Dermoscopic features of three cases of Langerhans cell histiocytosis

Sir,

The first case was a 10-month-old male child who was brought by his parents with complaints of scalp scaling of 2 months duration associated with fever. Cutaneous examination revealed multiple skin-colored to erythematous papules with greasy scales all over the scalp [Figure 1a]. Other cutaneous and mucosal examination was within normal limits. Dermoscopic examination (HEINE DELTA20® Dermatoscope, 10 × magnification) under nonpolarized contact dermoscopy revealed irregular vascular blotches and brown structure-less area and crust [Figure 2].

The second case was a 14-month-old male child who presented with asymptomatic skin rash over the face and trunk of 6 months duration. Cutaneous examination revealed multiple hypopigmented papules, few with central umbilication distributed over the face and trunk [Figure 1b]. The umbilicated papules under nonpolarized contact dermoscopy (10x magnification) showed central brown dots, and structure-less area surrounded by white structureless area and the hypopigmented papules showed diffuse white structure-less area [Figure 3].

Case 3 was a 1-year-old male child who presented with history of asymptomatic skin rash over the trunk of 4 months duration. It was associated with swelling over the frontal scalp and loss of appetite. Cutaneous examination revealed multiple skin-colored to hypopigmented papules of size ranging from 1 mm × 1 mm to 3 mm × 3 mm over the anterior and posterior aspect of the trunk [Figure 1c]. Under nonpolarized contact dermoscopy (10x magnification) the papules revealed perifollicular white homogenous areas in amoeboid (amoeba-like pseudopods extending at the periphery), nebuloid (indistinct margins, merging into surrounding skin) and petaloid pattern (well-defined borders) [Figure 4a][1]. At places, the white homogenous areas coalesced to form an irregular geographic pattern. Few papules demonstrated vascular blotches [arrows, Figure 4b].

In all cases, a systemic work-up was done, and histopathological examination of the skin biopsy showed dermal infiltration of histiocytes with a reniform nucleus and abundant eosinophilic cytoplasm consistent with a diagnosis of Langerhans cell histiocytosis, which was confirmed by

Table 1: The dermoscopic features of various dermatosis that were considered in the differential diagnosis of our cases

Our cases	Dermoscopic features	Differential diagnosis	Dermoscopic features
Case 1	Irregular vascular blotches, brown homogenous area and crusts	Seborrheic dermatitis	Yellow scales, patchy dotted vessels, arborizing red lines, atypical red vessels, glomerular vessels, twisted red loops and perifollicular white scale[5]
		Scalp psoriasis	White scale, homogenous dotted vessels, arborizing red lines and atypical red vessels[5]
		Tinea capitis	Comma hairs, corkscrew hairs, black dots, broken hairs, scale, peripilar cast, alopecia and pustule[6]
		Folliculitis decalvans	Tufted hairs, follicular pustules, perifollicular scale, crust and interfollicular twisted capillary loops[7]
Case 2	Central brown dots and structureless area surrounded by white structureless area	Pityriasis lichenoides et varioliformis acuta	Central whitish patch/crusted brownish structure surrounded by ring of dotted/linear/glomerular vessels and red globules[8]
Case 3	Vascular blotch and perifollicular white homogenous area in amoeboid, nebuloid and petaloid pattern	Guttate psoriasis	White scale and homogenous dotted vessels[9]
		Atopic dermatitis	Structureless erythema, scales, patchy dotted vessels and linear vessels[10]
		Exanthematous drug eruption	Dotted/linear irregular vessels[11]
		Pityriasis lichenoides chronica	Orange-yellowish structureless area, focal dotted vessels and linear branching and irregular vessels and milky-red globules[9]
		Juvenile xanthogranuloma	Setting sun appearance, clouds of paler yellow globule, whitish streak, smaller diameter linear branched vessels and subtle pigment network[12]
		Sarcoidosis	Pink homogenous background, orange yellow area/orange globules, arborizing vessels/linear vessels and central scar-like area and white fine scale[13,14]

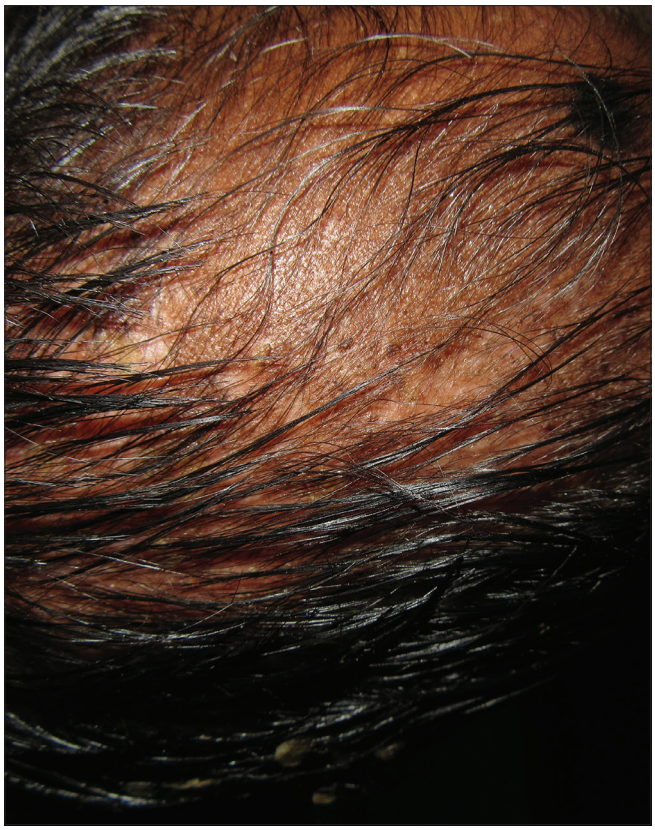

Figure 1a: Multiple skin-colored to erythematous papules over the scalp with greasy scale

