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CASE REPORT

Two Cases of Linear Alopecia on the Occipital Scalp

Chin-Ho Rhee, M.D., Seong-Min Kim, M.D., Myung Hwa Kim, M.D.[1], Yong Woo Cinn, M.D.[1], Chull-Wan Ihm, M.D.

Department of Dermatology, Chonbuk National University Medical School, Jeonju, [1]Dankook University Hospital, Cheonan, Korea

Alopecia of a scalp shows various shapes and extents of hair loss, from a small round patch to polymorphous patches or total global alopecia. But alopecia of a linear shape is very rare. Only a few such cases have currently been reported in the medical literature. We recently had the chance to observe and treat two cases of linear alopecia that developed on the occipital scalp. The lesions themselves were like alopecia areata that shows a smooth bald area without any abnormality except the hair loss, but histopathologically, the lesions were compatible with lupus erythematosus profundus. **(Ann Dermatol 21(2) 159～163, 2009)**

-Keyword-
Linear alopecia

INTRODUCTION

Regardless of its causes, alopecia of a scalp shows a great variety of the shape and extent of the lesions from a small round patch to extensively involved irregular geographic patches or total global alopecia. Considering this clinical variety of alopecic lesions, there would be no reason that a linear shaped lesion should be impossible, although only a few of such cases have been reported in the medical literature.

In terms of a linear lesion in dermatology, quite a few dermatoses could show such a linear distribution on any part of the body. These dermatoses include linear scleroderma, linear lupus erythematosus, linear lichen planus, linear psoriasis, linear epidermal nevus etc[1]. All of the lesions of these dermatoses show visible cutaneous changes such as papules, scales, depression or elevation, hardening etc. When they occur on the scalp, they may extend to the adjoining smooth skin over the hair lines or they may involve other parts of the body simultaneously. Considering all of the above facts it was interesting that we experienced a long linear lesion that was limited only on a scalp without other cutaneous changes except for the loss of hairs with no involvement of any other parts of the body.

We recently observed two cases of linear alopecia on the scalp. The patients were virtually without any other skin changes. In the medical literature there are only two reports for the linear alopecia on the scalp[2,3].

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Reprint request to: Chull-Wan Ihm, M.D., Department of Dermatology, Chonbuk National University Hospital, 634-18, Geumam-dong, Deokjin-gu, Jeonju 561-712, Korea. Tel: 82-63-250-1976, Fax: 82-63-250-1970, E-mail: cwihm@chonbuk.ac.kr

CASE REPORT

Case 1

A-14-year-old boy presented a linear bald patch that traversed almost his entire occipital scalp at the level of his ears for the past 9 months. It was 14 cm long and 2 cm wide (Fig. 1A). The alopecic area was smooth and bald and the skin was normal in color and consistency without any other skin changes such as erythema, scales, atrophy or induration. The lesional skin itself was like that seen in alopecia areata. The hair pull test and the perilesional trichogram were normal. Besides the linear alopecia, the boy was otherwise healthy and he showed no other skin lesions, and there was no preceding trauma or illness. There was no family history of alopecia or connective tissue diseases.

The histopathological findings of the lesional skin were as follows: In the epidermis, hyperkeratosis and follicular keratotic plugs were seen. In the dermis, the most prominent findings were perifollicular lymphoid cell infiltrates and frequent catagen hair follicles (Fig. 1B, C). About two thirds of the hair follicles were in their catagen stage, but the catagen follicles still showed the transient part of the hair follicle, and around which there were dense lymphocytic infiltrates. Less frequently, telogen follicles with hair

