravenous methylprednisolone	30 mg/kg/day (max 1g/day)	On three consecutive days *per* month for at least 3–6 months	Kreuter *et al.* [8]	Weight gain and stretch marks [2]
	pulsed high-dose-30 mg/kg	Various administration schedules	Zulian *et al.* [3]	
	30 mg/kg/dose (max 1 g)	Schedule 1: 3 consecutive daily doses/month × 3 months Schedule 2: 1 dose/week × 12 weeks	Li *et al.* [11]	
Oral prednisone	0.5–2 mg/kg bodyweight/ day (max 60 mg/day), preferably in 2–3 daily doses	For 2–4 weeks, tapering the dosage thereafter	Kreuter *et al.* [8]	
	1–2 mg/kg/day	For a period of 2–3 months with subsequent gradual tapering	Zulian *et al.* [3]	
	2 mg/kg/day (max 60 mg)	Min 2 weeks, max 4 weeks with tapering schedule: week 8–50% initial dose (divided), week 16–25% initial dose (daily), week 24 12.5% initial dose (daily), week 48 – discontinuation of the drug	Li *et al.* [11]	

BSA – body surface area, MTX – methotrexate

tainty that can inform the development of standardised treatment guidelines for juvenile localised scleroderma.

DISCLOSURES

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