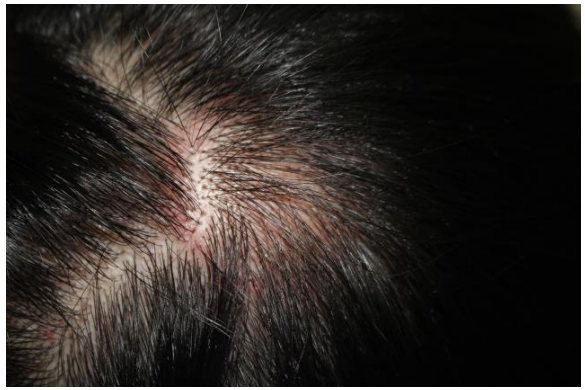
Case 8
A 19 year-old Thai woman from Bangkok
Chief complaint: Localized non-pruritic plaque on scalp for 6 months



Present illness:
The patient developed solitary well-defined erythematous pruritic annular edematous plaque concomitant with non-scarring alopecia on the lesion for 6 months. She was otherwise in good health.

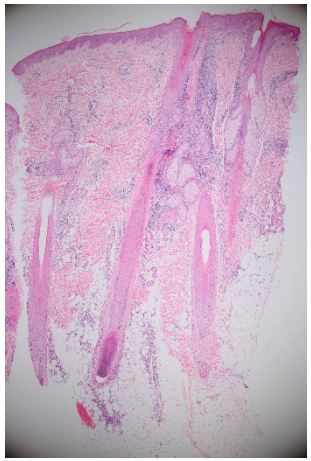
Past history: None

Family history: There was no family history of similar cutaneous lesions, autoimmune diseases or malignancies

Skin examination:
- Solitary well-defined scaly annular erythematous edematous plaque with non-scarring alopecia on the lesion
- Hair pull test was negative

Physical examination:
- Physical examination other than skin lesion revealed no abnormality.

Histopathology: (S16-8270A, scalp; vertical and horizontal section)



- Dense perivascular, perifollicular and interstitial inflammatory-cell infiltrate of mainly lymphocytes in the dermis, some of those extend into fat lobules of subcutaneous tissue.
- Interface changes with vacuolar alteration and occasional necrosis of basal keratinocytes.

Diagnosis: Annular lupus panniculitis of the scalp

Investigations:
Laboratory tests
- 10% KOH examination: negative
- CBC: Hct 38.9%, WBC 5970 (N68%, L25%, M6%, Eo1%),

Plt 271,000/mm^3, MCV 87.4 fL, MCH 28.8, RDW 12.0%

- BUN 9 mg/dL, Cr 0.78 mg/dL
- LFT: ALP/GGT 73/22, AST/ALT 22/21, TB/DB 0.7/0.3, TP/Alb 78.0/40.3
- UA: sp.gr. 1.016, protein and glucose: negative, WBC 0-1/HPF, RBC 3-5/HPF, Epi 0-1/HPF
- ANA, anti-dsDNA and anti-Sm were negative

Imaging studies

- Chest X-ray: unremarkable

Treatment:

- Hydroxychloroquine 200 mg/day
- 0.05% Clobetasol propionate ointment apply on scalp lesions twice daily
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Presenter: Phatcharawat Chirasuthat, MD
Consultant: Poonkiat Suchonwanit, MD

Discussion:

Lupus erythematosus panniculitis (LEP), also called lupus erythematosus profundus, is a rare variant of chronic cutaneous lupus erythematosus (CCLE). LEP was first described by Kaposi in 1883[1] and termed lupus erythematosus profundus when manifesting epidermal involvement by Irgang in 1940.[2] LEP affects approximately 2-3% of patients with systemic lupus erythematosus[3] (SLE) while the incidence of SLE in patients with LEP has been only about 10% to 41 %,[4-7] with the highest incidence seen in a Japanese[5] and Singaporean[7] series. LEP typically affects young women in their late 30s and early 40s. Like other forms of CCLE, there is female predominance with a female to male ratio of approximately 2:1.[8]

LEP often manifests as tender subcutaneous nodules and plaques with overlying normal skin, or forms skin lesions ranging from erythematous patch to discoid lupus erythematosus (DLE).[9] Lesions can be single or multiple, but generalized form is extremely rare.[4] Commonly involved sites are the face particularly cheeks, upper arms, scalp, hips and trunk.[4-7] Lesions tend to resolve with permanent atrophic scars and significant disfigurement. Untreated lesions of LEP can ulcerate, approximately 28% of the LEP patients.[10]

There is an uncommon clinical presentation of LEP localized at the scalp. The variant occurs much more common in Asian child and adolescent patients.The Lesion is usually non-scarring and sometimes located following the Blaschko's lines. It also tends to resolve without leaving atrophic areas.[11-12] Sometimes, LEP simulating alopecia areata presents as circumscribed non- scarring alopecia with probably containing exclamation mark hairs. Scalp tenderness, erythema of the overlying skin and histological findings of LEP are the key diagnosis of this variant of LEP.[13]

The diagnosis of LEP is confirmed by clinicopathologic correlation. More than 50% of cases, histopathology shows the form of typical changes of chronic DLE; including epidermal atrophy; interface change; thickened basement membrane; superficial and deep, perivascular and periadnexal lymphocytic inflammation; and increased dermal mucin. The remainder of cases, findings consist of a predominantly lobular lymphocytic panniculitis in the absence of typical epidermal and dermal changes of lupus.[14] Other features that could be useful clues for the diagnosis of LEP are lymphoid follicles with germinal center formation and hyaline sclerosis of lobules with focal extension into intralobular septa.[15]

Immunofluorescence studies typically demonstrate linear deposition of immunoglobulin M (IgM) and C3 along the dermoepidermal junction, even in the absence of DLE changes.[14] The immune deposition can be seen in the vessel wall located in the deep dermis and subcutis. Importantly, when a biopsy of a clinically typical lesion of LEP shows equivocal histopathology, a

positive direct immunofluorescence (DIF) stain along the dermoepidermal junction supports a diagnosis of LEP.[15-16] Antinuclear antibodies present in approximately 50% of patients with the disease.

The first therapeutic option of LEP is antimalarials, such as hydroxychloroquine and chloroquine. They are effective but require at least 3 months of treatment.[4] Systemic corticosteroids are often useful for severe cases accompanied by SLE. Other reported systemic therapies include thalidomide, dapsone, cyclosporine, intravenous immunoglobulins, and rituximab.[17]

Our patient presented with annular lupus panniculitis of the scalp which is an uncommon presentation of LEP. To the best of our knowledge, our case is the third reported case of annular LP localized to the scalp since the first case was reported by Bacanli et al [18] in 2005 and the second case was reported by Mitxelena et al [19] in 2013. Our patient did not meet the criteria for SLE. She was treated with hydroxychloroquine 200 mg/day and high potency topical steroids for lesions on the scalp. The lesions showed partial improvement after 4 weeks of treatment.

References

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