Linear, Blaschkoid, Hypopigmented and Childhood Mycosis Fungoides

Ceyda Çaytemel, Begüm Çalım Gürbüz[1]

From the Department of Dermatology and Venereology, Acibadem University, School of Medicine, Istanbul, [1]Department of Pathology, Istanbul Basaksehir Cam and Sakura City Hospital, Istanbul, Turkey
E-mail: ceydacaytemel@gmail.com

Indian J Dermatol 2023:68(6):727

Sir,

A 6-year-old Syrian girl presented with pigment loss on her right leg for 1 year that began as a small area, then expanding distally without history of previous erythema. Examination disclosed linear, hypopigmented and non-scaly patch starting from the lateral right knee and extending to the right big toe in a pattern following Blaschko's lines [Figure 1a and b]. She denied systemic or constitutional symptoms. The physical examination (including lymph nodes, liver and spleen) was normal. In the biopsy of lesional skin, large, hyperchromatic, atypical lymphocytes with cerebriform nuclei were detected in the basal layer, showing a single array pattern and forming nests in some areas addition to band-like lymphocytic infiltration [Figure 2]. These lymphoid cells showed positive expression with CD2, CD3, CD4, CD5, CD7 and CD8 [Figure 3]. Similar findings were seen in the biopsy from the right foot. With these clinical and pathological findings, the patient was diagnosed with hypopigmented mycosis fungoides. There were no abnormal findings in the laboratory examinations, peripheral blood smear, chest radiograph and abdominal ultrasonography. Narrowband ultraviolet B light therapy was started 3 days a week in addition to topical clobetasol dipropionate ointment. Regression was noticed in the lesions after 30 sessions [Figure 1c], and the light theraphy was continued once a week. After 1 year, the patient's lesions were less conspicuous. No systemic involvement was detected.

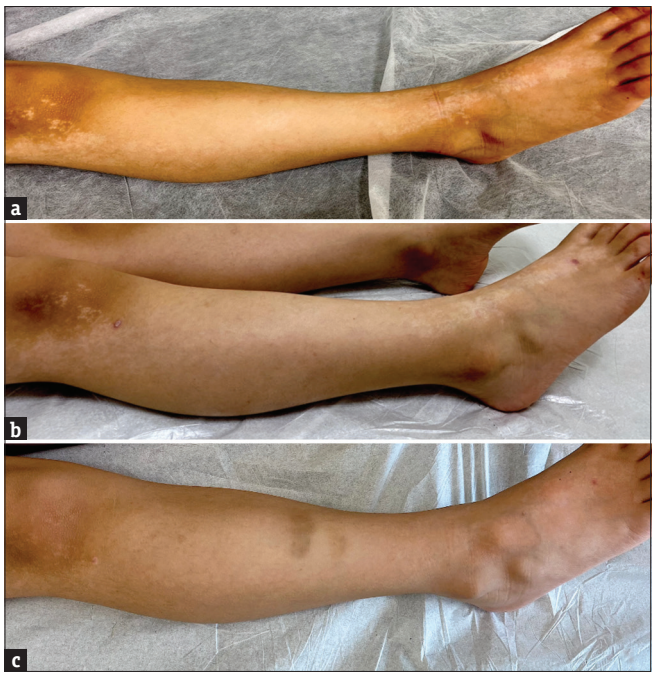

Figure 1: Linear hypopigmented patch extending from the outside of the right knee to the big toe (a and b) and post-treatment image of lesions (c)

Hypopigmented micosis fungoides (HMF) is relatively rare among all MF cases, but it is the most common form of MF observed in paediatric population.[1] It is more common in dark-skinned individuals, including Middle Eastern.[2] Although this paediatric and Syrian case report fits the classical definition of HMF, it is clinically distinct in its distribution.

Unilateral cases are extremely rare among all cutaneous lymphomas.[3] In addition to being unilateral, this case showed a linear blaschkoid distribution in the lower extremity. Happle suggested that blaschkoid dyspigmentation disorders are the result of genetic mosaicism.[4] Dermatoses following Blaschko's lines mostly affect ectodermal components such as melanocytes, keratinocytes and epidermal appendages.[5] However, some studies have stated that mosaicism can also be demonstrated in lymphocytes and/or skin fibroblasts.[6] Environmental factors, trauma, infections and genetic predisposition are blamed for linear lesions due to mosaicism.[5] It has been suggested that viral infections trigger T cell response by inducing cutaneous antigen expression and forming mosaicism.[7] In the hypopigmented variant of MF, impaired melanogenesis and damaged melanocytes are considered to cause hypochromic patches. Affected melanocytes of ectodermal origin and T cells may have caused linear, blaschkoid and

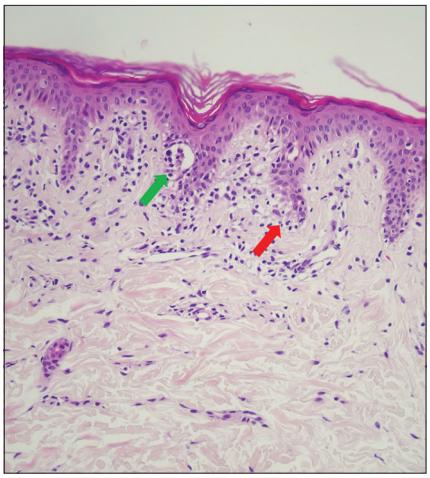

Figure 2: Atypical lymphocyte infiltration/epidermotropism in the basal layer that line up one by one (red arrow), atypical lymphoctes with clear halos make Pautrier microabcess (green arrow) in some areas (HE ×100)