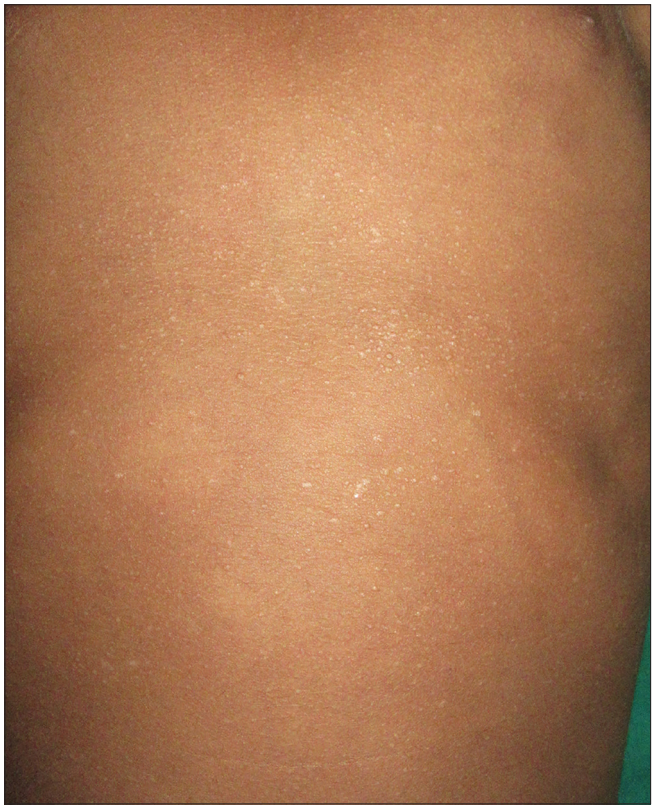

Figure 1c: Multiple skin-colored to hypopigmented papules over the trunk

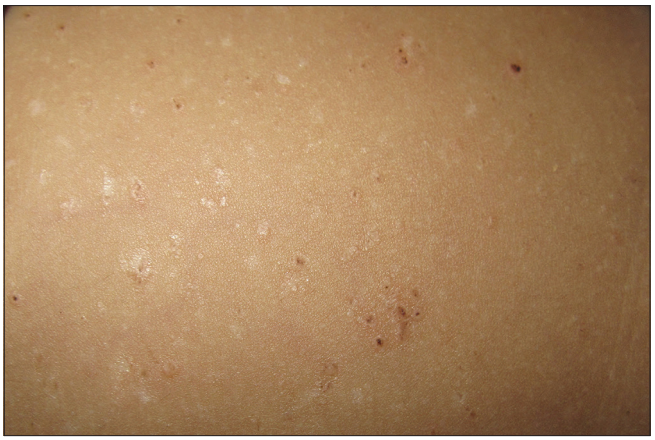

Figure 1b: Multiple hypopigmented papules with few having central umbilication

positive S100 and CD1a and negative CD68 immunostaining [Figure 5a-c].

Langerhans cell histiocytosis is a rare proliferative Langerhans cell disorder of unknown etiology. Cutaneous presentation of Langerhans cell histiocytosis can have a diverse morphological presentation that includes papules, pustules, vesicles, nodules, petechiae and ulcers. The clinical morphology of Langerhans cell histiocytosis can mimic inflammatory dermatosis such as seborrheic dermatitis to malignant condition such as leukemia cutis. Consequently, the diagnosis and treatment of the condition is delayed.[2] Out of the three cases described by us, the first case had only scalp involvement and the second and third cases had only cutaneous involvement but with different morphology.

Dermoscopy is a simple, rapid, noninvasive outpatient procedure that unfolds the diagnosis of a particular dermatosis by giving a certain subtle clue, thus alleviating the need for biopsy. The data regarding the dermoscopic features of Langerhans cell histiocytosis is very scanty and includes widespread red-blue lacunes of different sizes, reddish-purple areas and brown dots and central white area with peripheral telangiectasia.[2-4] The dermoscopic features of various dermatological conditions that were considered in the differential diagnosis of each of our cases are delineated in Table 1.

The dermoscopic features that may help in differentiating Langerhans cell histiocytosis involving the scalp from the scalp seborrheic dermatitis and scalp psoriasis are the presence of vascular blotch, brown homogenous area/crust and the absence of dotted and nondotted vessels.[5] The vascular blotch clinically corresponds to the purpuric tinges associated with the lesion. Interestingly, the papules in the third case were not clinically associated with any purpuric tinges, but few of them demonstrated vascular

Figure 2a: Dermoscopy of the scalp papule showing irregular vascular blotches, brown structure-less area and crust

Figure 2b: Dermoscopy of the area around the papule showing irregular vascular blotch

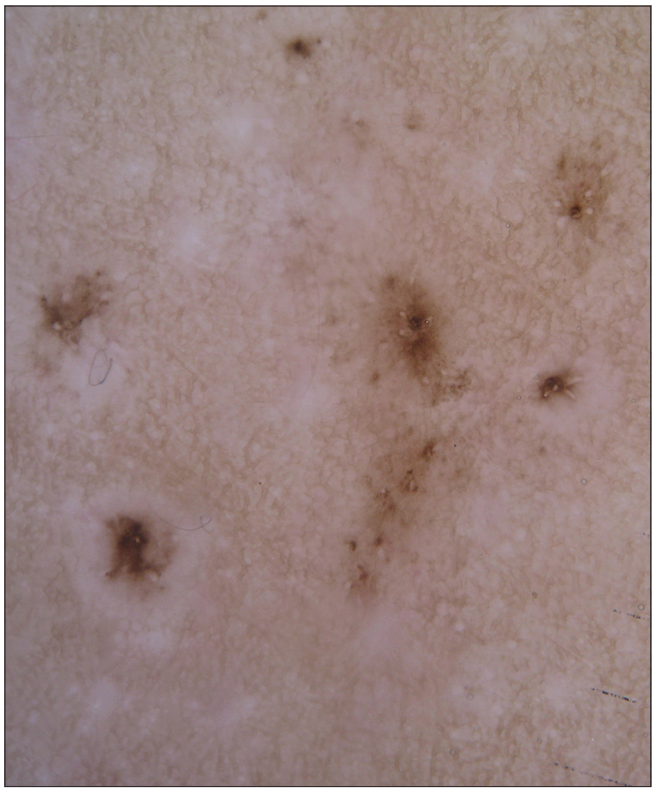

Figure 3a: Dermoscopic examination of the umbilicated papules showing central brown dots/structure-less area surrounded by white structure-less area

