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LINEAR MORPHEA- A RARE CASE SERIES

Dermatology

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ABSTRACT

INTRODUCTION:
Linear morphea is rare localized form of scleroderma. The lesions are characterized by localized hard/smooth ivory-coloured immobile areas of skin. The appearance is hidebound skin, known as andrew's disease of skin. These linear lesions often develop along the line of blaschko.

METHOD:
A cross-sectional,hospital-based clinical study of patients with linear morphea attending the Outpatient Department of tertiary care centre was done over a span of 3 months. Detailed history was taken along with clinical examination. After counselling and after recording their consent, skin biopsy done along with routine investigations and results were evaluated.

RESULTS:
We have come across six cases in 3 months. Age group ranged from 10-20 years with 2 male and 4 female patients. Average age of onset was 14.5 years. Two cases had lesions involving limbs, two had lesions on forehead and one patient had extensive lesions involving entire right half of body including face. Serum ANA titre was positive in one case,which also showed changes in CT scan and bone scan. EEG changes were seen in two out of three cases of facial lesions. Skin biopsy in all cases were consistent with morphea.

CONCLUSION:
Though linear morphea is a local disorder without systemic association generally,patients with Serum ANA positivity and EEG changes in facial lesions need to be observed for permanent CNS damage if not counteracted with immunosuppressives. As it has higher morbidity and predilection in younger population,cosmetic and functional disability caused by it affects the quality of life.

KEYWORDS

linear morphea, ANA, Line of Blaschko

INTRODUCTION

Linear lesions in dermatology are commonly encountered. They vary in cause; being congenital or acquired; can be present as macules, papules, patches, plaques, vesicles or nodules; they can be inflammatory or non-inflammatory; it can be a single lesion which is linear or multiple lesions which are arranged in a linear pattern. The causes for occurrence in a linear pattern include lesions following Blaschko's lines, blood vessels, lymphatics and dermatomes; due to Koebner's phenomenon and auto-inoculation; external factor such as plants, chemicals, allergens.

Linear Morphea is characterized by skin thickening with increased quantities of collagen in indurative lesion usually along Blaschko's lines.[1] The underlying etiology is poorly understood [2], although studies have suggested that morphea represents a mosaic form of systemic sclerosis limited to the skin. Linear morphea may have a self-limited course, but it frequently has a relapsing or chronic nature [3] despite aggressive medical treatments during the active phase of the disease. Delayed diagnosis and subsequent permanent cosmetic sequelae are common. 40-70% cases of Linear morphea are often observed in children and adolescents [4].

MATERIALS AND METHODS:

A total of 6 cases of different types of linear morphea which presented to our department were analysed. Detailed history including onset of lesion, its duration including signs and symptoms were taken in each case. All required routine investigations were done. In all cases, serum ANA titre was done. Additional investigations like CT SCAN, BONE SCAN were done in selected cases. Patients were followed up regularly for any complication related to linear morphea. Skin biopsy for histopathology was done and special stains were used wherever necessary.

RESULTS:

Out of 6 patients of LINEAR SCLERODERMA, 2 were male and 4 female. Youngest was 10 years old female and oldest was 27 years old female. Most common age group distribution was between 10 to 20 years of an age of which 2 was male and 2 was female (equal sex distribution). 2 patients had only limb involvement and 4 patients had scalp and limb involvement both. ANA positivity was present in 4 patients. EEG changes were observed in 2 patients with facial lesions.

CASE REPORTS:

Case 1: Linear Morphea Upper Limb

- **Clinical Presentation:** A 18 years old female patient presented with pain in affected limb and functional deformity with linear atrophic lesion from shoulder to thumb on right side and few discrete morphea patches on right thigh since 2 years, gradually increasing in extent and severity, leading to subcutaneous atrophy, change in girth of limb and hyperpigmentation.
- **Investigation:** S. ANA was ++ with normal ANA profile and normal haematological investigations.
- **Complication:** Patient is under regular follow up and has no evidence of any other sequelae related to the disease.

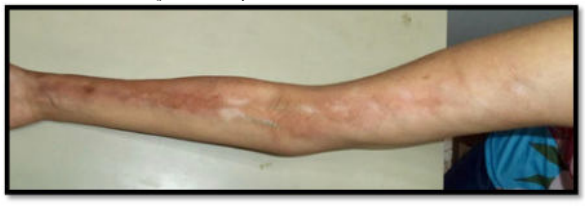

Figure1 : Linear morphea of upper limb

Case 2: Linear Morphea Lower Limb

- **Clinical Presentation:** A 14 years old male patient presented with linear atrophic lesion with evident cutaneous vasculature on right lower limb extending on trunk on same side since 10 months.
- **Investigation:** Haematological investigations were normal. S. ANA was negative.

Complication:

Limb shortening and decreased girth of the affected limb was evident.

