Dermoscopy of Primary Cutaneous Anaplastic Lymphoma Kinase Negative Large T-Cell Lymphoma

Keshavmurthy A. Adya, Ajit B. Janagond, Arun C. Inamadar, Satish Arakeri[1]

Departments of Dermatology, Venereology and Leprosy, [1]Pathology, Shri B M Patil Medical College, Hospital and Research Center, BLDE (Deemed to be University), Vijayapur, Karnataka, India

Anaplastic lymphoma kinase (ALK) negative T-cell lymphomas are a rare subset of CD30 positive anaplastic large cell lymphomas (ALCL). The systemic ALCL can be both nodal and extranodal affecting a wide age group. These tumors can be ALK-positive (presence of chromosomal rearrangements of the *ALK* gene at 2p23) which is associated with a good prognosis. The ALK-negative forms have variable clinical and genetic features and are associated with a poorer prognosis. The primary cutaneous ALCL (PC-ALCL) are typically ALK-negative and have a favorable prognosis.[1] In this report, we describe the clinical, dermoscopic, and immunohistochemical features of the disease in an elderly Indian male along with a discussion of the differential diagnoses.

A 65-year-old farmer presented with a persistent, slow-growing, exophytic, fleshy ulcerated nodule, measuring about 2 × 2 cm over the left extensor forearm from the past 6 months [Figure 1]. Patient also informed about similar but smaller lesions adjacent to the nodule which regressed spontaneously. Polarized dermoscopy (using DermLite™ DL3, 3Gen Inc., San Juan Capistrano, CA, USA) revealed yellow-white structureless areas, polymorphic linear vessels (serpentine, coiled, single and complex looped), and red structureless areas (hemorrhagic spots) over a red-pink background with a peripheral thick scaly collarette [Figure 2]. Based on the history, clinical and dermoscopic findings, differential diagnoses of certain non-neoplastic (deep fungal infection) and neoplastic (poroma, undifferentiated squamous cell carcinoma, and amelanotic melanoma) disorders were considered. Deep fungal infection was preferentially considered because of the patient's occupation and the infrequent occurrence of malignancies in our skin type. Histopathology revealed diffuse dermal infiltrate of atypical lymphoid cells with mitotic figures and prominent nuclei suggestive of cutaneous lymphoma. At high magnification, large cells with abundant pale cytoplasm, nuclear polymorphism, and prominent nucleoli were seen [Figure 3]. Immunohistochemically, the cells were positive for CD30/CD2/CD5 and negative for CD3/CD7/CD45RO/LCA (CD45)/CD56/ALK/CD25 [Figure 4]. Based on these findings, anaplastic large T-cell (ALK-negative) lymphoma was diagnosed and the patient was referred to an oncologist. Systemic workup revealed no evidence of extracutaneous disease and thus a final diagnosis of primary cutaneous ALK-negative anaplastic large T-cell lymphoma was established.

The PC-ALCL accounts for about 30% of all the primary cutaneous lymphomas and commonly affects males in their 6th decade. It is characterized by solitary or multiple grouped indolent nodular or noduloulcerative lesions mostly confined to the skin with only about 10% disseminating as extracutaneous disease accounting for a good prognosis. Some of the lesions may spontaneously regress as well. The common histology is that of a diffuse dermal infiltrate of large atypical cells with abundant pale cytoplasm and round or horseshoe-shaped nuclei with prominent nucleoli. Immunohistochemistry typically shows membranous or globular pattern of CD30 expression. Anaplastic lymphoma kinase is mostly negative which differentiates PC-ALCL from the systemic form. Management of PC-ALCL includes surgical, radiotherapy, and chemotherapeutic modalities depending

How to cite this article: Adya KA, Janagond AB, Inamadar AC, Arakeri S. Dermoscopy of primary cutaneous anaplastic lymphoma kinase negative large t-cell lymphoma. Indian Dermatol Online J 2022;13:254-6.

Received: 08-Jul-2021. **Revised:** 09-Aug-2021. **Accepted:** 10-Aug-2021. **Published:** 03-Mar-2022.

