

## LINEAR SCLERODERMA: A SERIES OF ALL CLINICAL VARIANTS

Keerthi Jampani, Akshaya Nagaraja, Srilakshmi Peddireddy,  
Shiva Kumar V.

*Department of Dermatology, Venereology & Leprosy, P.E.S Institute of Medical  
sciences and Research, Kuppam, AP, India*

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Corresponding author: Dr. Keerthi Jampani, MD

[Keerthi.jampani88@gmail.com](mailto:Keerthi.jampani88@gmail.com)

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### Abstract

Morphea is a fibrosing disorder of the skin and subcutaneous tissues, wherein the overabundant collagen deposition destroys adnexal structures and hair follicles. In linear morphea erythematous or violaceous patches or plaques are seen with central sclerosis and active red border distributed in linear configuration which over time become sclerotic, white or hypopigmented. We report a series of cases encompassing all the three clinical variants of linear morphea. Case one had en coup de sabre deformity, case two had progressive facial hemiatrophy and case three had linear limb morphea with involvement of the face. This series is being reported for its rarity.

**Key words:** Morphea; En coup de sabre; Parry Romberg syndrome; Linear limb morphea

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### Introduction

Morphea, also known as localized scleroderma, is a fibrosing disorder of the skin and subcutaneous tissues, wherein the overabundant collagen deposition destroys adnexal structures and hair follicles [1]. The common clinical variants are circumscribed, generalized, bullous, linear and deep [2]. Plaque-type morphea is the most common type affecting adults in the mid-40s. The most common variant in children is linear morphea, with equal sex distribution, presenting as erythematous or violaceous patches or plaques with central sclerosis and active red border distributed in linear configuration which over time become sclerotic, white or hypopigmented.

### Case Report

#### Case 1

A 16-year-old male patient presented to our outpatient department with an asymptomatic, right-sided-facial atrophy and alopecia involving frontal region of scalp since 2 years (Fig. 1). Skin over the involved areas was smooth in texture and could be pinched up easily on palpation. There was no history of seizures, visual abnormalities or trauma. Examination showed a linear depressed groove on the frontoparietal region extending onto scalp, producing a linear zone of alopecia. This case was managed with topical corticosteroids.

#### Case 2

A 46-year-old female patient presented with a hyperpigmented depression over the left side of the face progressing upwards onto the scalp and downwards to involve the mandibular area since 20 years (Fig. 2). There was history of trauma to the head following which lesions started. History of three episodes of seizures was present since 1 year. She also had mild blurring of vision in the left eye. The patient was managed with intralesional corticosteroid injections.

#### Case 3

A 26-year-old female patient came with depressed groove over the left-side of the face, left arm and left forearm since 10 years (Fig. 3A, B). It initially started over the face and gradually progressed on to arms and forearms involving the elbow joint which restricted her joint mobility. The case was managed with topical calcipotriol and corticosteroids.

Skin biopsy was done in all 3 cases which showed histopathological features suggestive of morphea. Epidermis showed basket-weave keratosis with ironed out rete ridges and follicular keratosis (Fig. 4). Dermis showed thick collagen bundles paralleled to one another extending from papillary dermis to deeper dermis with lymphocyte infiltration around blood vessels and sweat ducts. Features suggestive of morphea.



Figure 1. En coup de sabre with a linear depressed groove on the frontoparietal region extending into the scalp, producing a linear zone of alopecia.



Figure 2. Parry Romberg syndrome with progressive facial hemiatrophy extending from left side of forehead upwards to the left side of the cheek downwards.



Figure 3A and B. Facial hemiatrophy with linear limb morphea.

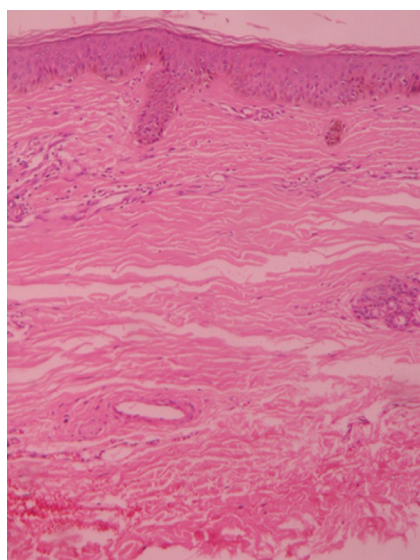


Figure 4. Epidermis showed basket-weave keratosis with ironed out rete ridges and follicular keratosis. Dermis showed thick collagen bundles paralleled to one another extending from papillary dermis to deeper dermis with lymphocyte infiltration around blood vessels and sweat ducts.

## Discussion

Linear morphea which is the most common type in children involves the lines of blaschko, suggesting genetic mosaicism. The three clinical variants are en coup de sabre (Fig. 1), progressive hemifacial atrophy (Fig. 2), and linear limb morphea (Fig. 3), all of which can be associated with underlying tissue atrophy. This case series represents all the varieties of linear morphea.

### En coup de sabre

The term en coup de sabre morphea refers to a lesion of linear morphea generally located in the frontoparietal scalp and/or the paramedian forehead, often resembling a stroke from a sword [3]. It usually occurs on the paramedian forehead. It can be associated with underlying ocular and central nervous system (CNS) involvement, including headaches and seizures. It typically follows Blaschko lines [4]. It can be associated with alopecia and present (less commonly) with more than 1 lesion.

### Progressive hemifacial atrophy (also known as Parry-Romberg syndrome)

It is an infrequent, acquired disorder characterized by progressive hemiatrophy of the skin and soft tissue of the face and, in some cases, results in atrophy of muscles, cartilage, and the underlying bony structures. It was first described by Parry in 1825 and Romberg in 1846 [5]. It can have overlap with en coup de sabre; thought to be different ends of the same condition. It may have underlying seizures [1].

### Linear limb morphea

It is associated with muscle atrophy, limb length discrepancies, and joint contractures. It is usually unilateral. Most likely to have extracutaneous manifestations with linear morphea. Linear morphea found to be associated with the following complications:

Articular disease (47.2%), neurologic (17.1%), vascular (9.3%), ocular (8.3%), gastrointestinal (6.2%), respiratory (2.6%), cardiac (1%), and renal (1%).

### Treatment

Various treatment modalities for linear morphea are tried like [6];

Topical corticosteroids: High-potency topical steroids can be applied once or twice daily to affected areas; lower-potency steroids should be used on the face or folds; care should be

taken to prevent atrophy of the uninvolved skin.

Topical vitamin D analogues: Calcipotriene ointment 0.005% under occlusion twice a day has been used, with good results.

Topical calcineurin inhibitors: Tacrolimus ointment (0.1%) is another effective topical treatment.

Intralesional corticosteroids: Triamcinolone, 5 mg/mL can be injected intralesionally once a month for 3 months.

Oral corticosteroids and Methotrexate given at 0.5 mg/kg weekly. It might be helpful to initiate a 1- to 2-month course of oral corticosteroids at the same time because of the slow onset of action for methotrexate.

Phototherapy: UVA and NB UVB is also tried with varied response.

## Conclusion

Because of the rarity of en coup de sabre morphea and parry-romberg syndrome, there is still much to be learned about them. Both en coup de sabre and Parry-Romberg syndrome are types of localised scleroderma that may have a similar pathogenesis. Neurological abnormalities, particularly seizures have been described in association with en coup de sabre and Parry-Romberg syndrome.

Antimalarials like hydroxychloroquine and methotrexate were found to be most effective of all.

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