Figure 3b: Dermoscopic examination of hypopigmented papules showing diffuse white structure-less area

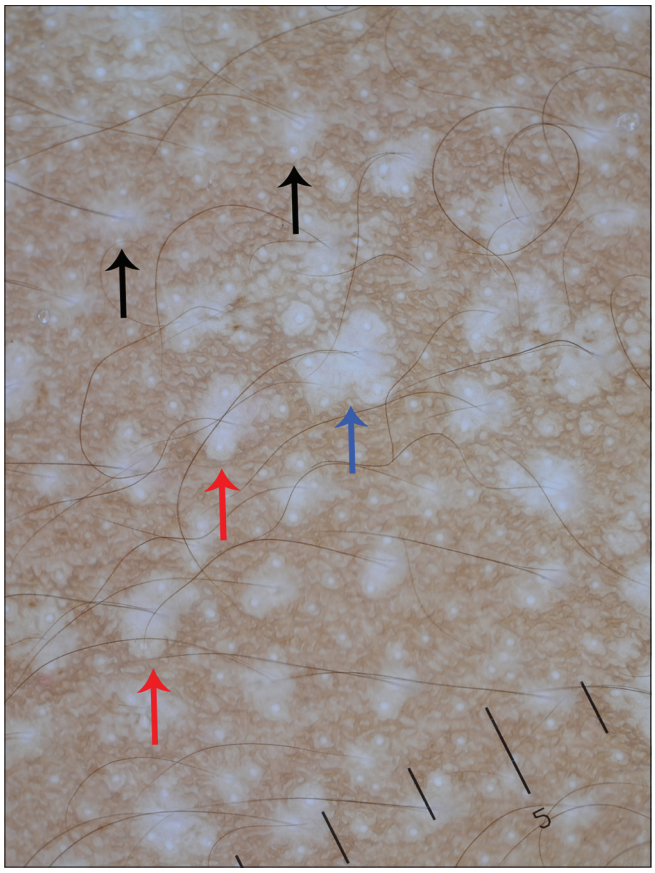

Figure 4a: Dermoscopic examination showing perifollicular white structure-less area in an amoeboid (red arrows), nebuloid (black arrows) and petaloid pattern (blue arrow)

