Case 8

A 28-year-old Thai woman from Bangkok

Chief complaint: An asymptomatic rash on forehead for 3 months


Fig.8.1

Present illness: The patient developed an asymptomatic linear erythematous to purplish atrophic patch on the forehead for 3 months. She had no history of trauma preceding the lesions or photosensitivity. She had no personal or family history of autoimmune connective tissue disease.

Past history: She had no underlying disease.

Current medication: None

Physical examination: Other systemic examination revealed no abnormality

Dermatological examination:

- Linear erythematous to purplish atrophic patch on the forehead following the lines of Blaschko (Fig.8.1)
- Other dermatological examinations were unremarkable including mucous membranes, scalp, hair and nails

Dermatoscopy:

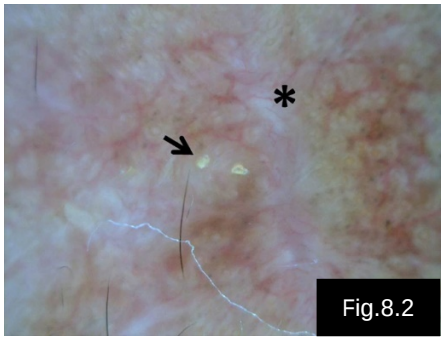
Fig.8.2

- Structureless whitish areas (asterisk), follicular keratotic plugs (arrow), telangiectatic vessels and brown to grayish pigmentation (original magnification x 50) (Fig.8.2)

Histopathology (S18-7140, forehead):

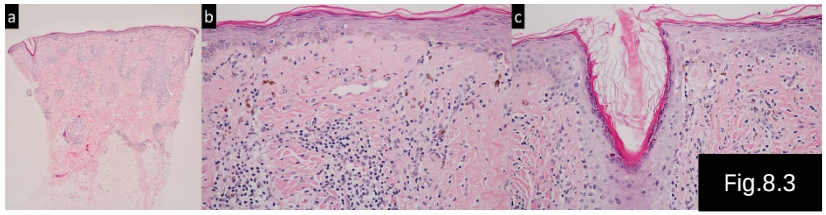

Fig.8.3

- Superficial and deep perivascular and periadnexal lymphocytic infiltrate (a) (original magnification x 40)
- Epidermal atrophy, hydropic degeneration of the basal cell layer, numerous melanophages, and telangiectasia (b) (original magnification x 400)
- Follicular interface change with follicular hyperkeratosis (c) (original magnification x 400) (Fig.8.3)

Direct immunofluorescence (S18-28787, forehead):

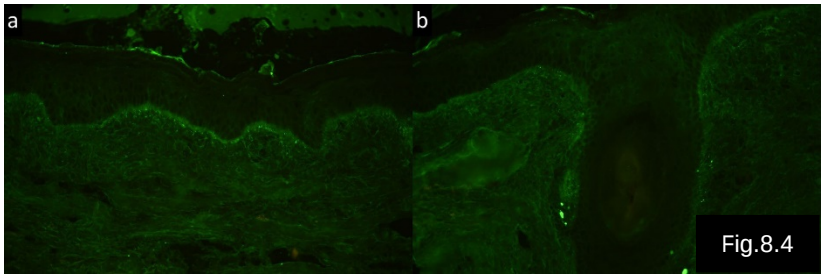

Fig.8.4

- Homogenous granular deposition of IgM and few cytoid bodies along the basement membrane (a) and follicular epithelium (b) (Fig.8.4)

Investigation:

- Antinuclear antibody titer was negative

Diagnosis: Linear discoid lupus erythematosus (LDLE)

Treatment:

- Strict sun protection
- Tacrolimus ointment 0.1% was applied on the lesion
- hydroxychloroquine 200 mg/day

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Discussion:

Cutaneous LE presenting with linear configuration is exceeding rare. A linear variant of CLE was first described in 1978 by Umbert and Winklemann[1] as a cutaneous mixed or overlap syndrome between linear scleroderma and DLE. Later in 1998, Abe et al[2] proposed the term linear cutaneous lupus erythematosus (LCLE) to described this rare variant of CLE.