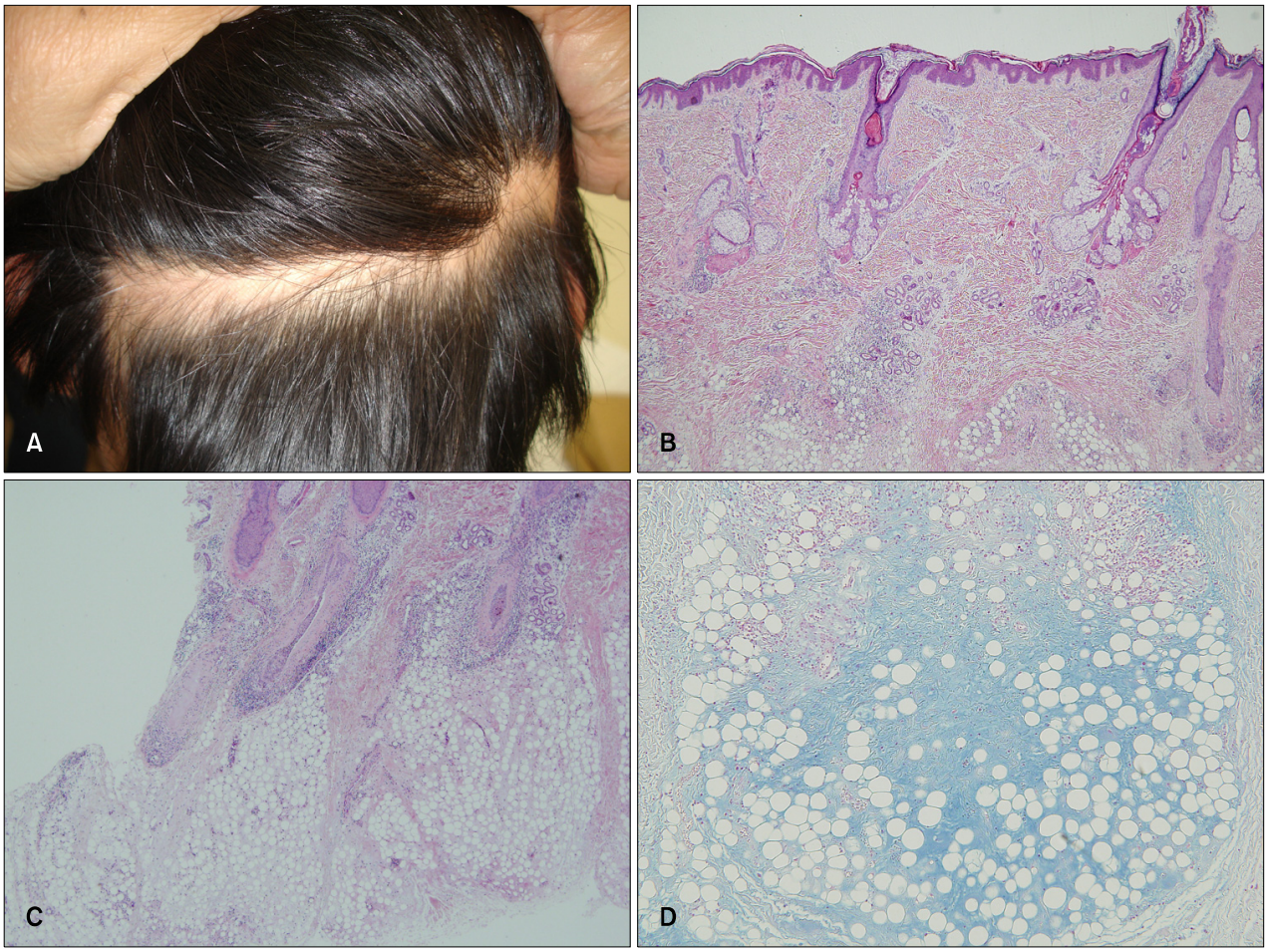

Fig. 1. The clinical and histopathological pictures of case 1. (A) A 2 cm wide and 14 cm long smooth linear bald band traversing the occipital scalp. (B) Microscopically, there are follicular plugs and hyperkeratosis in the epidermis. The upper segments of the hair follicles, the dermoepidermal junctions and the upper dermis show no appreciable inflammatory findings. Inflammatory infiltrates are seen in the area of the eccrine glands and at the bottom of the shrinking hair follicles (H&E, ×40). (C) In the lower dermis and fat tissue, prominent lymphoid cell infiltrates are surrounding the hair bulbs and the transient parts of the follicles. Fat tissue hyaline degeneration is also well seen (H&E, ×100). (D) The fat tissue degeneration was actually abundant mucin deposition (Alcian blue staining, pH 2.5, ×100).

germs and anagen follicles were found. Inflammatory cell infiltrates were also present in the area of the sweat glands. In the subcutaneous fat tissue, there was abundant mucin deposition within and between the fat lobules with some lymphoid cell infiltrates (Fig. 1C). Perineural inflammatory cell infiltrates were also found below the fat lobules. The routine lab tests were normal. Direct immunofluorescent testing of the tissue for IgM, IgG, IgA, C3 and fibrinogen was negative.