Address for correspondence:
Dr. Arun C. Inamadar, Department of Dermatology, Venereology and Leprosy, Shri B M Patil Medical College, Hospital and Research Center, BLDE (Deemed to be University), Vijayapur - 586 103, Karnataka, India.
E-mail: aruninamadar@gmail.com

Access this article online

Website: www.idoj.in

DOI: 10.4103/idoj.idoj_441_21

Quick Response Code:

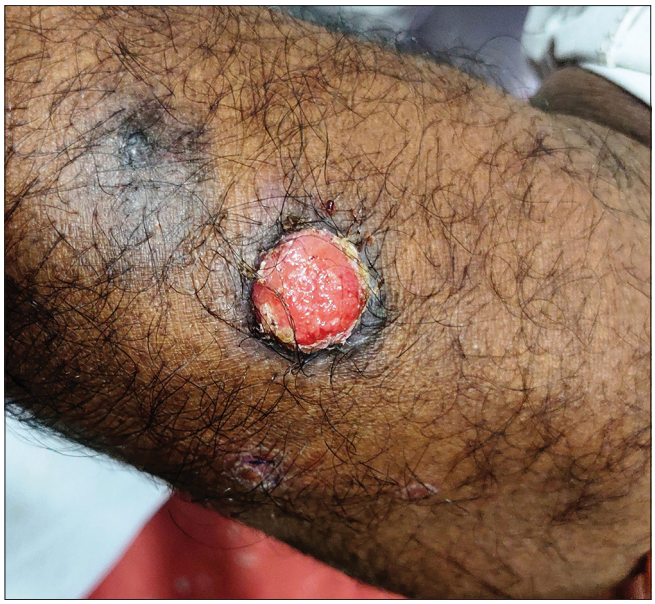

Figure 1: Exophytic noduloulcerative lesion on the extensor aspect of the left forearm

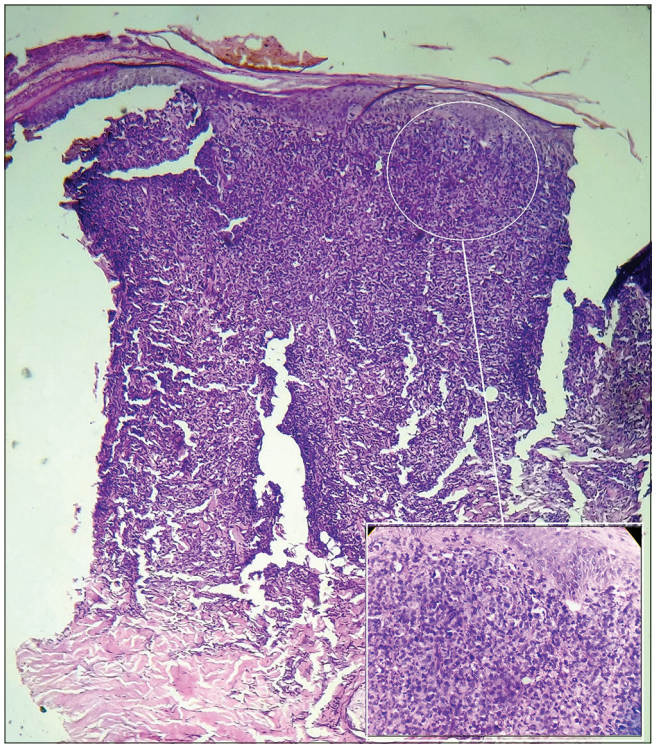

Figure 3: Photomicrograph showing diffuse dense dermal atypical mononuclear cells with prominent nuclei (H&E, ×10). In set: Higher magnification showing large cells with pale cytoplasm, mitotic figures, nuclear polymorphism and prominent nucleoli. [H&E, Original magnification × 10 (in-set ×40)]

on the disease extent and presence or absence of extracutaneous dissemination.[2-4]

Various dermoscopic patterns are described in different cutaneous lymphomas.[5-7] The predominant dermoscopic features of papular and nodular lesions include salmon-colored background, polymorphic vessels (serpentine and arborizing), orange color, shiny white structures, and follicular plugs.[6,7] While some authors believe that the features are characteristic of specific lymphoma types, others are of the opinion that they only suggest a possibility of cutaneous lymphoma in general.[5,6] In regard to PC-ALCL, we found only two case reports describing the dermoscopic features—a pinkish yellow-brown background with arborizing-to-polymorphous, and helical vessels.[8,9] The features in our case are pretty much identical except for the morphology of the vessels being predominantly serpentine and looped rather than branching and helical. The yellow-white structureless areas correspond to the dense dermal infiltrate and the pinkish background hue to the vasodilatation seen on histology.

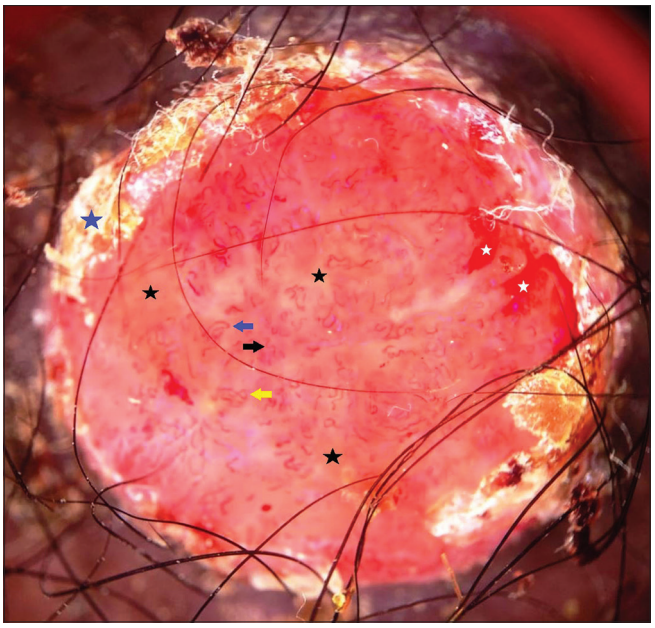

Figure 2: Polarized dermoscopy shows yellow-white structureless areas (black stars), polymorphic vessels (serpentine [black arrow], coiled [yellow arrow] and loop [blue arrow]) and hemorrhagic spots (white stars) over a red-pink background with a peripheral thick scaly collarette (blue star). [Original magnification ×10]

As to the differential diagnoses for our case, a deep fungal infection such as fixed cutaneous sporotrichosis was considered owing to the patient's occupation and dermoscopic features of yellow-white structureless areas with telangiectatic vessels being suggestive of a granulomatous lesion.[10] In regard to the neoplastic differential diagnoses, poroma was considered due to the pink-red background, complex loop and coiled vessels, and presence of collarette.[11,12] Finally, malignant tumors such as undifferentiated squamous cell carcinoma and nodular amelanotic melanoma were considered as they exhibit pink-white or pink-red background, hemorrhage, and polymorphic vessels as observed in the lesion.[13]

To conclude, primary cutaneous ALCL is an uncommon entity that, as per the current understanding,

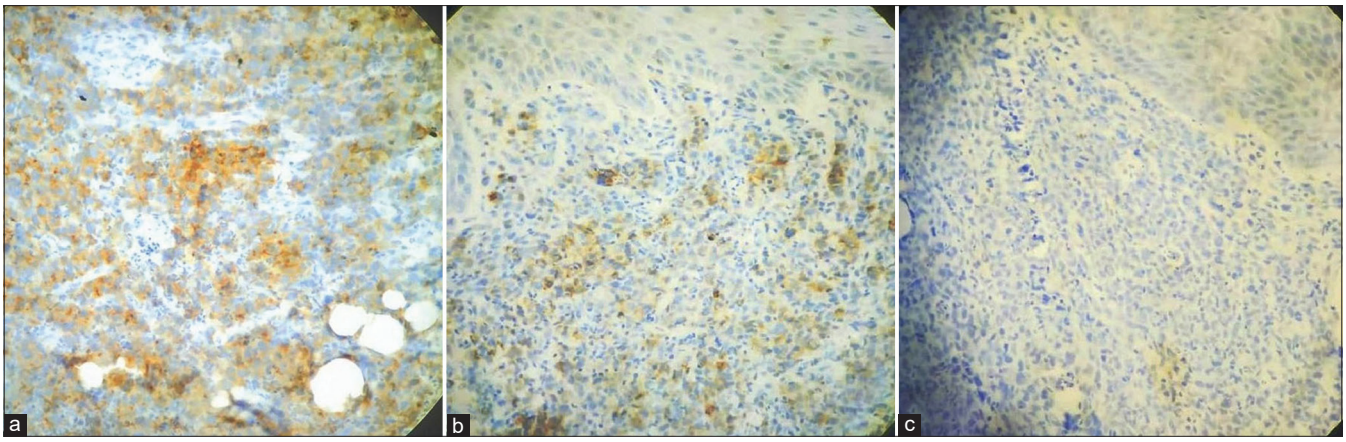

Figure 4: Immunohistochemistry shows positivity with CD5 (a), CD30 (b) and negative ALK (c). [Original magnification ×10]

dermoscopically exhibits features such as pink-brown background and polymorphic vessels as described generally in cutaneous lymphomas irrespective of a specific type. We tend to agree with the opinion that such features are non-specific and can only suggest a possibility of cutaneous lymphoma which makes it imperative to interpret them contextually. We report this case not only for its rarity but also to suggest that although primary cutaneous malignancies are uncommon in our skin, it may be worthwhile to include cutaneous lymphoma among the differential diagnoses for the clinical and dermoscopic presentation as in this case.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Kao EY, Mukkamalla SK, Lynch DT. ALK negative anaplastic large cell lymphoma. StatPearls. Treasure Island (FL): StatPearls Publishing; 2021.
2. Jacobsen E. Primary cutaneous anaplastic large cell lymphoma. Available from: https://www.uptodate.com/contents/primary-cutaneous-anaplastic-large-cell-lymphoma. [Last accessed on 2021 Jul 08].
3. Moodley N, Nombona P, Mosam A. Primary cutaneous anaplastic large-cell lymphoma. Dermatopathology (Basel) 2019;6:163-9.
4. Brown RA, Fernandez-Pol S, Kim J. Primary cutaneous anaplastic large cell lymphoma. J Cutan Pathol 2017;44:570-7.
5. Ghahramani GK, Goetz KE, Liu V. Dermoscopic characterization of cutaneous lymphomas: A pilot survey. Int J Dermatol 2018;57:339-43.
6. Geller S, Marghoob AA, Scope A, Braun RP, Myskowski PL. Dermoscopy and the diagnosis of primary cutaneous B-cell lymphoma. J Eur Acad Dermatol Venereol 2018;32:53-6.
7. Navarrete-Dechent C, Del Puerto C, Abarzúa-Araya Á, Molgó M, Geller S, Andreani S, *et al*. Dermoscopy of primary cutaneous B- and T-cell lymphomas and pseudolymphomas presenting as solitary nodules and tumors: A case-control study with histopathologic correlation. Int J Dermatol 2019;58:1270-6.
8. Uzuncakmak TK, Akdeniz N, Karadag AS, Taskin S, Zemheri EI, Argenziano G. Primary cutaneous CD 30 (+) ALK (-) anaplastic large cell lymphoma with dermoscopic findings: A case report. Dermatol Pract Concept 2017;7:59-61.
9. Jeter J, Lewis W, Wohltmann W. A case of primary cutaneous anaplastic large cell lymphoma presenting with corkscrew vessels on dermoscopy. Available from: https://apps.dtic.mil/sti/citations/AD1081629. [Last accessed on 2021 Jul 8].
10. Vinay K, Bhattacharjee R, Bishnoi A, Chatterjee D, Rudramurthy S, Dogra S. Dermatoscopic features of cutaneous sporotrichosis. J Eur Acad Dermatol Venereol 2020;34:e718-20.
11. Chessa MA, Patrizi A, Baraldi C, Fanti PA, Barisani A, Vaccari S. Dermoscopic-histopathological correlation of eccrine poroma: An observational study. Dermatol Pract Concept 2019;9:283-91.
12. Takada T. Diagnostic features of a non-pigmented eccrine poroma with a collarette: Histopathological and dermoscopic correlation. Clin Case Rep 2021;9:1601-4.
13. Adya KA, Ankad BS. Premalignant and malignant tumors. In: Ankad BS, Mukherjee SS, Nikam BP editors. Dermoscopy-Histopathology Correlation. 1st ed. Singapore. Springer. p. 251-78.