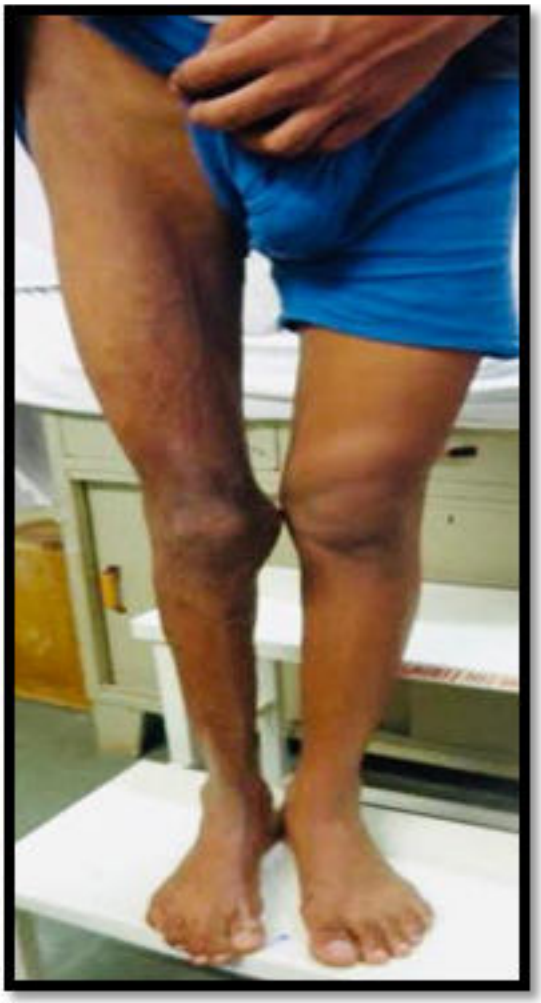

Figure2: Linear Morphea of Lower limb

Case 3: En Coup De Sabre

- **Clinical Presentation:** A 18 years old male presented with single linear indurated hyperpigmented lesion over forehead extending up to scalp with linear bony depression and there was NO history of convulsion.
- **Investigations:** CT SKULL denoted marked thinning of skin and bone and EEG demonstrated abnormal frequent sharp waves and occasional slow waves. S. ANA was +++.
- **Complications:** Patient was given topical therapy and there was no evidence of any further abnormality on regular follow up.

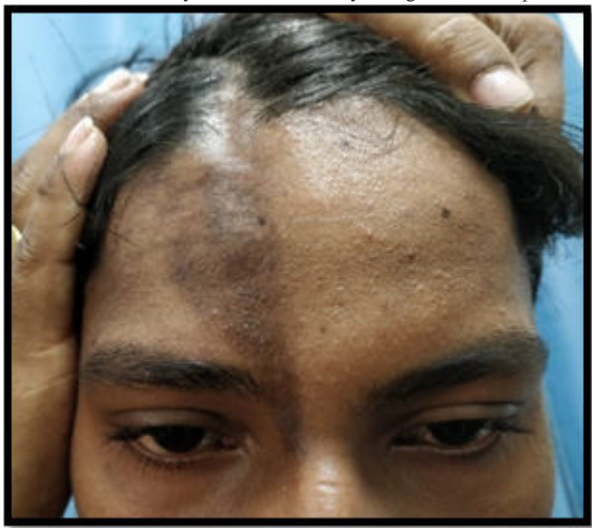

Figure 3: En Coup De Sabre

Case 4: Parry Romberg Syndrome

- **Clinical Presentation:** A 20 years old female presented with depression over right frontal and parietal region with patches of alopecia with facial hemiatrophy and muscle wasting of right upper limb without cutaneous lesions of morphea on other body parts.
- **Investigations:** Nerve conduction study and EMG was normal.
- **S.ana Was Positive Homogenous Pattern**
- CT Scan showed thinning of facial bones over right side leading to asymmetry, wasting of periorbital soft tissues and 3D BONE SCAN-RIGHT FACIAL HEMI-ATROPHY.
- **Complications:** There was no further progression of the disease and patient was referred to plastic surgeon for further management.

Figure4A and 4B : Parry Romberg Syndrome

CASE 5

- **Clinical Presentation:** A 27 years old female came to our OPD with Atrophic linear patch over left side of forehead.
- **Investigations:** S. ANA was positive ++ with nucleolar pattern.

CASE 6

- **Clinical Presentation:** A 10 years old female presented with linear morphea present over left paramedian side of forehead.
- **Investigation:** S. ANA was negative with normal hematological investigations.
- **Complication:** These lesions were static on a regular 6 monthly follow up with no systemic complaints, patients were treated with topical therapy.

DISCUSSION:

In our study, we inferred that out of total number of cases, there were 2 male and 4 females with M:F ratio=1:2. There is usually female predominance in occurrence of linear morphea[5].

The age group of our patients ranged from 10 -30 years, average age of onset being 14.5 years. 2 patients were in paediatric age group and 4 were in adult age group.

The site involved in our study was head and neck area in 3 cases, Upper limb in 1 case, Lower limb in 1 case, Hemi-atrophy of body in 1 case. Serum ANA was positive in 4 patients. Homogenous pattern was in 1 case and nucleolar pattern in 1 case. EEG Changes were seen in 2 patients with facial lesions, which guided us to regularly check for central nervous system related complications as neurological complications are most commonly associated with linear morphea.[6] Linear morphea can lead to significant morbidity and functional disability in young patients due to its complications. Early diagnosis and meticulous timely evaluation is needed to identify the complications and thereby plan its management.

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