Linear Lupus Erythematosus Profundus on the Scalp Following the Lines of Blaschko in an Adult

Chun-Ping Wu   Tsen-Fang Tsai

The lines of Blaschko describe distribution patterns which may represent embryologic development pathways. The distribution may be followed by some congenital and acquired skin disorders. Lupus erythematosus profundus (LEP) is a rare disease characterized by deep nodules and plaques, most commonly localized on the buttocks, arms, thighs and face. There is only one reported case of LEP on the scalp following the lines of Blaschko. We herein report the second case. The patient was a 21-year-old female presenting with a spiral-shaped alopecia on the scalp for one year. Skin biopsy taken from the lesion showed fat degeneration with mucin deposit and perieccrine infiltration of mononuclear cells. She was treated with oral hydroxychloroquine (200 mg/day) for 7 weeks. Complete regrowth of hair on the previous alopecic area was noted in the following one year. (Dermatol Sinica 22: 166-172, 2004)

Key words: Lupus erythematosus profundus, Linear cutaneous lupus erythematosus, Lines of Blaschko, Alopecia

布拉許口氏線是一種特殊分布的型態。此種型態是表現胚胎發育的路徑。有些先天以及後天的皮膚疾病是沿著這種型態分布。深部紅斑性狼瘡是一種少見的疾病，臨床表現特徵爲紅色或是膚色的深層結節和斑塊，特別好發在臀部、手臂、大腿和臉部。只有一個在頭皮沿著布拉許口氏線分布的線狀深部紅斑性狼瘡在文獻中被報告過。我們在此報告第二例。一位21歲女性，在頭皮呈現一年時間的螺旋狀禿髮。在病灶處的切片檢查顯示脂肪壞死合併黏液沉積以及汗腺旁的單核細胞浸潤。病人接受口服200 mg hydroxychloroquine治療七個星期。追蹤一年之後發現先前禿髮之處頭髮完全再生。（中華皮誌22: 166-172, 2004）
Fig. 1A
A spiral-shaped alopecia on the frontal and parietal areas.

Fig. 1B
Another arch-shaped alopecia on the right temporal area.

Fig. 2
(A) Photomicrograph showing perieccrine aggregation of plasma cells and lymphocytes in the dermo-subcutaneous junction(H & E, original magnification x 20).
(B) Higher magnification of fat lobules showing infiltration of plasma cells and lymphocytes(H & E, original magnification x 200).
INTRODUCTION

Lupus erythematosus profundus (LEP), also known as lupus erythematosus panniculitis (LEP), is characterized histologically by chronic periadnexal and perivascular inflammation deep in the dermis which extends into subcutaneous tissue, often with no underlying epidermal abnormality. Linear configurations of cutaneous lupus erythematosus profundus are rarely described. Only 4 cases of linear LEP (LLEP) had been reported in the English literature\(^1\)-\(^4\) and only one of them had a linear pattern alopecia on the scalp following the lines of Blaschko.\(^4\) We report the fifth case of LLEP and the second case of LLEP on the scalp following the lines of Blaschko in a young female.

CASE REPORT

A 21-year-old female presented with a 1-year history of progressive hair loss of the scalp in 2001. She had been in low mood and under stress. There was no history of trauma or excessive exposure to sunlight. She had no family history of connective tissue disease. Physical examination revealed a spiral-shaped alopecia on the frontal and parietal areas (Fig. 1A). There was another arch-shaped alopecia on the right temporal area (Fig. 1B). These two lines of alopecia were not continuous. The overlying skin showed normal color without atrophic or sclerotic change. Laboratory studies showed that the complete blood cell counts, anti-ENA (anti-SS-A, SS-B, Sm, Scl-70, and RNP), anti-nuclear antibody (ANA) were all within normal limits. A biopsy taken from the lesion on the scalp showed perieccrine aggregation of plasma cells and lymphocytes in the dermo-subcutaneous junction (Fig. 2A & B). The epidermis was normal with no liquefactive degeneration. There were no vacuolar alterations at the basement membrane zone. Fat degeneration was observed along with slight lymphocytic infiltration in the subcutaneous tissue. Special stains of alcian blue demonstrated abundant deposition of mucoid material through the dermis and the fat tissue (Fig. 3). Direct immunofluorescence (DIF) study was not performed. Since these clinical and laboratory findings do not fulfill the diagnosis of systemic lupus erythematosus (SLE). Linear LEP on the scalp was diagnosed mainly based on the histological findings. She was treated with hydroxychloroquine 200 mg/day for 7 weeks. Repeated biopsy for pathological examination and direct immunofluorescence (DIF) study was per-

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Fig. 3
Photomicrograph showing abundant deposition of a mucoid material through the dermis and the fat tissue (alcian blue, pH 2.5; original magnification x 40).

Fig. 4
Complete resolution of hair loss was noted in the following one year.

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formed one year later and no evidence of lupus erythematosus (LE) and alopecia was noted. Hence, complete resolution of hair loss was noted in the following one year (Fig. 4).

**DISCUSSION**

LEP is a rare variant of cutaneous LE involving the deep dermis and subcutaneous tissue. The lesions are preferentially localized on the face, buttocks, arms and thighs. The overlying skin can be unchanged or may show the lesions of LE. The histological features of LEP are lymphocytic panniculitis with hyaline fat necrosis, perivascular and periadnexal dense infiltrates of lymphocytes with some plasmal cells, hydropic degeneration of the basal layer in the epidermis, and fibrinoid degeneration of the vessels in the dermis. Approximately only 50% of patients with LEP have positive DIF stainings at the dermal-epidermal junction and the basement membrane of hair follicles, especially with granular depositions of IgM and C3 and linear fibrin deposits. Evidence of mucin deposition is also common in LEP.

