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A Potpourri of Atrophic Skin Diseases – A Case Series

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1. INTRODUCTION

The term 'scleroderma' is derived from Greek, wherein *skleros* means hard and *derma* means skin. Localized scleroderma encompasses a group of disorders characterized by inflammation, fibrosis, sclerosis and atrophy of skin and subcutaneous tissue. [1]Morphoea is a localized type of scleroderma characterized by progressive sclerosis of the skin. It is characterized by inflammatory 'bruise-like' indurated patches of skin involving the head, neck, trunk and extremities. [2]It has an annual incidence ranging from 4-27 new cases per million people. [3] It exhibits a bimodal age distribution with an onset between 2-14 years in children and 44-47 years in adults. [1][3] The disease also shows a female preponderance with a female-to-male ratio of 4:1.[3]

Morphoea is primarily confined to the skin and subcutaneous tissues such as fat, fascia, muscle, bone, and joints. Although predominantly a cutaneous disorder, it may also display extracutaneous manifestations leading to musculoskeletal, neurological, ocular, oral and dental complications. [1][3] Morphoea exhibits various clinical presentations and the most commonly used classification was described by Laxer and Zulian. [2] The subtypes of morphoea under this classification include linear, circumscribed, plaque-type, pansclerotic and mixed varieties. Circumscribed morphoea is the most common variant and is further categorized into superficial and deep forms. [2]

Lichen sclerosis et atrophicus (LSA) is also known as Csillag disease. It is a chronic inflammatory dermatosis characterized by porcelain-white, sclerotic plaques or patches. It has a world-wide prevalence of 0.03%. LSA can be differentiated into anogenital (83-98%) and extragenital variants (15-20%). [4] [5] Extragenital LSA commonly occurs over the neck, shoulder, and upper trunk. Other sites such as palms, soles, scalp, and face are less commonly implicated. It is usually asymptomatic, but may sometimes cause itching. [4]Extragenital LSA is more commonly seen in post-menopausal women with a female-to-male ratio that varies from 10:1 to 6:1.5. [6]

The co-existence of morphoea and LSA is thought to be controversial as some authors consider LSA to be a superficial variant of morphoea, while others consider it as a distinct entity. A possible relationship between the two with genetic, autoimmune and infectious factors has been proposed. [7] [8][9] In this research study, we present ten cases with varying clinical aspects of the localized scleroderma spectrum.

Case 1: Extragenital LSA with tinea versicolor-like presentation

A 58-year-old female presented to us with itchy hypopigmented lesions over the upper back, central chest, and both forearms for the past 7 months. On examination, multiple discrete, hypopigmented, atrophic macules and patches were noted over bilateral forearms, upper chest, and back. (Figure 1) Stretch skin test was negative for scales. Positive Koebner's phenomenon was noted within the lesions. Probable differential diagnosis included tinea versicolor, idiopathic guttate hypomelanosis, and confetti vitiligo. Histopathological examination of the biopsy specimen showed epidermal atrophy with basal cell vacuolar degeneration and dermis with homogenized dermal collagen and mild perivascular lymphocytic infiltrates, confirming the diagnosis of LSA. The patient was started on oral methotrexate 10 mg weekly once with concomitant narrowband-UVB phototherapy (NBUVB) and topical tacrolimus 0.1%.

