Linear and Annular Lupus Panniculitis of the Scalp with Annular Lupus Panniculitis of the Face in a Child

Abstract

Linear and annular lupus panniculitis of the scalp is a rare form of lupus panniculitis recently reported in literature. It presents as linear or annular or arciform areas of nonscarring alopecia of the scalp with minimal surface changes. We report a 4-year-old Indian female child with arciform erythematous plaque over the forehead extending on the scalp with nonscarring alopecia with annular erythematous plaque over the cheek. Histology showed lobular lymphocytic panniculitis with abundant mucin deposition. Antinuclear antibody and systemic screen for lupus were negative. After treatment with oral corticosteroids, complete remission was achieved with good regrowth of scalp hair with no relapse during the follow-up period of 6 months. This is the youngest reported case of linear and annular lupus panniculitis of the scalp.

Keywords: *Alopecia, lupus erythematosus, lupus panniculitis*

Narayanan Baskaran, Prashant Bharti, Sudheer Arava[1], Sujay Khandpur

Departments of Dermatology and Venereology, [1]Pathology, All India Institute of Medical Sciences, New Delhi, Delhi, India

Introduction

Lupus panniculitis is a form of cutaneous lupus erythematosus, accounting for 1–3% of cases.[1] It presents as indurated erythematous nodules and plaques, with a predilection for face, arms, buttocks, and thighs. Scalp involvement is seen in 16.7% of the cases.[2] Linear and annular lupus panniculitis of the scalp (LALPS) is a distinctive form of lupus panniculitis affecting the scalp, presenting along the lines of Blaschko as linear or arciform areas of nonscarring alopecia.[3] Most cases of linear lupus panniculitis of the scalp reported in literature have isolated involvement of the scalp.[4]

Case Report

A 4-year-old female child presented with an asymptomatic linear curved erythematous plaque over the forehead extending on to the scalp with overlying alopecia and an annular erythematous plaque over the right side of face for 1 year. She also gave history of a similar asymptomatic erythematous annular plaque over the left cheek, which had resolved with depression a year back. There were no systemic symptoms or significant personal and family history.

Physical examination revealed an arciform, erythematous, nontender, and mildly indurated plaque extending from the left side of the forehead to the parieto-occipital region of the scalp with overlying nonscarring alopecia [Figure 1a]. Trichoscopy revealed intact follicular ostia with loss of follicles, miniaturized hair, and mild interfollicular erythema. There were no prominent or abnormal vascular patterns noticed in trichoscopy. An annular erythematous plaque with mild central depression was present near the right angle of the mouth above the jawline [Figure 2a].

Routine hematological and biochemical investigations were within normal limits. Skin biopsies from both the plaques showed superficial and deep dermal perivascular and perieccrine aggregates of lymphocytes, histiocytes, and occasional plasma cells [Figure 3a]. Lobular panniculitis with a dense infiltrate of lymphocytes and plasma cells was prominent [Figure 3b]. There was nodular moderately dense inflammation around the hair follicles, composed of lymphocytes and histiocytes without any follicular scarring in the biopsy from the scalp [Figure 3c]. Special stains revealed the presence of thickened basement membrane and abundant mucin in the subcutis [Figure 3d]. The connective tissue profile including anti-nuclear antibody, anti-ds DNA antibody and anti-Smith

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Address for correspondence:
Dr. Sujay Khandpur, Department of Dermatology and Venereology, All India Institute of Medical Sciences, New Delhi - 110 029, Delhi, India.
E-mail: sujay_khandpur@yahoo.com

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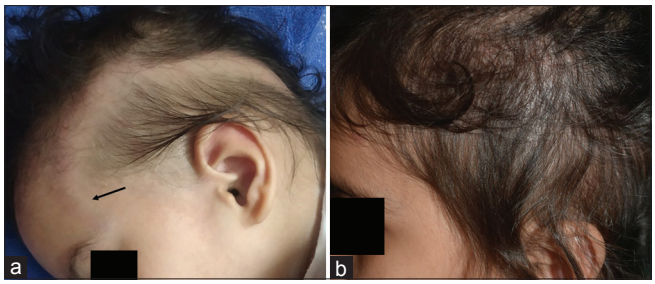

Figure 1: (a) Arciform erythematous plaque over the forehead (black arrow) extending on to the scalp with nonscarring alopecia. (b) Complete resolution of plaque with regrowth of scalp hair post-treatment

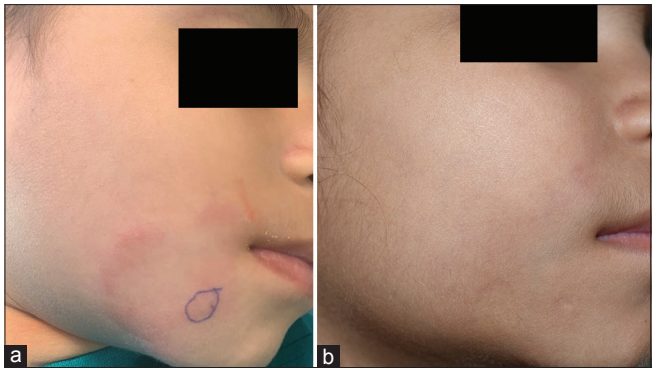

Figure 2: (a) Annular erythematous plaque over the right cheek. (b) Complete resolution of the plaque post-treatment