According to the literature review, a total of 102 cases diagnosed with LCLE has been reported. The most common pathological diagnosis of CLE in the linear distribution was discoid lupus erythematosus (DLE) (38.2%), followed by lupus erythematosus panniculitis (LEP) (21.6%), lupus erythematosus tumid (LET) (2.9%), subacute lupus erythematosus (SCLE) (2%) and bullous lesions of SLE (1%)[3-12]. The majority of cases were children and young adults. The mean age onset was 22.1 years (1 - 68 years). Unlike the female predominance typically seen in SLE patients, no sex preference was noted. The female to male ratio was 1.3:1. The typical clinical manifestations were single or multiple linear asymptomatic erythematous plaques along the lines of Blaschko. Most commonly, lesions developed on head (66.7%). However, the neck, trunk, and extremities may also be affected. Antinuclear antibodies are mostly negative. Photosensitivity and progression to SLE are rarely observed[6, 8, 9].

The exact pathogenesis of LCLE is unknown. Alfred Blaschko first described the lines of Blaschko in 1901 and represent the developmental growth pattern of the embryonic ectodermal cells. In the linear variant of CCLE, genetic mosaicism/epigenetic modification of keratinocytes and the immune system in the lines of Blaschko may play a potential role in development of this specific LCLE variant[6, 13-15]. The keratinocytes in the lines of Blaschko triggered by trauma, irritation, or ultraviolet light may express antigens and introduce a wide range of stimuli crucial for the development of CCLE. Keratinocyte apoptosis has also been indicated as a key event in initiating CLE through various apoptotic pathway such as p53, TNF-α, and Fas/FasL. However, it remains a speculation whether these genetically variant keratinocytes are indeed lacking proteins essential for regulation of apoptosis, are there immunological distinct showing aberrant MHC expression, or are there relaxing abnormal aberrant cytokines[8, 15-17].

The differential diagnosis of LCLE includes other acquired inflammatory skin diseases distributed along the lines of Blaschko, such as linear morphea, linear lichen planus, lichen striatus, linear psoriasis, linear lichen sclerosus and linear granuloma annulare[9, 16, 17].

Although, the final diagnosis of CLE was achieved by histopathological examination, dermatoscopic findings also play an important role to identify the accurate diagnosis. Lallas et al. described the dermatoscopic criteria of DLE located on the face, trunk and extremities, and correlated them to the underlying histopathology. Perifollicular whitish halo, follicular keratotic plugs and telangiectasia were the most common dematoscopic findings in DLE[18].

In our patient, some dermatoscopic features correspond well to the histopathological findings. Dermatoscopic findings showed follicular keratotic plugs, telangiectatic vessels, pigmentation, and structureless whitish areas, representing histopathologic features of follicular hyperkearatosis, telangiectasia, pigmenatary incontinence, and diffuse dermal fibrosis, respectively. In addition to dematoscopic and histopathologic diagnosis, direct immunofluorescence (DIF) of lesional skin is also the useful adjunction for diagnosing LDLE. These findings are granular depositions of IgG, IgM, IgA, and C3 along the dermoepidermal junction and periadnexal structure. Nonetheless, a negative DIF result does not exclude the diagnosis[9, 16, 17, 19, 20].

In linear DLE, topical high-potency corticosteroids and sun protection are the mainstay of therapy, whereas topical calcineurin inhibitors are reserved for long-term maintenance. In active discoid lesions, intralesional injection of corticosteroid can provide benefit. In cases presenting with widespread lesions unresponsive to topical therapy, systemic therapy may be considered. Antimalarials, is the first-line drug for LDLE, especially hydroxychloroquine. Other systemic treatments including oral corticosteroids, dapsone, thalidomide, mycophenolate mofeti, azathioprine and retinoids have been reported to be benificial [6, 20].