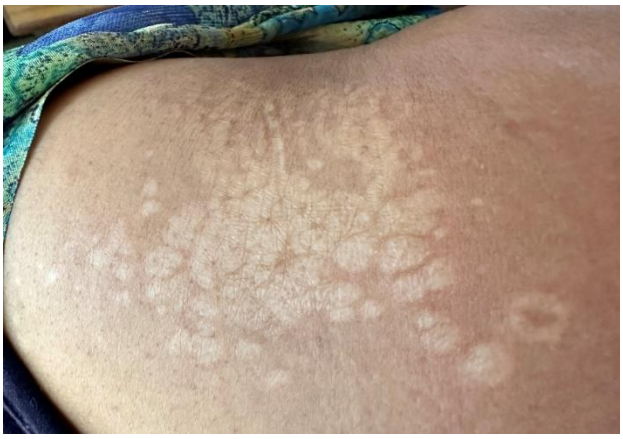

Figure 1: Multiple hypopigmented atrophic macules and patches over upper back

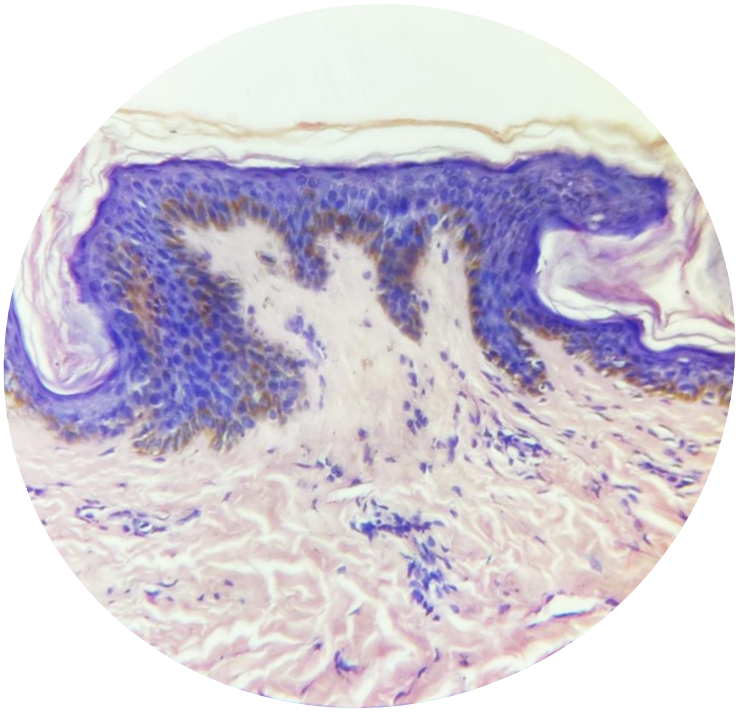

Figure 2: Papillary dermal oedema with homogenized collagen

Case 2: Linear morphoea mimicking linear atrophic lichen planus

An 18-year-old female presented with complaints of asymptomatic hyperpigmented lesions over the right lower limb for 4 months. Cutaneous examination revealed ill-defined, hyperpigmented, atrophic, indurated plaques distributed in a linear fashion over the anterolateral aspect of the right thigh extending to the lower leg. Histopathological evaluation showed a thinned-out epidermis along with thick eosinophilic collagen bands and scattered lymphocytic infiltrates in the dermis consistent with morphoea. The patient was subsequently treated with a weekly regimen of 15 mg methotrexate

subcutaneous injections along with 0.1% topical tacrolimus. We also administered intralesional triamcinolone acetonide 10 mg/ml injection 3 weeks apart over the indurated plaques.

Case 3: Extragenital segmental LSA imitating discoid lupus erythematosus and confetti vitiligo

A 75-year-old female presented with mildly pruritic skin lesions over the lower back for 3 months. On cutaneous examination, a single hypopigmented, atrophic plaque with an ill-defined hyperpigmented border, surrounded by multiple hypopigmented, atrophic macules, was noted over the right side of the lower back. (Figure 2) Discoid lupus erythematosus and confetti vitiligo were kept in mind as possible differentials. On histopathological examination, the epidermis showed mild thinning and basal vacuolar degeneration, and the papillary dermis revealed smooth eosinophilic collagen bundles with sparse perivascular infiltrates. We came to a conclusive diagnosis of extragenital segmental LSA and started the patient on methotrexate 10 mg per week and topical tacrolimus 0.1%.

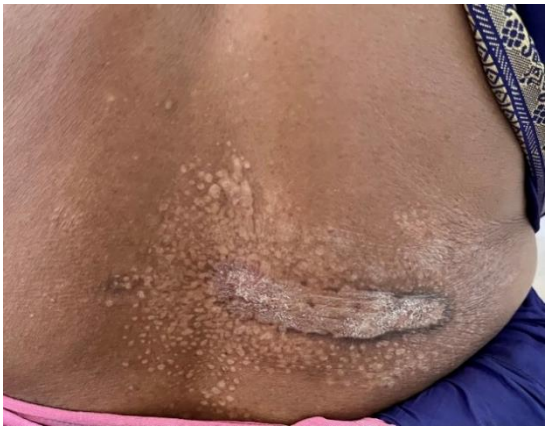

Figure 3: Single hypopigmented, atrophic plaque with an ill-defined hyperpigmented border, surrounded by multiple hypopigmented, atrophic macules present over right lower back

Case 4: Generalized morphea with discoid lupus erythematosus-like lesions over the malar areas

A 24-year old female presented with pruritic hyperpigmented skin lesions over the face for a duration of one year. She also complained of multiple similar lesions over the right forearm, left thigh, and abdomen. Cutaneous examination revealed symmetrical, hyperpigmented, and atrophic patches over the malar areas resembling discoid lupus erythematosus (DLE). (Figure 3) She also had diffuse, hyperpigmented, indurated plaques over the right upper limb, left thigh, and abdomen. Histopathological confirmation of morphea was made, and the patient was treated with a weekly dose of subcutaneous 15 mg methotrexate injections and a daily dose of prednisolone 30 mg, which was subsequently tapered over one month. NBUVB and topical tacrolimus 0.1% were coadministered along with systemic medications.

Figure 4: DLE like hyperpigmented and atrophic patches over bilateral malar area

Case 5: En Coup de Sabre with coexisting madarosis and scarring alopecia

A 29-year old female presented to the Dermatology OPD with complaints of asymptomatic lesions involving her face and

scalp for the past 5 months. A single well-defined, hyperpigmented atrophic patch was noted over the right forehead extending on to the scalp leading to scarring alopecia. Associated madarosis was also observed. Parry Romberg syndrome and linear lipoatrophy were considered as probable differential diagnoses. On histopathological examination, the epidermis showed atrophy and basal vacuolar degeneration at the dermoepidermal junction, and the dermis showed perivascular and periadnexal lymphocytic infiltrate with sclerosis and reduction in appendages consistent with en coup de sabre. She was treated with oral methotrexate 15 mg weekly once and 0.1% topical tacrolimus.

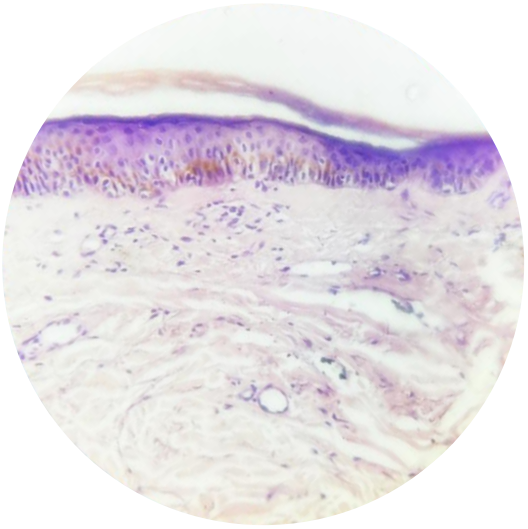

Figure 5: Epidermal atrophy with flattening of rete ridges

Case 6: Guttate extragenital LSA resembling tinea versicolor

A 6-year old girl was brought in with complaints of asymptomatic hypopigmented lesions confined to the chest for the past one year. (Figure 4) There was no family history of similar skin lesions. On examination, multiple confluent, hypopigmented to depigmented macules with mild wrinkling were noted over the chest. Other differential diagnosis considered were achromic tinea versicolor, dyschromatosis, and idiopathic guttate hypomelanosis. Histopathological examination showed orthokeratotic hyperkeratosis, focal follicular plugging, atrophic epidermis with flattening of the rete ridges, and focal basal cell vacuolization with the dermis showing thick hyalinized collagen bundles confirming LSA. The patient was subsequently treated with topical 1% methotrexate gel and 8% methoxypsoralen lotion.

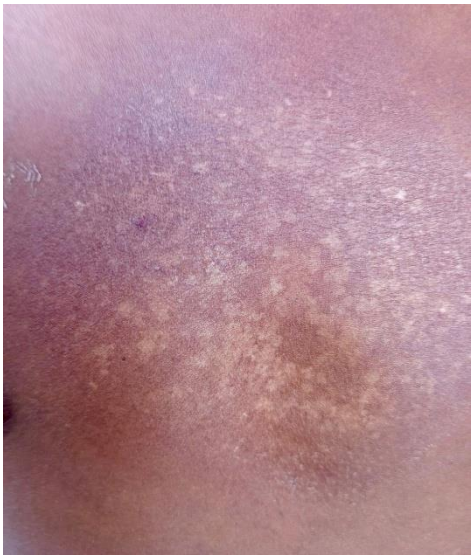

Figure 6: Multiple confluent, hypopigmented to depigmented macules with mild wrinkling over the chest.