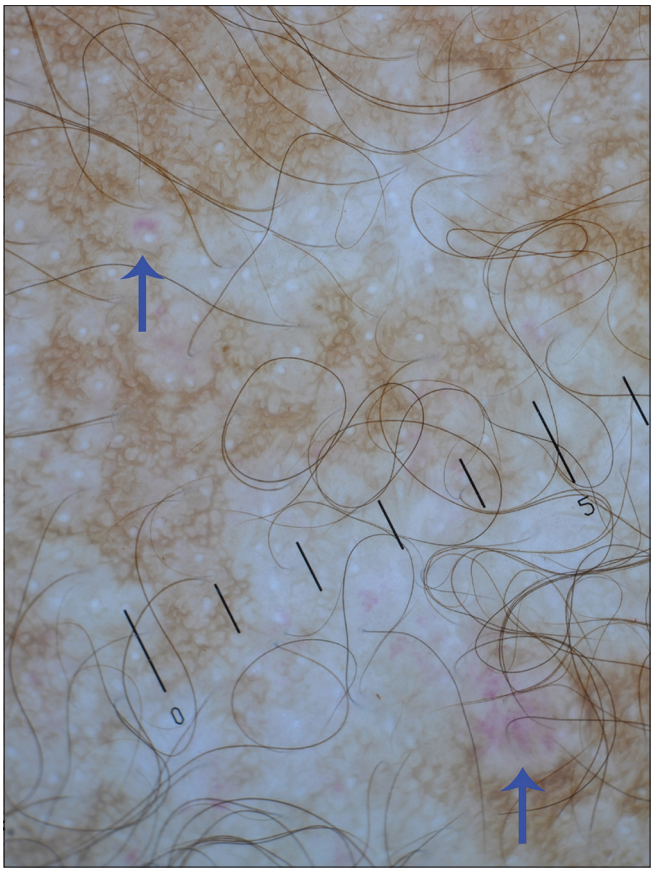

Figure 4b: Dermoscopic examination showing the presence of vascular blotches (arrows)

blotch under the dermoscope (blue arrows, Figure 4b), which suggests that dermoscopy can be useful in unveiling the subclinical hemorrhages that cannot be appreciated by the naked eye. The presence of a white structure-less area and absence of a yellow structure which corresponds to the infiltration of xanthomized histiocytes can be an important clue in differentiating Langerhans cell histiocytosis from non-Langerhans cell histiocytic disorders such as juvenile xanthogranuloma, benign cephalic histiocytosis and reticulohistiocytosis.[12]

Analyzing the dermoscopic features of previously reported cases and our present cases, the presence of vascular blotch seems to be a common dermoscopic finding. The cases reported by Rubio-González *et al*.[2] and Murata *et al*.[4] demonstrates vascular blotch in the dermoscopic images, although they have not mentioned the same in the text. The vascular blotch present under dermoscopy histologically corresponds to dermal hemorrhage [Figure 5b]; the white homogenous area corresponds to the dermal Langerhans cell infiltration without epidermal involvement and brown dots/structure-less area/crust to the epidermal infiltration and necrosis by Langerhans cells.

In conclusion, Langerhans cell histiocytosis, similar to its clinical presentation, can have diverse dermoscopic features. In this report, we are describing various dermoscopic features in three cases of Langerhans cell histiocytosis, and we hope that further studies on a larger case series will be helpful in revealing additional dermoscopic features and diagnostic clues.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

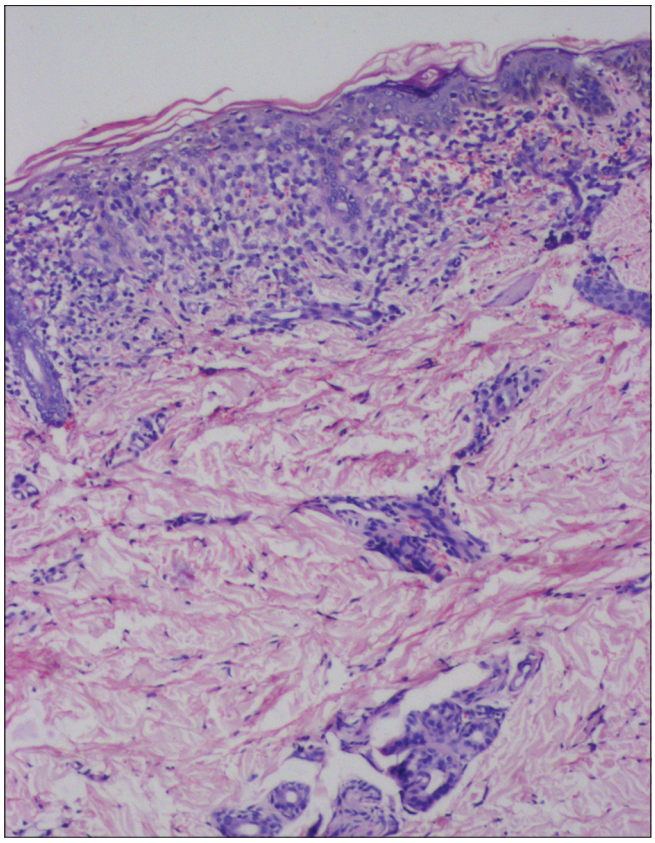

Figure 5a: Dermal infiltration of Langerhans cells with areas of hemorrhage (Hematoxylin and eosin, ×100)