The treatment for the alopecic lesion was as follows: hydroxychloroquine 400 mg/day was administered for 5 weeks. After this, the hydroxychloroquine was switched to prednisolone 20 mg/day for 2 weeks because of facial hyperpigmentation that was thought to be due to hydroxychloroquine. During this period, intralesional injections of triamcinolone suspension (5 mg/ml) were done three times. Hair regrowth was noted from the 8th week of the treatment. The dose of the predinisolone was adjusted to 7.5 mg daily thereafter. Approximately 3 cm long terminal hairs grew at 5 months after the start of the treatment. Recurrence of the lesion was not seen at one year after the end of treatment. The diagnosis of the case was most compatible with lupus erythematous profundus.

Case 2

A 32-year-old presented a linear alopecia on his occipital scalp. It was about 2 cm wide and 12 cm long, and this was present for the previous two years (Fig. 2A). The lesion was remarkably similar to that of case 1 in its shape and location. The alopecic area was also smooth and it

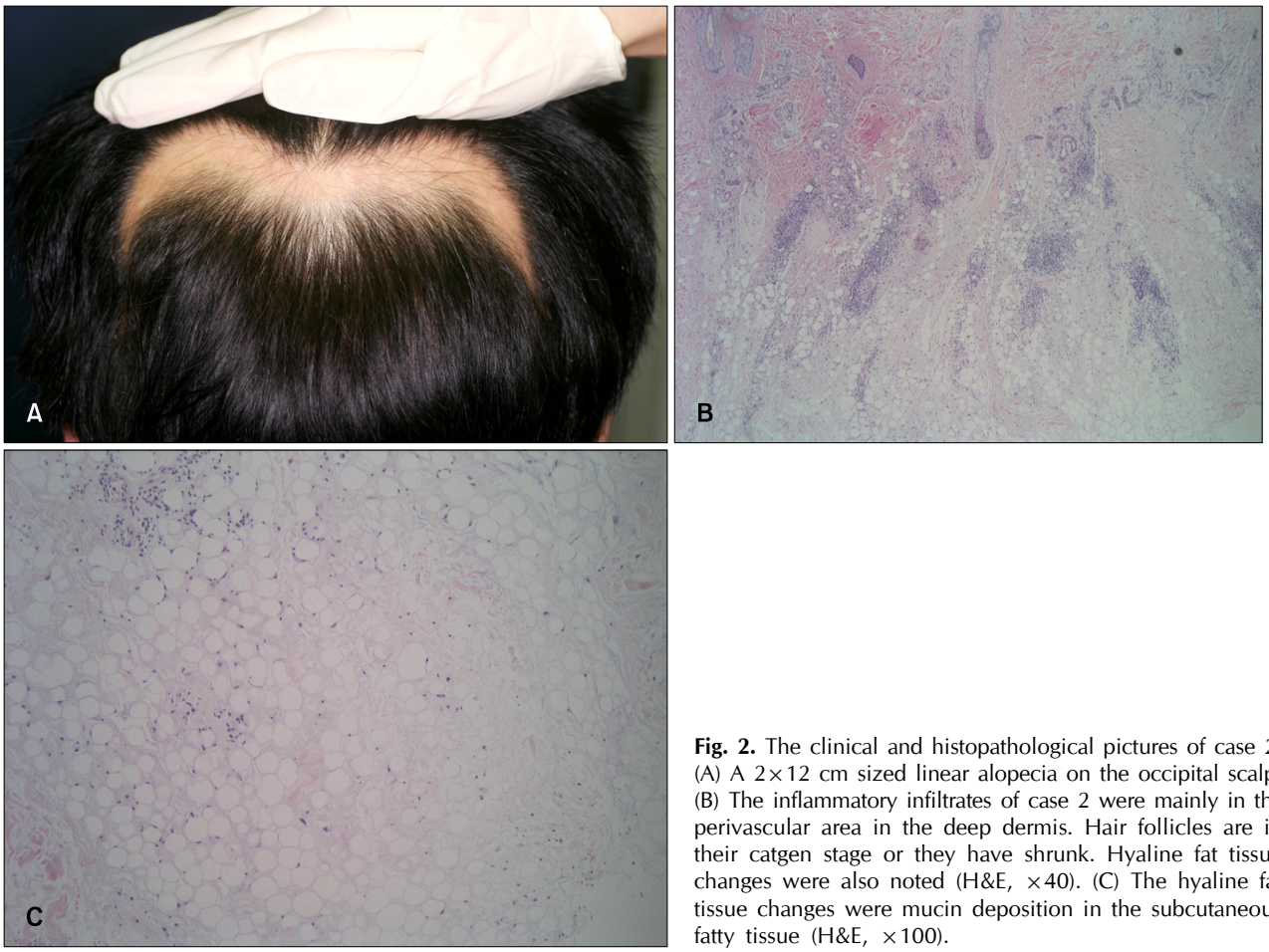

Fig. 2. The clinical and histopathological pictures of case 2. (A) A 2×12 cm sized linear alopecia on the occipital scalp. (B) The inflammatory infiltrates of case 2 were mainly in the perivascular area in the deep dermis. Hair follicles are in their catgen stage or they have shrunk. Hyaline fat tissue changes were also noted (H&E, ×40). (C) The hyaline fat tissue changes were mucin deposition in the subcutaneous fatty tissue (H&E, ×100).

showed only very slight erythema and a slight depression. The histopathological findings of the lesional skin showed deep dermal perivascular lymphoid infiltrates (Fig. 2B) and mucin deposition in the subcutaneous fat tissue (Fig. 2C). The hair follicles were generally small in size and they were frequently in the catagen stage, but there were no significant perifollicular infiltrates. Direct IF was negative for IgM, IgG, IgA, C3 and fibrinogen. The diagnosis of case 2 was most compatible with lupus erythematous profundus. For the treatment, dapsone (daily 50 mg) was administered for 12 weeks and regrowth of the terminal hairs then started. Further follow-up was not done.

DISCUSSION