Richarz et al. described a 48-year-old man diagnosed of cutaneous LE following the lines of Blaschko, but other authors questioned the veracity of this diagnosis. We did not include this case in the summary of patients with linear cutaneous lupus erythematosus (LCLE) (Table I). The mean age at onset in 16 cases including ours was 14.4 years. The female-to-male ratio was 1:1. ANA test was positive in only 4 patients. Eight cases occurred on the face, six on the extremities, three on the trunk, and two on the scalp. DIF studies were positive in nine cases. In all cases, no preceding trauma or excessive light exposure and no family history of LE were reported. In six patients, the lesions developed on two different areas. Four
cases of LEP with linear distribution have been reported and only one who had a linear lesion on the scalp. The patients' age of onset ranged from 3 to 37 years and in ten cases, the lesions developed in childhood. Twelve cases noted the relationship with Blaschko's lines. Therefore several important characteristics are noted in the linear variant of cutaneous LE: childhood onset, Blaschko line distribution, and low rate of progression to systemic disease.

The clinical diagnosis of LLEP is difficult especially when the other features of LE are absent. The histopathological and special stains of Alcian blue favored the diagnosis in this patient. The cause for linearity remained unknown and trauma or Kobner phenomenon had been suggested as possible causes.1-3 This variant of LEP is extremely rare and to our knowledge, only four cases had been reported.

The differential diagnosis of LLEP should include a variety of acquired linear dermatoses.11-13, 19 In our patient, the main differential diagnosis is linear scleroderma. However, Linear scleroderma is characterized by sclerotic change of the skin, homogenization of collagen bundles in the dermis without significant interface change. Alopecia areata has not been reported to occur in a linear pattern.

Topical corticosteroid1 and intralesional injection of steroid3 were used to treat LLEP in previous cases. Some cases of linear cutaneous lesions were improved considerably with oral dapsone4, 10, 11, 16 or anti-malarial drugs.1, 15 Our case was treated with oral hydroxychloroquine with nearly complete improvement.

In 1901, Blaschko first described acquired nevoid skin disease following constant lines.20 The nature of Blaschko lines remains unknown. One possible embryologic interpretation is that they correspond to the direction of growth in clones of cutaneous cells.20, 21 The cutaneous lesions that follow Blaschko lines reflect a mosaic condition deriving from postzygomatic somatic mutation or lionization.21 The Blaschko's line pattern is characteristic on the trunk and extremities.20 A description of Blaschko's lines on the head and neck was reported by Happle.22 In the preauricular area, the lines of Blaschko intersect at an angle of 90°. From the upper margin of the ear the lines run horizontal-

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age of onset (years)/Sex</th>
<th>Diagnosis</th>
<th>Location</th>
<th>ANA</th>
<th>DIF</th>
<th>Association with Blaschko lines</th>
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<tr>
<td>Umbert et al.7</td>
<td>7/female</td>
<td>DLE + morphea</td>
<td>Hand, forearm</td>
<td>—</td>
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<td>Tada et al.6</td>
<td>9/male</td>
<td>LLEP</td>
<td>Leg</td>
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<td>Bullous SLE</td>
<td>Hand, forearm</td>
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<td>LEP</td>
<td>Arm, breast</td>
<td>+</td>
<td>+</td>
<td>—</td>
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<tr>
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<td>3/female</td>
<td>LCLE</td>
<td>Face</td>
<td>+</td>
<td>—</td>
<td>+</td>
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<tr>
<td>Abe et al.9</td>
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<td>LCLE</td>
<td>Face</td>
<td>—</td>
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<td>Arm</td>
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<td>Face</td>
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<td>Face</td>
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<td>Face</td>
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<td>+</td>
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<td>Scalp</td>
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<td>Our case</td>
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<td>LLEP</td>
<td>Scalp</td>
<td>—</td>
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<td>+</td>
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ANA, antinuclear antibody; DIF, direct immunofluorescence study; DLE, discoid lupus erythematosus; LLEP, linear lupus erythematosus profundus; LCLE, linear cutaneous lupus erythematosus; SLE, systemic lupus erythematosus; ND, not done.
ly to the external canthus. In the preauricular area, they are crossed by lines that run downward in an almost perpendicular direction and subsequently form large caudally convex bends converging at the mouth. On the scalp, the lines show a spiral configuration that culminates on the vertex. The direction of the spiral apparently does not concur with the direction of the hair whorl. The number of pertinent cases, however, is very limited and therefore we cannot determine whether the direction of the spiral is constant. It is obvious that the lesions of our patient followed the lines of Blaschko (Fig. 5A & B).

In conclusion, we presented a rare case of LLEP, which is the fifth reported case of LLEP and the second reported case of LLEP following the lines of Blaschko on the scalp. In our cases, the clinical diagnosis of LEP was difficult because of the linear distribution of the lesions that did not show the typical findings of the diseases. The diagnosis of LLEP was established based on the histopathological findings. The good therapeutic response to oral hydroxychloroquine favors the diagnosis of LLEP lesions. However, because of the rarity of such cases, the lack of serological markers, and negative immunoglobulin deposit, the exact nosology of such cases remains to be clarified.

REFERENCES

20. Bologna JL, Orlow SJ, Glick SA: Lines of