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Case Report

A case report on late onset linear discoid lupus erythematosus on the face mimicking linear morphoea

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ABSTRACT

50-year-old female with no known comorbidities presented to the dermatology OPD with complaints of slowly progressive asymptomatic skin coloured linear atrophic plaque over left side of face since, 5 years. Cutaneous examination revealed well demarcated, linear atrophic plaque with hidebound skin over left side of face extending from scalp margin to tip of the nose, involving the lateral wall of nose along the lines of Blaschko. Dermoscopic examination showed structureless areas and loss of follicles as well as eccrine openings. Laboratory investigations including ANA-IFA were negative. A clinical diagnosis of linear morphoea (En- coup de sabre) was made. Further investigation with punch biopsy was performed from the lesion site which gave a histopathological diagnosis of Discoid Lupus Erythematosus and was confirmed with direct immunofluorescence study. Full physical examination, review of systems and laboratory workup showed no features to suggest systemic lupus. She was managed with hydroxychloroquine and sun protective measures. Linear discoid lupus erythematosus can clinically mimic linear morphoea, but histopathological examination provides distinctive features that aid in accurate diagnosis and differentiation between the two conditions. This case gave us valuable insight into keeping DLE as a differential for linear morphoea on the face**.**

Keywords: Direct immunofluorescence, Linear morphoea, Linear discoid lupus erythematosus

INTRODUCTION

Linear DLE is a rare subtype of Lupus erythematosus usually in children and young adults. Lesions are seen as linear unilateral erythematous plaques, usually distributed on the face along the lines of Blaschko.[1] Linear morphoea is a rare chronic autoimmune fibrosing disorder with multisystem involvement and is usually seen in children. Adult morphoea is of even rarer incidence.[2] Both these conditions can have similar clinical presentations and the definitive diagnosis is made by the characteristic histopathological and DIF findings. Sun protection remains the mainstay of treatment with concomitant usage of topical agents like corticosteroids, calcineurin inhibitors and retinoids in cases of linear DLE, whereas aggressive treatment with methotrexate or mycophenolate mofetil is indicated for cases of Linear morphoea due to its rapid progression and resultant complications.[3,4] We report a case highlighting the diagnostic challenges posed by Linear DLE's similarity to Linear Morphoea (En Coup de Sabre). Biopsy and histopathological examination proved crucial in establishing an accurate diagnosis, emphasizing their importance in distinguishing between clinically overlapping conditions.

CASE REPORT

50-year-old female with no known comorbidities presented to the dermatology OPD with complaints of slowly progressive skin coloured linear atrophic plaque over left side of face since, 5 years. No c/o itching/pain/photosensitivity. No h/o trauma prior to onset of skin lesions. The illness started as faint, reddish raised skin lesion over forehead which later progressed to involve

the left lateral wall of nose and nostril. No h/o similar complaints in family.

Physical examination

Cutaneous examination revealed well demarcated, linear atrophic plaque with hidebound skin over left side of face extending from scalp margin to tip of the nose, involving the lateral wall of nose (Figure 1A) along the lines of Blaschko (Figure 1B).

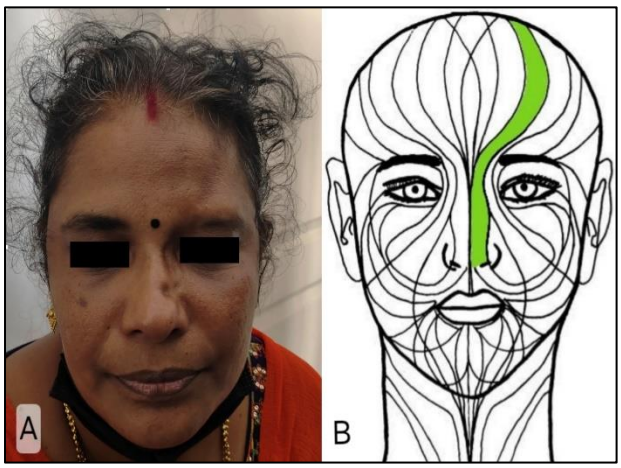

Figure 1: (A) Well demarcated, linear atrophic plaque with hidebound skin over left side of face extending from scalp margin to tip of the nose, involving the lateral wall of nose. Affected areas show loss of hair and atrophy. (B) Diagrammatic representation of Lines of Blaschko on the face and the pattern of distribution of lesions in our case (green).