Table 1: Differential diagnosis of hypopigmented mycosis fungoides

Primary hypopigmented lesions	Blaschkoid hypopigmented lesions	Post-inflammatory linear hypopigmentation; previous history of papulosquamous rash
Vitiligo; aqquired, sharply demarcated macules and/or patches	Ito hypomelanosis; starts during infacy, nonprogressive, neurological and ocular abnormalities	Linear psoriasis
Pityriasis versicolor; fine scale, seborrheic distribution, positive skin scrabbing	Nevus depigmentosus; congenital, nonprogressive	Linear lichen planus
Pityriasis alba; fine scale, poorly demarcated lesions, history of atopy	Incontinentia pigmenti-stage 4; previous vesicular, verrucous, and hyperpigmentation stages, developmental anomalies of the skin, hair, nail, teeth and eyes	Lichen striatus
Idiopathic macular hypomelanosis; ill- defined, nummular, symmetric lesions without scale		Blaschkitis
Syphilis; positive serological tests		Pityriasis lichenoides chronica
Leprosy; birth or residence in an endemic region, peripheral neuropathy, patches of alopecia		Inflammatory linear verrucous epidermal nevus

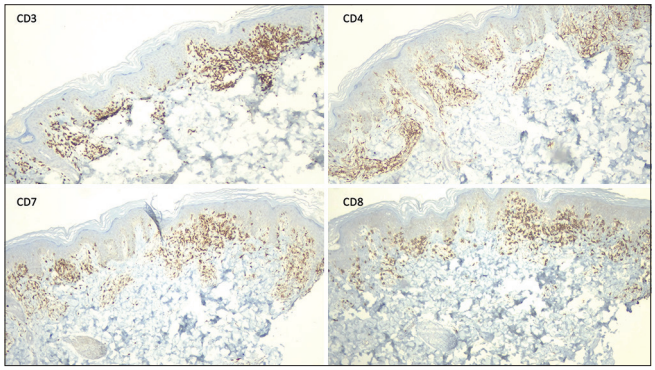

Figure 3: Immunohistochemical staining with CD3, CD4, CD7 and CD8 (×100)

hypopigmented lesions as seen in this patient (see Table 1 for differential diagnosis). Kaplan *et al.* presented a case of MF with distribution in a linear blaschkoid pattern in 2020, in which they detected an acquired mosaic GNAS gene mutation as a segmental superimposed manifestation on an existing congenital mutation suggested by Happle in acquired skin diseases.[8]

We presented a case of HMF due to its rare clinical appearance and we think genetic mosaicism seen in blaschkoid lesions may contribute to understanding the pathogenesis of HMF cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Pradhan D, Jedrych JJ, Ho J, Akilov OE. Hypopigmented mycosis fungoides with large cell transformation in a child. Pediatr Dermatol 2017;34:e260-4.
2. Martínez Villarreal A, Gantchev J, Lagacé F, Barolet A, Sasseville D, Ødum N, *et al.* Hypopigmented mycosis fungoides: Loss of pigmentation reflects antitumor ımmune response in young patients. Cancers (Basel) 2020;12:2007.
3. Hodak E, Phenig E, Amichai B, Feinmesser M, Kuten A, Maron L, *et al.* Unilesional mycosis fungoides: A study of seven cases. Dermatology 2000;201:300-6.
4. Cohen J 3rd, Shahrokh K, Cohen B. Analysis of 36 cases of Blaschkoid dyspigmentation: Reading between the lines of Blaschko. Pediatr Dermatol 2014;31:471-6.
5. Kartal Durmazlar SP, Altunel CT. Dermatoses following the lines of Blaschko. Dermatoz 2014;3:1–28.
6. Thomas IT, Frias JL, Cantu ES, Lafer CZ, Flannery DB, Graham JG Jr. Association of pigmentary anomalies with chromosomal and genetic mosaicism and chimerism. Am J Hum Genet 1989;45:193-205.

7. Lipsker D, Stark J, Schneider GA. Blaschkolineäre Dermatitis im Erwachsenenalter (Morbus Grosshans-Marot) mit antinukleären Antikörpern [Blaschko-linear dermatitis in adulthood (Grosshans-Marot disease) with antinuclear antibodies]. Hautarzt 2000;51:774-7.
8. Kaplan Y, Sarig O, Rabinowitz T, Mohamad J, Shomron N, Sprecher E, *et al*. Linear Mycosis fungoides ın a blaschkoid distribution. Harefuah 2020;159:25-8.

Access this article online

Quick Response Code:

Website: www.e-ijd.org

DOI: 10.4103/ijd.ijd_270_23

How to cite this article: Çaytemel C, Çalım Gürbüz B. Linear, blaschkoid, hypopigmented, and childhood mycosis fungoides. Indian J Dermatol 2023;68:727.

Received: March, 2023. **Accepted:** August, 2023.