Case 7: Atrophoderma of Pacini and Pierini

A 14-year old girl was brought in with complaints of asymptomatic lesions over the back for 7 years. On examination, multiple hyperpigmented atrophic patches along with a few non-indurated plaques were noted over the midline of the back and bilateral arms. (Figure 5) Superficial morphoea and atrophic lichen planus were considered as differential diagnosis.

Histopathological examination revealed mild spongiosis in the epidermis and sclerosed collagen bundles with mild perivascular mononuclear cell infiltrates in the dermis.The final diagnosis of atrophoderma of Pacini and Pierini was made, and she was treated with a weekly dose of oral 7.5 mg methotrexate, along with topical tacrolimus 0.1%, and calcitriol 0.0003% plus clobetasol proprionate 0.05% combination.

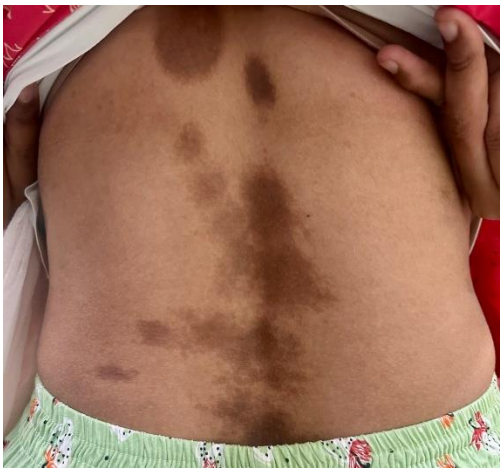

Figure 7: Multiple hyperpigmented atrophic patches and plaques noted over the midline of the back

Case 8: Linear LSA imitating segmental vitiligo

A 20-year old male presented to us with complaints of asymptomatic lesions over the back for 10 months. Cutaneous examination showed multiple hypopigmented, atrophic macules coalescing to form patches arranged in a linear pattern over the back. (Figure 6) Histopathological examination showed an atrophic epidermis with follicular plugging and focal basal cell vacuolation, and the papillary dermis showed areas of sclerosis with mild perivascular and periadnexal mixed infiltrates consistent with LSA. The patient was started on NBUVB and 0.1% topical tacrolimus.

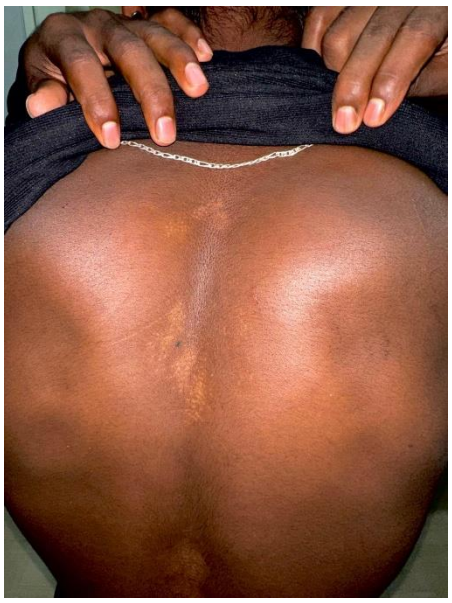

Figure 8: Linear arrangement of hypopigmented, atrophic macules and patches over the back.

Case 9: Localized early morphoea resembling DLE

A 14-year old girl was brought in with complaints of a single asymptomatic lesion over the face for 6 months. On examination, there was a single hyperpigmented, infiltrated plaque over the left side of her face. (Figure 7) Differential diagnoses included were DLE, atrophic lichen planus, and atrophoderma. Histopathological examination showed a thinned-out epithelial lining and a dense eosinophilic band of collagen in the papillary dermis with few scattered periadnexal lymphocytic infiltrates. Her diagnosis was finalized to be localized early morphoea and was treated with 0.05% topical desonide and 1% topical methotrexate.

Figure 9: Single hyperpigmented, infiltrated plaque over the left side of face.

Case 10: Generalized morphoea

A 46-year old female presented with complaints of asymptomatic lesions over bilateral upper limbs for 7 months. Cutaneous examination revealed multiple ill-defined, annular, hyperpigmented, atrophic plaques present over the extensor aspects of bilateral upper limbs. Atrophic lichen planus and generalized morphoea were considered as possible differential diagnoses. On histopathology, there was mild thinning of the epidermis and a band-like collagenous area in the dermis with mild-to-moderate perivascular infiltrates. Her diagnosis was finalized as morphoea, and was treated with oral methotrexate 15 mg weekly once and adjuvant NBUVB.

Table 1: Summary of all cases

	Age & Sex	**Clinical Examination**	**Histopathological features**	**Diagnosis**	**Treatment**
Case 1	58/F	Multiple hypopigmented, atrophic macules over bilateral forearms, upper chest and back.	Atrophic epidermis with basal cell vacuolar degeneration. Homogenous dermal collagen with mild perivascular lymphocytic infiltrates in superficial dermis	Tinea versicolor-like extragenital LSA	T. MTX 10 MG (W/O) Tacrolimus 0.1% ointment NB-UVB phototherapy
Case 2	18/F	Hyperpigmented, indurated, atrophic patches arranged in a linear fashion over the right leg	Thinned out epidermis along with a thick eosinophilic collagen band and scattered lymphocytic infiltrates in the dermis	Atrophic lichen planus-like morphoea	Inj. MTX 15 mg S/C (W/O) I/L Triamcinolone acetonide 10 mg/ml Tacrolimus 0.1% ointment

Case 3	75/F	A single hypopigmented, atrophic plaque and multiple guttate hypopigmented, atrophic macules over the lower back	Epidermis showed mild thinning and basal vacuolar degeneration. Papillary dermis showed smooth eosinophilic collagen bundles with sparse perivascular infiltrates.	DLE and confetti vitiligo like extragenital LSA	T. MTX 10 MG (W/O) Tacrolimus 0.1% ointment
Case 4	24/F	Hyperpigmented, atrophic patches over the malar areas and hyper-pigmented, indurated plaques over the right upper limb, left thigh and abdomen	Focal thinning of epidermis. Dermis showed densely distributed, thickened hypocellular collagen bundles with perivascular and periadnexal lymphocytic infiltrates.	DLE-like morphoea	Inj. MTX 15 mg S/C (W/O) T. Prednisolone 30 mg OD Tacrolimus 0.1% ointment NB-UVB phototherapy
Case 5	29/F	A single hyperpigmented atrophic patch along with scarring alopecia over the scalp.	The epidermis showed atrophy and vacuolar degeneration at the dermoepidermal junction and the dermis showed perivascular and periadnexal lymphocytic infiltrates with sclerosis and reduction in follicular units.	En coup de sabre	T. MTX 15 mg (W/O) Tacrolimus 0.1% ointment
Case 6	6/F	Multiple hypopigmented macules with mild wrinkling were noted over the chest.	Epidermis showed orthokeratotic hyperkeratosis, focal follicular plugging, reduced layering of the squamous epithelium and flattening of the rete ridges with focal basal cell vacuolization. Dermis showed thick hyalinized collagen bundles.	Guttate extragenital LSA resembling TV	Methotrexate 1% gel Methoxypsoralen lotion
Case 7	14/F	Multiple hyperpigmented patches with few non-indurated plaques over the back and bilateral arms.	Mild spongiosis in the epidermis and sclerosed collagen bundles with mild perivascular mononuclear cell infiltrates in the dermis.	Atrophoderma of Pacini and Pierini	T. Methotrexate 7.5 mg (W/O) Tacrolimus 0.1% ointment 0.0003% Calcitriol + 0.0.5% Clobetasol propionate combination ointment
Case 8	20/M	Multiple hypopigmented, atrophic macules coalescing to form patches over the back.	Epidermis showed atrophy, follicular plugging and focal basal cell vacuolation.The papillary dermis showed areas of sclerosis with mild perivascular and	Segmental vitiligo-like extragenital LSA	T. Methotrexate 7.5 mg (W/O) Tacrolimus 0.1% ointment

