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Lupus panniculitis of the scalp presenting with linear alopecia along the lines of Blaschko

S. Lueangarun,[1] U. Subpayasarn,[1] P. Chakavittumrong,[1] T. Tempark[2] and W. Suthiwartnarueput[3]

[1]Division of Dermatology, Chulabhorn International College of Medicine, Thammasat University, Bangkok, Thailand; [2]Department of Pediatrics, Chulalongkorn University Faculty of Medicine, Bangkok, Thailand; and [3]Department of Pathology and Forensic Medicine, Thammasat University Faculty of Medicine, Bangkok, Thailand

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Lupus panniculitis of the scalp (LPS) is a rare presentation of lupus erythematosus panniculitis (LEP), an inflammatory disorder of the subcutaneous fat, mostly found in 1–3% of patients with lupus erythematosus[1] LPS has the distinctive clinical feature of distribution along the lines of Blaschko, giving rise to linear, arched or annular alopecia. We report successful treatment in a rare case of LPS with linear alopecia along the lines of Blaschko.

A 28-year-old Burmese man presented with a 1-year history of progressive linear-pattern hair loss, starting as a focal patch at the left parietal area, and gradually proceeding in length and width along the vertex to the right parietal area and eventually over both sides of the parietal area and vertex. There was no itching or burning on the alopecic area, and there were no other symptoms.

Physical examination showed a linear band of balding approximately 25 mm in width, distributed across the parietal and vertex areas, with the hairs being small and thin. A few scaly erythematous patches were observed at the vertex and right temporal scalp region without skin atrophy or sclerotic plaques (Fig. 1). Hair-pull test was negative, and the rest of the physical examination was unremarkable.

Histopathological examination of a biopsy from the scalp revealed a dense perifollicular lymphoid infiltration in the infudibuloisthmic portion of the hair follicle with a vacuolar interface to the infundibular epithelium, epidermal atrophy, follicular plugging, apoptotic keratinocytes, mild melanin incontinence, superficial and deep perivascular infiltrates, and necrosis of fat lobules. Alcian blue staining revealed interstitial mucin in the dermis and subcutaneous fat.

Laboratory examinations including complete blood count, blood urea nitrogen, creatinine and syphilis (VDRL) were normal, whereas a positive speckled pattern of antinuclear antibody (ANA) with a titre of 1 : 320 was noted.

Additionally, dermoscopic examination showed miniaturization of hairs and empty follicles in the alopecic patch, as well as the follicular openings with yellow dots, perifollicular white scale and some patchy erythematous areas. Perifollicular white dots associated with a honeycomb pigment pattern was seen in some alopecia areas.

Linear LPS was diagnosed on the basis of these results. Treatment was started with oral prednisolone 40 mg and hydroxychloroquine 400 mg daily, intralesional triamcinolone acetonide injection monthly, and 5% minoxidil lotion twice daily. Improvement was marked at the 2-week and 3-month follow-ups (Fig. 1), with disappearance of erythema and an increase in dermoscopic hair count from 34 to 124 per cm^2 at the biopsy area.

The distinct clinical features of LPS lesion morphologies are linear, annular, arch-shaped and ulcerated areas, involving all areas of the scalp (parietal, frontal, temporal and vertex), mainly along the lines of Blaschko.[1,2] The LEP pathogenesis pertaining to the Blaschko line distribution is not explicit, but it is hypothesized that there is cellular mosaicism resulting from a mutation during embryogenesis, producing abnormal cell clones that then migrate into deeper structural components such as lipocytes or fibroblasts.[3] These genetically abnormal cell clones may not induce clinical signs initially, but later appear to the immune system as new antigens, producing a local inflammatory response

Correspondence: Dr Suparuj Lueangarun, Division of Dermatology, Chulabhorn International College of Medicine, Thammasat University, 95 Moo 8 Paholyotin Road, Amphur Klongluang, Pathumthani Klongluang, Bangkok, 12120, Thailand
E-mail: saoraya180@gmail.com