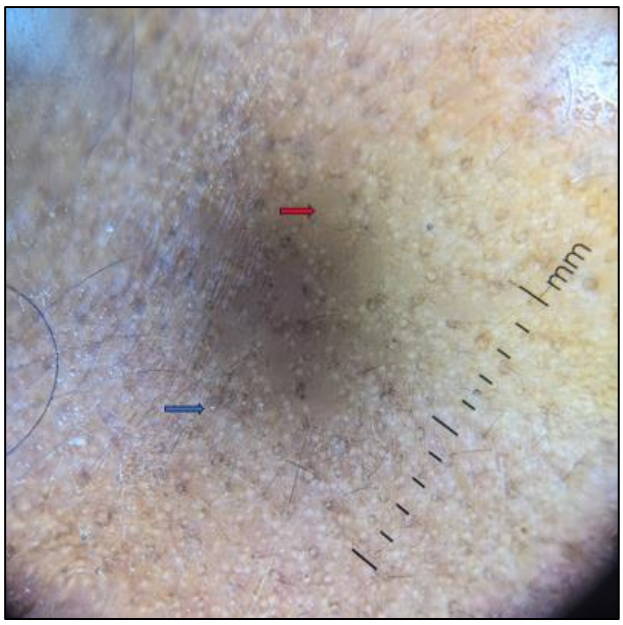

Figure 2: Dermascopic image showing peri eccrine brownish pigmentation (blue arrow) and brownish grey structureless areas with loss of follicular opening and eccrine opening (red arrow).

Loss of hair and atrophy present over affected sites. Systemic examination was within normal limits. Dermoscopic examination showed structureless areas and loss of follicles as well as eccrine openings (Figure 2). Hence, a clinical diagnosis of linear morphoea (En- coup de sabre) was made.

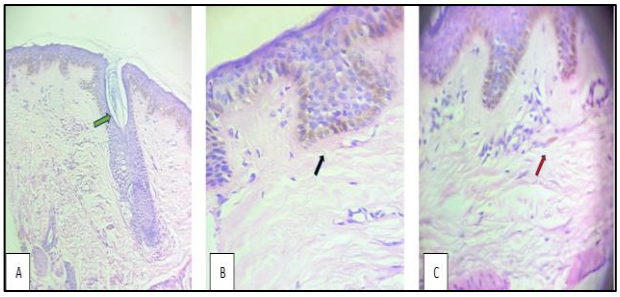

Figure 3: (A) Histopathological image in low power view showing hyperkeratosis, follicular plugging (green arrow), flattening of rete ridges and focal basal cell degeneration. (B) High power microscopic view showing thickening of basement membrane (black arrow). (C) Melanin pigment incontinence (red arrow) and perivascular lymphocytic infiltrate.

Investigations

Laboratory investigations including ANA-IFA (Anti-nuclear antibody-immunofluorescence assay) were negative. Histopathological examination with Punch biopsy performed from the lesion site showed basement membrane thickening, epidermal hyperkeratosis, perivascular and periadnexal infiltration of mononuclear cells which pointed towards a diagnosis of DLE (Figure 3). The diagnosis of DLE was confirmed with Direct Immunofluorescence study which was positive for IgG, IgM, IgA and C3 deposits at the dermoepidermal junction (Figure 4).