A case of linear alopecia due to lupus erythematosus profundus was reported in 2007 in the Korean medical literature[3]. The patient, a 20-year-old, showed linear alopecia on his occipital scalp. At first it was diagnosed as alopecia areata by the clinical findings, but one year later the lesion showed erythema, scale and atrophy. A subsequent biopsy revealed the findings of lupus erythematosus profundus showing basal liquefaction, mucin deposition in the dermis and subcutis with periappendageal inflammatory infiltrates. Before this Korean report, a Japanese girl with linear alopecia was reported on with the title of "Linear lupus erythematosus profundus on the scalp following the lines of Blaschko" (2003)[2]. The lesion of the Japanese girl consisted of two disrupted linear alopecia areas on the left temporal and the parietal scalp, but they also involved the forehead skin as band-like erythema. Histopathologically, it also showed abundant deposition of mucoid material and atrophic hair roots with slight lymphocytic infiltrates. Both the Korean and Japanese cases failed to regrow hairs at four months and at one and a half years, respectively.

The clinical lesions of the present two cases and the first Korean case mentioned above are all similar to each other. The skin lesion of case 1 was like the lesional skin of alopecia areata because it showed only a smooth bald surface without any other skin changes. Case 2 also showed only slight erythema and a depression, which could be seen in the lesion of alopecia areata. We think it

is understandable that the previous Korean case was at first misdiagnosed as alopecia areata before it developed erythema and scales one year later.

The diagnoses of the two cases are most compatible with lupus erythematosus profundus based on their histopathological features. Case 1 showed lymphocytic infiltrations not only in the perifollicular areas, but also in the eccrine sweat gland areas. In the subcutaneous fatty tissue, there was prominent mucin deposition and hyalinization of the adipose lobules. Although the lymphoid cell infiltrates in the adipose tissue were not so severe, it is known that the intensity of the inflammatory infiltrates in adipose tissue may lessen over time as the hyalinization progresses[4]. Perineural lymphocytic infiltrates were also seen in case 1. In case 2 the periappendageal infiltrates were not prominent, but the deep dermis showed prominent patchy lymphocytic infiltrates. The adipose tissue findings were similar to those of case 1. In addition to the histopathological findings, their clinical responses to the drugs (chloroquine, predinisolone and dapsone) also favored of the diagnosis of lupus erythematosus profundus.