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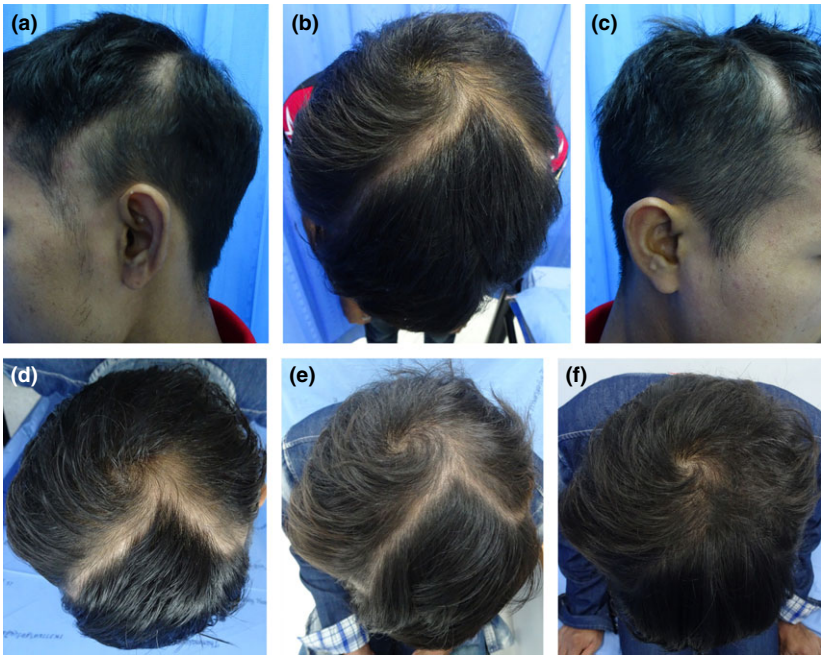

Figure 1 (a–c) Linear lupus panniculitis of the scalp with linear, nonscarring alopecia in linear and curved patterns along the along the lines of Blaschko. (d–f) clinical improvement at 1, 2 and 3 month(s) after treatment.

resulting from trigger stimulation and immune tolerance depletion due to local LEP inflammation.[4]

Combination therapies that have been reported in most of the published cases with deep LPS involvement have yielded complete response or some improvement, although having the possible drawbacks of scarring alopecia and skin atrophy, therefore we treated our patient with a combination of oral prednisolone, hydroxychloroquine, intralesional steroid injection and topical minoxidil lotion to decrease the disease-related inflammation and stimulate hair regrowth, resulting in significant improvement.

In conclusion, we report a rare and interesting case of LPS of the scalp with nonscarring linear alopecia along the lines of Blaschko. The combination treatment of oral prednisolone, hydroxychloroquine, intralesional steroid injection and topical minoxidil lotion yielded remarkable improvement. However, the patient will remain under continuous follow-up because of the ANA positivity, high possibility of recurrence, and possibility of developing systemic lupus erythematosus.[5]

References

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CPD questions

Learning objective

To demonstrate up-to-date knowledge about lupus panniculitis of the scalp.

Question 1

Which of the following is (are) the clinical presentation(s) of lupus panniculitis of the scalp (LPS)?

(a) Hair loss at the vertex and parietal areas.
(b) Sclerotic skin changes.
(c) Oddly shaped patches of hair loss with different hair lengths.
(d) Nonscarring alopecia with exclamation-mark hair.
(e) Linear alopecia along the lines of Blaschko.

Question 2

Which of the following is the presumptive pathology of a distribution along Blaschko lines of lupus panniculitis of the scalp (LPS)?

(a) Mutation during embryogenesis causes cellular mosaicism.
(b) Ultraviolet radiation.
(c) Inflammatory cell infiltration.
(d) Local trauma.
(e) Immune tolerance depletion results from local lupus erythematosus inflammation.

Instructions for answering questions

This learning activity is freely available online at http://www.wileyhealthlearning.com/ced

Users are encouraged to

- Read the article in print or online, paying particular attention to the learning points and any author conflict of interest disclosures
- Reflect on the article
- Register or login online at http://www.wileyhealthlearning.com/ced and answer the CPD questions
- Complete the required evaluation component of the activity

Once the test is passed, you will receive a certificate and the learning activity can be added to your RCP CPD diary as a self-certified entry.

This activity will be available for CPD credit for 2 years following its publication date. At that time, it will be reviewed and potentially updated and extended for an additional period.