			periadnexal mixed inflammatory infiltrates.		
Case 9	14/F	A single hyperpigmented, infiltrated plaque over the left side of the face.	Thinned out epithelial lining and a dense eosinophilic band of collagen in the papillary dermis with few scattered periadnexal lymphocytic infiltrates.	Localized early morphoea resembling DLE	Desonide 0.05% gel Methotrexate 1% gel
Case 10	46/F	Multiple hyperpigmented, atrophic plaques over bilateral upper limbs	Mild thinning of the epidermis and a band-like collagenous area in the dermis with mild-to-moderate perivascular lymphocytic infiltrates.	Generalized morphoea	Inj. MTX 15 mg S/C (W/O) NB-UVB phototherapy

(MTX – Methotrexate; S/C – subcutaneous; I/L – intralesional; W/O – weekly once; Inj – Injection)

2. DISCUSSION

The present study included 10 patients between the ages of 6 years to 75 years with differing features of the localized scleroderma spectrum, including 4 cases of LSA and 6 cases of morphoea. Majority of the cases were females (9:1) in accordance with previous reports. There was a female predominance noted with the female-to-male ratio varying between 6:1 and 10:1 in cases of Csillag disease, and it is around 4:1 in cases of morphoea. [1][6][10]

Morphoea is known to have various different presentations, including linear, circumscribed, plaque-type, pansclerotic, and mixed variants. [2] Atrophoderma of Pacini-Pierini has also been considered as another rare variant of morphoea. [1] LSA commonly presents in the genital area, but it may affect other areas too. Postmenopausal females and premenarchal girls are commonly affected by LSA. Extragenital LSA is a rare disorder that mostly affects women. It affects around 15% of LSA patients. Around 6% of LSA patients may have only extragenital lesions. The co-occurrence of LSA and morphea suggests the possibility that both conditions may be part of a spectrum that share similar etiopathogenesis. [11]

Histopathological features indicating morphoea include squared-off punch biopsy specimens, heavy band-like sclerotic collagen bundles involving the dermis with destruction of appendages, and dense inflammatory infiltrates around the perivascular and periadnexal structures. Histopathological clues pointing towards LSA include an atrophic epidermis with hyperkeratosis, follicular plugging, basal layer vacuolar degeneration, and characteristic homogenized sclerosis of collagen bundles in the superficial dermis along with variably dense lymphoid infiltrates. [4] Dermoscopy of LSA shows porcelain white structureless areas, comedo-like openings, and a network of dotted blood vessels. [8]

A rare case of extragenital LSA- morphoea overlap masquerading as lupus vulgaris has been reported recently. Table 2 summarizes the atypical case reports of morphoea and LSA reported in past literature. Histopathological analysis is extremely important in such cases to confirm the diagnosis.

Table 2: Summary of previous case reports

Author	Age & Gender	Clinical Findings	Possible diagnosis	Actual diagnosis
Prasanna S et al, 2023[8]	56/M	Erythematous plaque with central atrophy with an advancing edge on one side and a regressive edge on other side	Lupus vulgaris	Morphoea LSA overlap
Go J et al, 2023[12]	63/F	Multiple erythematous to dusky patches on the	Patch-type mycosis fungoides	Early morphoea

		trunk and thighs.		
Mendiratta V et al, 2021[13]	5/F	Asymptomatic progressive hollowing of soft tissue over the posterior aspect of left ankle, calf, thigh, and buttocks.	Linear lipoatrophy	Subcutaneous morphoea
Kiran C et al, 2015[6]	45/F	Multiple itchy, hypopigmented plaques with hyperpigmented borders over both the legs since five years.	Discoid lupus erythematosus	Extragenital LSA
Patel B et al, 2015[5]	39/F	Multiple itchy, violaceous to hyperpigmented maculo-papular lesions over the lower limbs, thighs and buttocks since 5 months.	Lichen planus	Extragenital LSA
Ganesan L et al, 2015[4]	54/F	A single linear vertical hypopigmented plaque on the forehead since one year.	En Coup de Sabre	Extragenital LSA

3. CONCLUSION

Both morphoea and LSA mimic several dermatological conditions as depicted in this case series and can be misdiagnosed easily. This highlights the importance of thorough clinical evaluation and histological confirmation for establishing the precise diagnosis so that appropriate treatment can be directed to all patients

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