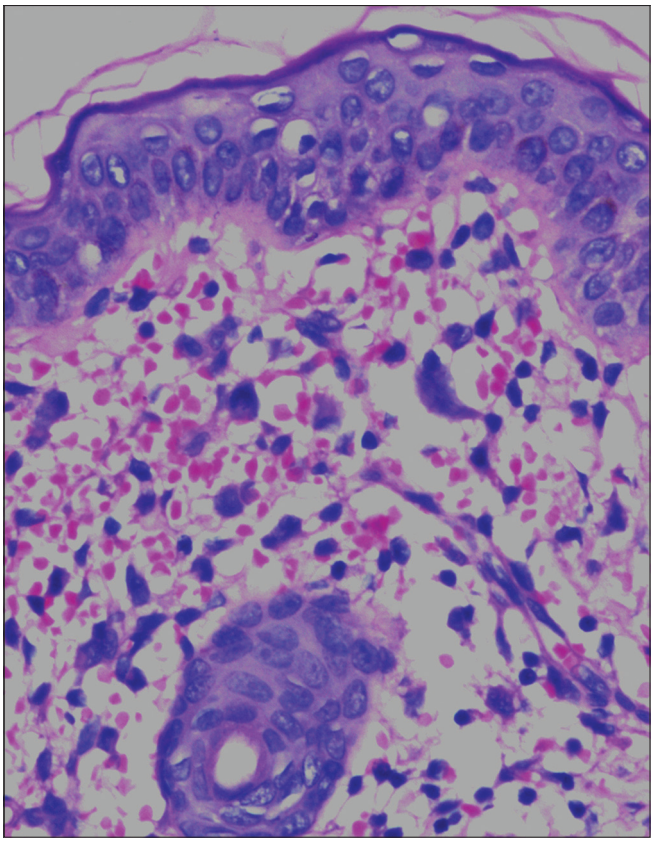

Figure 5b: The Langerhans cells with a reniform nucleus and abundant eosinophilic cytoplasm (Hematoxylin and eosin, ×400)

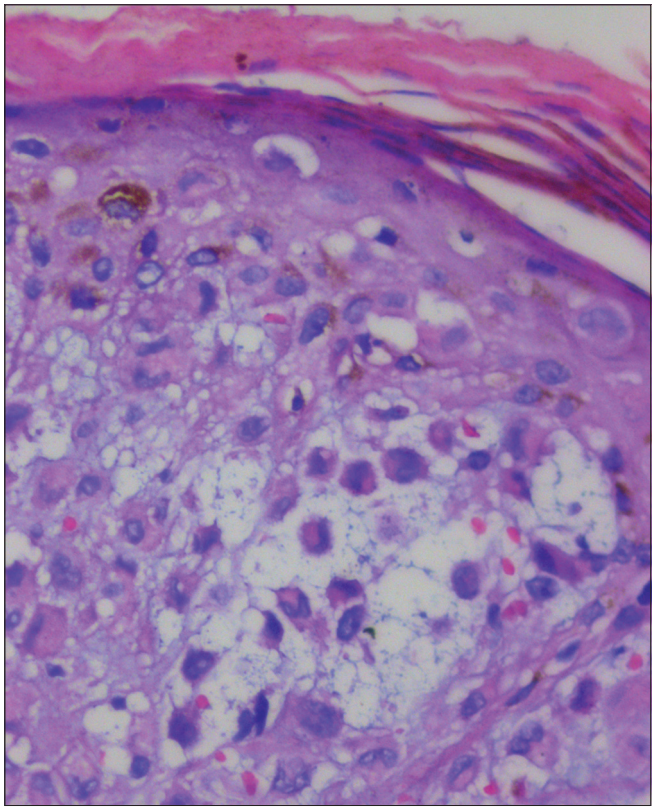

Figure 5c: Epidermal infiltration by Langerhans cell (Hematoxylin and eosin, ×400)

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