Case No. 18

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Patient: A 24-year-old Thai woman, from Bangkok

Chief complaint: Progressive hair loss on occipital area for 2 years

Present illness:
The 24-year-old woman presented with hair loss on occipital area for 2 years without previous severe illness and stress. She denied pulling her hair. She did not have any other skin or oral lesions. She had no photosensitivity, arthritis, or constitutional symptoms. In the absence of treatment, her symptoms became worsen. Therefore, she came to see the doctors at the Institute of Dermatology.

Past history: She had no illness or abnormality. She did not use any medication or supplement.

Family history: No family member experienced the same condition with the patient.

Physical examination:
   HEENT: no pale conjunctiva, no icteric sclera, no thyroid gland enlargement
   Heart: normal s1s2, no murmur
   Lungs: normal breath sound both lungs, no adventitious sound
   Abdomen: soft, not tender, no hepatosplenomegaly
   Extremities: no edema, no arthritis

Dermatological examination:
   Scalp: linear transverse band of non-scarring alopecia on the occiput involves about 10% of scalp area, hair pull test: positive
   Skin: no skin lesion
   Oral cavity: no oral ulcer
   Nails: normal nails

Investigation:
   CBC: Hb 13.1 g/dL, Hct 39.5%, WBC 6,000/mm³ (N 50%, L 39%, M 10%),
   Platelet 311,000/mm³
   FANA: negative
   LFT: AST 11 U/L, ALT 10 U/L, ALP 75 U/L
   Eye examination: no maculopathy

Histopathology: Slide No. 59-3345A, B
   Sections of vertical and horizontal cuts display unremarkable epidermis. The dermis shows a subtle superficial perivascular infiltration of lymphocytes. There is less inflammation of the subcutaneous fat lobules. No atypia is noted. No distinct vasculitis is observed. Hyaline necrosis of the fat cells is identified. The features are suggestive for lupus erythematosus panniculitis.

Diagnosis: Linear lupus erythematosus panniculitis of the scalp

Treatment: She had been treated with oral hydroxychloroquine, topical desoximetasone and topical minoxidil.
Discussion:

Lupus erythematosus panniculitis (LEP) is a rare variant of cutaneous lupus erythematosus (CLE) that characterized by inflammation of subcutaneous fat. LEP develops in 1% to 3% of patients with CLE. The common sites of LEP are areas with high density of adipose tissue which are face, upper arms, shoulders, breasts, buttocks and thighs. Lupus erythematosus panniculitis of the scalp (LPS) is a rare presentation of LEP. Most reported patients of LPS are Asian. Therefore, genetic or ethnic predisposition are suggestive.

Hallmark clinical presentation of LEP is non-scarring alopecia along the Blaschko lines on any area of the scalp. Shape of the lesions include linear, annular, or arc. Genetic mosaicism is proposed to play an important role on pathogenesis of linear distribution. Overlying skin can be normal or erythematous. Discoid lupus erythematosus (DLE) signs such as follicular plugging and scaling can also be found.

Characteristic histological finding in LEP is lobular panniculitis with lymphoplasmacytic infiltration. Majority of cases show hyaline necrosis and mucin deposition. Some cases show lymphocytic vasculitis and DLE of the overlying epidermis and dermis. Presence of DLE with LEP is diagnosed as lupus profundus.

In comparison with typical LEP, LPS shows relatively sparse inflammatory infiltrate, more abundant mucin in the fat lobules and a higher degree of hyalinized fat necrosis. Most cases of LPS patients have negative findings by immunofluorescent study. In contrast with LEP, positivity rate of antinuclear antibody (ANA) is low in LPS (17%).

Differential diagnosis of LPS include alopecia areata, trichotillomania, traction alopecia, linear morphea and subcutaneous panniculitis-like T-cell lymphoma (SPTCL). Clinical and histological finding, including laboratory investigation are essential for diagnosis.

Up to date, there is no standard recommendation for LPS treatment. Oral hydroxychloroquine, corticosteroids (topical, oral, and intralLesional) and oral dapsone are most commonly used with variable results. Treatment should be started in an inflammatory stage to prevent skin depression and alopecia.

Prognosis of LPS is varied from complete remission (67%), partial remission (33%) and recurrence (33%). To our knowledge, no LPS case that progressed to systemic lupus erythematosus (SLE) had been reported.

In conclusion, we present the case with rare manifestation of LEP. The patient presented with progressive linear transverse band of non-scarring alopecia. She had no systemic involvement. Histopathological of the lesion showed inflammation of the subcutaneous fat lobules and hyaline necrosis of the fat cells. ANA titer was negative. The diagnosis was LPS. Her alopecia was improved after treatment with oral hydroxychloroquine, topical steroid and topical minoxidil.

References: