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

lymphoma Anaplastic 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 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 vellow-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

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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 CD3/CD7/CD45RO/LCA (CD45)/CD56/ALK/CD25 Figure 4]. Based these findings. anaplastic 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 multiple grouped indolent nodular 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, chemotherapeutic modalities

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

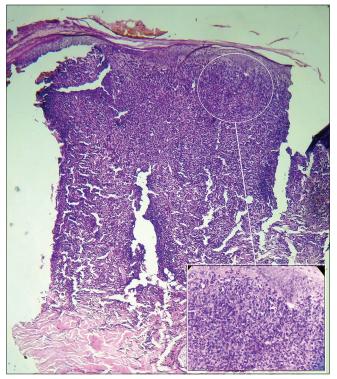


Figure 3: Photomicrograph showing diffuse dense dermal atypical mononuclear cells with prominent nuclei (H&E, ×10). In set: Higher magnification showing large cells with pale cytoplasm, mitotic figures, nuclear polymorphism and prominent nucleoli. [H&E, Original magnification × 10 (in-set ×40)]

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

Various dermoscopic patterns are described in different cutaneous lymphomas.<sup>[5-7]</sup> The predominant dermoscopic features of papular and nodular lesions

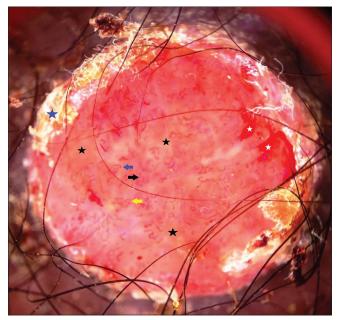


Figure 2: Polarized dermoscopy shows yellow-white structureless areas (black stars), polymorphic vessels (serpentine [black arrow], coiled [yellow arrow] and loop [blue arrow]) and hemorrhagic spots (white stars) over a red-pink background with a peripheral thick scaly collarette (blue star). [Original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

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.<sup>[5,6]</sup> 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.

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,

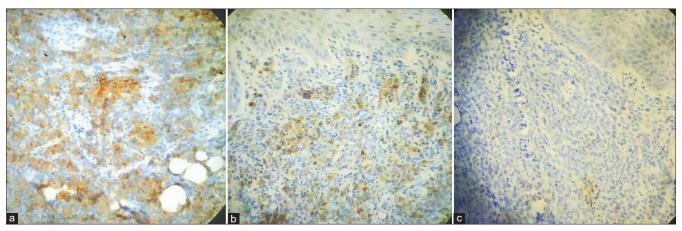


Figure 4: Immunohistochemistry shows positivity with CD5 (a), CD30 (b) and negative ALK (c). [Original magnification ×10]

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.

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#### Conflicts of interest

There are no conflicts of interest.

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