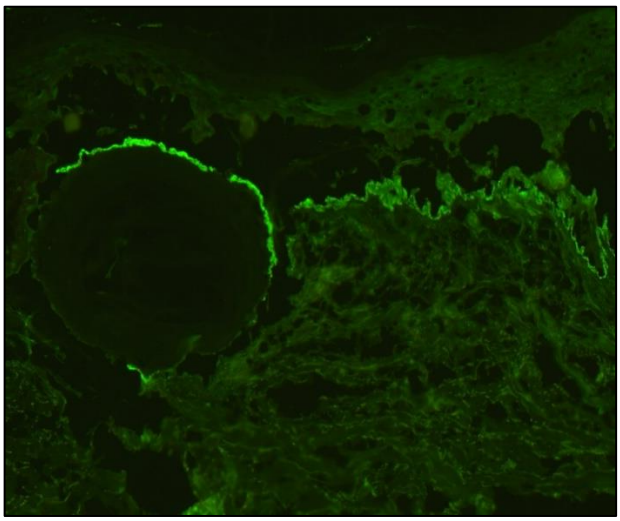

Figure 4: Direct Immunofluorescence study images showing granular staining of basement membrane zone for IgG, IgM, IgA and C3 deposits.

Diagnosis

Based on the clinical features and dermoscopic findings even though a clinical diagnosis of Linea morphoea was made initially, the final revised definitive diagnosis of Linear DLE was made based on histopathological and DIF findings.

Treatment and follow-up

Patient was treated with strict sun protective measures, topical tacrolimus (calcineurin inhibitor) and oral hydroxychloroquine 200 mg twice daily for 1month. Patient is currently on follow-up with no significant disease progression.

DISCUSSION

Linear DLE is a rare subtype of Lupus erythematosus with infrequent progression to systemic Lupus erythematosus. Linear DLE is mostly reported in children and young adults, frequently on the face.[1] Lesions are seen as linear unilateral erythematous plaques, usually distributed on the face along the lines of Blaschko which represents the embryological pathways of epidermal cell migration.[5] The diagnosis of linear DLE is made by histopathological examination which demonstrates characteristic findings such as follicular plugging, basement membrane thickening, dermal mucin deposition, epidermal hyperkeratosis, perivascular and periadnexal infiltration of mononuclear cells. DIF reveals deposits of IgG, IgM and IgA at the dermoepidermal junction.[6] Sun protection remains the mainstay of treatment with concomitant usage of topical agents like corticosteroids, calcineurin inhibitors and retinoids. Other treatment options include intralesional steroids, systemic antimalarials like hydroxycholoroquine, methotrexate, thalidomide and mycophenolate mofetil.[3]

Linear morphoea is a chronic autoimmune disorder characterised by linear atrophic depressed dyschromic cutaneous lesions usually seen in children. These are rare fibrosing disorder involving the muscle, connective tissue, bone and brain.[2] Linear morphoea is reported in adults in approximately 32%.[4] The histopathological findings that aid in diagnosis of linear morphoea includes thickened horizontally arranged dermal collagen bundles and skin sclerosis.[7] Aggressive treatment with methotrexate or mycophenolate mofetil is indicated in this condition due to its rapid progression and resultant complications like facial hemiatrophy.[4]

The presence of linear lesions in both these conditions are attributed to the exposure of genetically unique keratinocytes to UV light which produces an inappropriate cytokine response. Genetic mosaicism/epigenetic modification of keratinocytes and the immune system plays a pivotal role in specific arrangement of lesions in the characteristic linear pattern.[8] There are only few reported cases in literature of adult onset DLE mimicking linear morphoea. The lack of characteristic morphology of DLE lesion in the linear plaque complicates the diagnostic process. Sindhusen S et al, reported a case of adult onset linear DLE on forehead mimicking En Coup de Sabre on an adult female Thai patient. However erythematous to violaceous plaques were noted on clinical examination, and follicular plugging was noted on dermoscopy consistent with DLE.[9] This difference in clinical presentation could probably be attributed to ethnic variations.

The clinical presentation of Linear DLE can masquerade as linear morphoea (En Coup de Sabre), necessitating its inclusion in differential diagnoses for linear lesions of the face and head/neck. This case underscores the critical role of histopathological assessment via biopsy and role of DIF in distinguishing between conditions with overlapping clinical features.

CONCLUSION

Linear DLE can mimic Linear morphoea (En- coup de sabre) in clinical presentation and hence it should always be kept in the list of differential diagnosis for linear facial and head and neck lesions. This case underscores the pivotal role of biopsy and exemplifies the indispensable value of histopathological examination in establishing an accurate diagnosis, highlighting their essentiality in differentiating between clinically similar conditions with ambiguous clinical presentations.

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