In 2002, Kossard reported a case of lupus panniculitis that clinically simulated alopecia areata of a common patchy type[5]. The case was a 27 year-old woman with a known history of lupus erythematosus. When the two small patches of alopecia developed on her scalp, 1 cm and 2 cm in diameter, respectively, there were also erythematous plaques on her face. With a prompt diagnosis and administering proper treatment with hydroxychloroquine, both the facial LE lesion and the alopecia of the scalp resolved well. The author described that the alopecic lesion showed only faint erythema and it rather simulated the lesion of alopecia areata, but the biopsy specimen of the lesion revealed a prominent lymphoid infiltrate targeting the deep follicular segments, the hair bulbs and the eccrine glands with hyalinization of the fat and mucinosis. The author mentioned that the prominent lymphoid infiltrate targeting the deep follicular segments and hair bulbs shared the histopathology of alopecia areata, and it induced localized patches of non-scarring hair-loss that clinically resembled alopecia areata. We observed similar findings in our two cases. Anyway, it was very interesting that such histopathological changes developed linearly and they traversed the occipital scalp of the patients. For the clinical aspect of the linear alopecia, such a sharply defined linear shape could be possible in a patient with trichotillomania, but this was definitely impossible according to the characteristic histopathology of the present cases.

Another interesting aspect of the present case report was that there are authors who have argued that there are true combinations of alopecia areata and lupus erythematous, and these two maladies are both known to be caused by autoimmune mechanisms[6]. They showed that alopecia areata developed in 10% of patients with lupus erythematosus in contrast to alopecia areata developing in to 0.42% of the general dermatologic patients. Among the two cases presented here, the microscopical slide of case 1 was reviewed by D. Whiting of the Baylor Hair Research and Treatment Center in Dallas. He suggested a possible combination of the two autoimmune diseases, alopecia areata and lupus profundus, based on the numerous catagen follicles with several terminal follicular hair bulbs surrounded by lymphocytes and the lymphohistiocytic infiltrates that invaded the eccrine glands and subcutaneous adipose tissue. The exact pathogenesis of the two autoimmune diseases is currently unknown. Regardless of the potential combination of the alopecia areata and lupus erythematosus, lupus panniculitis may be clinically misinterpreted as alopecia areata when the process is concentrated around the deep temporary segments of the hair follicles and the panniculus produces temporary hair loss. In case 1, we found that the smooth alopecic lesion showed an inflammatory process mostly in deep segments of the hair follicles and the eccrine glands with mucin deposition in the fat tissue. In case 2, we found that the inflammatory infiltrates were mostly in the deep dermal perivascular and eccrine gland areas with mucin deposition in the fat tissue. The hair follicles were generally miniaturized or in the catagen stage, and they showed no prominent infiltrates. We think that the relatively sparse inflammatory process of the upper segment of the hair follicles, where stem cells are present, and of the upper dermis may explain the successful hair regrowth in the two cases. In lupus panniculitis, if the process extends into the dermis or up to the epidermis, then it would produce erythema, telangiectasis, ulceration or scarring that leads the typical clinical lesion of alopecia due to lupus erythematosus. In such cases, it would be likely that the alopecia may remain as scarring.

Approximately 70% of the patients with LE profundus also have typical DLE lesions that often overly the panniculitis lesions. The typical subcutaneous lesions present as firm nodules 1 to 3 cm in diameter. The overlying skin often becomes attached to the subcutaneous nodules and it is drawn inward to produce deep, saucer-shaped depressions. In the absence of overlying DLE, the concurrent facial involvement can simulate the appearance of lipoatrophy, and if seen on the breast, this can simulate breast carcinoma. Roughly 50% of the patients with LE profundus have evidence of SLE[7].

Several cases of linear lupus profundus have been cur-

rently reported, but most of them were on the extremities and less frequently, on the face[8-11]. In the field of dermatology, it is known that many naevoid skin lesions and dermatoses display an arrangement following the Blaschko lines[12]. On the occipital scalp, the transverse line of the present cases may be compatible with the lines of Blaschko, but any explanation for the reason why LE profundus develops along these lines is beyond the scope of this paper. However, we think it is fascinating that all the four cases of such linear alopecia due to LE profundus, including the present two cases, have been found only among the Korean and Japanese people.

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