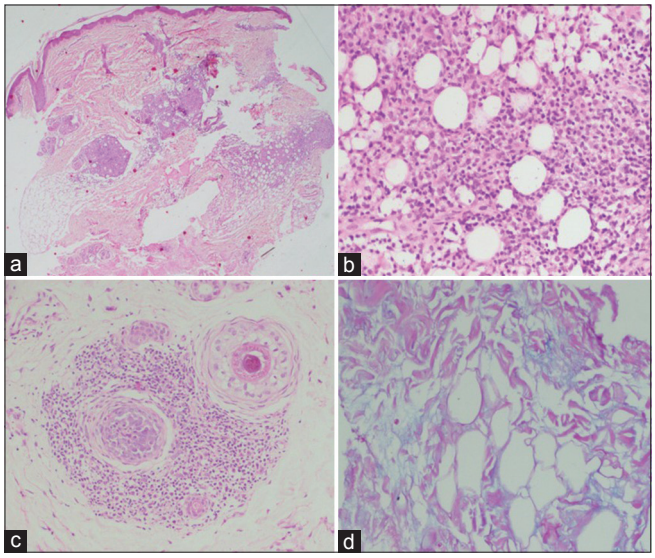

Figure 3: (a) Superficial and deep dermal perivascular and perieccrine infiltrate with lobular panniculitis (H&E, 40x). (b) Lobular panniculitis with dense infiltrate of lymphocytes, plasma cells, and histiocytes (H&E, 400x). (c) Perifollicular nodular infiltrates composed of lymphocytes and histiocytes (H&E, 100x). (d) Alcain blue stain showing abundant mucin in the subcutis (Alcain blue, 400x)

antibody were negative. The scalp was diagnosed with LALPS with annular lupus panniculitis of the face.

The child was started on oral prednisolone 1 mg/kg, which was tapered and stopped over a period of 8 weeks. There was complete resolution of the plaques and significant regrowth of hair overlying the plaque on the scalp with the therapy [Figures 1b and 2b]. Complete regrowth of hair occurred over five months. No recurrences were noticed during the follow-up period of 6 months.

Discussion

LALPS is a recently described distinctive form of lupus panniculitis. The first such case was described by Nagai *et al.*,[5] in 2003, and the entity was named by Mitxelena *et al.*,[6] in 2013. Subsequently, many such cases have been reported in literature, mainly affecting the East Asian race.[4]

LALPS has peculiarities that distinguish it from classical lupus panniculitis. It has a lower mean age of onset (21.5 years versus 35.6 years) and a male preponderance.[4] Importantly, the two features that distinguish it from classical lupus panniculitis are the linear or arciform presentation and the nonscarring nature of alopecia.[3,4]

Morphological presentations include annular, linear, and arc-shaped, and can affect any area of the scalp. Surface changes are generally absent, but erythema (70%) and induration (25%) can be seen.[3] Trichoscopy shows empty follicular ostia with loss of hair shafts, perifollicular whitish scaling, and patchy erythema.[3] Most cases reported have isolated scalp involvement, with only a few reports of associated annular plaques of lupus panniculitis elsewhere.[3] Park *et al.*, reported a case of LALPS with annular cutaneous nodules on left upper arm in geometric configuration.[7] Pandhi *et al.* reported a case of annular LALPS with concomitant involvement of the parotid area and the scalp, which further progressed to systemic lupus erythematosus.[8]

Histology shows lymphoplasmacytic lobular panniculitis. In comparison with the classical cases, LALPS has been shown to have more abundant mucin in the fat lobules and a higher degree of hyaline fat degeneration.[4] The antinuclear antibody positivity is lesser when compared to classical cases, and most cases do not progress to systemic lupus erythematosus.[3,4]

The differential diagnoses include linear alopecia areata, traumatic or tractional alopecia, and linear morphea. Clinical examination of the overlying surface changes, trichoscopic features, and histopathology are useful in distinguishing these entities. The location on face and scalp, annular and linear morphology of plaques rather than subcutaneous nodules, overlying erythema, and alopecia help to differentiate LALPS from subcutaneous granuloma annulare.

Treatment options include intralesional or systemic corticosteroids, hydroxychloroquine, dapsone, thalidomide and other immunosuppressants. Response to treatment is seen after 8 weeks, and there is good regrowth achieved with therapy. Around two-thirds of the cases go into

complete remission. Recurrence is seen in around 35% of the cases.[3,4]

In conclusion, LALPS is a unique form of lupus panniculitis presenting as linear or arciform nonscarring alopecia of the scalp. It can coexist with annular lupus panniculitis elsewhere. It should be included in the list of differentials of linear nonscarring hair loss, even in children. Screening for systemic lupus erythematosus and regular follow-up are necessary, even though such risk is low.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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