References:

1. Umbert P, Winkelmann RK. Concurrent localized scleroderma and discoid lupus erythematosus. Cutaneous 'mixed' or 'overlap' syndrome. Arch Dermatol. 1978;114:1473-8.
2. Abe M, Ishikawa O, Miyachi Y. Linear cutaneous lupus erythematosus following the lines of Blaschko. Br J Dermatol. 1998;139:307-10.
3. Bernardez C, Prieto-Torres L, Macias E, Ramirez-Bellver JL, Haro-Ramos R, Diaz-Recuero JL, et al. Concurrent presentation of cutaneous lesions of deep linear morphoea and discoid lupus erythematosus. Lupus. 2016;25:204-8.
4. Campos-Munoz L, Fueyo-Casado A, Conde-Taboada A, Carranza-Romero C, Lopez-Bran E. Clinical Images: Progressive linear blaschkoid lupus erythematosus. Arthritis Rheumatol. 2017;69:2250.
5. Elbendary A, Griffin J, Li S, Tlougan B, Junkins-Hopkins JM. Linear Sclerodermoid Lupus Erythematosus Profundus in a Child. Am J Dermatopathol. 2016;38:904-9.
6. Jin H, Zhang G, Zhou Y, Chang C, Lu Q. Old lines tell new tales: Blaschko linear lupus erythematosis. Autoimmun Rev. 2016;15:291-306.
7. Lueangarun S, Subpayasarn U, Chakavittumrong P, Tempark T, Suthiwartnarueput W. Lupus panniculitis of the scalp presenting with linear alopecia along the lines of Blaschko. Clin Exp Dermatol. 2017;42:705-7.
8. Mao QX, Zhang WL, Wang Q, Xiao XM, Chen H, Shao XB, et al. Linear cutaneous lupus erythematosus/discoid lupus erythematosus in an adult. Postepy Dermatol Alergol. 2017;34:177-9.
9. Marinho AK, Ramos TB, Barbosa DM, Kozmhinsky V, Takano DM, Figueira MM. Linear cutaneous lupus erythematosus following the lines of Blaschko - Case report. An Bras Dermatol. 2016;91:510-3.
10. Ogawa M, Muro Y, Sugiura K, Sakakibara A, Akiyama M. Magnetic resonance imaging findings are useful for evaluating the three-dimensional development and follow-up of linear lupus erythematosus profundus. Lupus. 2015;24:1214-6.
11. Meena M, Balai M, Mehta S, Bharti G. Chronic Cutaneous Lupus Erythematosus in a Blaschkoid Pattern over Face. Indian J Dermatol. 2018;63:180-1.

12. Saha A, Seth J, Pradhan S, Dattaroy S. Disseminated discoid lupus erythematosus with a linear lesion on the forearm and hand: A rare presentation and review of literature. Indian J Dermatol Venereol Leprol. 2017;83:586-9.
13. Daldon PE, Lage R. Linear chronic discoid lupus erythematosus following the lines of Blaschko. An Bras Dermatol. 2011;86:553-6.
14. Lin JH, Dutz JP, Sontheimer RD, Werth VP. Pathophysiology of cutaneous lupus erythematosus. Clin Rev Allergy Immunol. 2007;33:85-106.
15. Seitz CS, Brocker EB, Trautmann A. Linear variant of chronic cutaneous lupus erythematosus: a clue for the pathogenesis of chronic cutaneous lupus erythematosus? Lupus. 2008;17:1136-9.
16. Engelman DE, Kotz EA, 3rd, Maize JC, Sr. Linear cutaneous lupus erythematosus in the lines of Blaschko. Pediatr Dermatol. 2007;24:125-9.
17. Green JJ, Baker DJ. Linear childhood discoid lupus erythematosus following the lines of Blaschko: a case report with review of the linear manifestations of lupus erythematosus. Pediatr Dermatol. 1999;16:128-33.
18. Lallas A, Apalla Z, Lefaki I, Sotiriou E, Lazaridou E, Ioannides D, et al. Dermoscopy of discoid lupus erythematosus. Br J Dermatol. 2013;168:284-8.
19. George PM, Hood AF, Rest EB. Histopathology and immunofluorescence of discoid lupus erythematosus in children. Pediatr Dermatol. 1996;13:269-73.
20. Okon LG, Werth VP. Cutaneous lupus erythematosus: diagnosis and treatment. Best Pract Res Clin Rheumatol. 2013;27:391-404.