Atypical clinical and dermoscopic features of hobnail hemangioma

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Sir,

A 36-year-old woman (Patient 1) with skin phototype-V, presented with a 10-month history of an asymptomatic pigmented lesion on the abdomen. Cutaneous examination revealed a solitary 1.5 cm × 1 cm, firm, non-tender, pigmented plaque on the abdomen [Figure 1a]. Differential diagnosis of dermatofibroma, dermatomyofibroma and melanocytic nevus was considered. Dermoscopic examination under nonpolarized contact mode (HEINE DELTA20® Dermatoscope, Germany, 10 × magnification) showed a cerebriform pattern. The dermoscopic features observed were a gray background, comedo-like openings, linear irregular crypts and keratotic plugs [Figure 1b]. The excisional biopsy of the lesion showed multiple lymphangioma-like dilated spaces with intraluminal projections lined by hobnail endothelial cells in the upper dermis, and slit-like vascular spaces dissecting the collagen bundles along with perivascular lymphocytic infiltration in the lower dermis. Perl's Prussian blue stain demonstrated dermal hemosiderin [Figure 2].

A 44-year-old man (Patient 2) with skin phototype-V had a slow-growing lesion on the abdomen for the last 6 months. Cutaneous examination revealed a solitary 5 mm × 5 mm firm non-tender shiny dome-shaped skin-colored nodule on the abdomen [Figure 3a]. Differential diagnosis of solitary neurofibroma, solitary circumscribed neuroma and fibroepithelial polyp was considered. Dermoscopy of the nodule demonstrated a gray homogenous area, along with brown peppering [Figure 3b]. Shave biopsy of the lesion showed, proliferating endothelial cells admixed with multiple dilated vascular spaces lined by hobnail endothelial cells. Perl's Prussian blue showed occasional hemosiderin deposition [Figure 4].

In both cases, a final diagnosis of hobnail hemangioma was made based upon the histopathological findings.

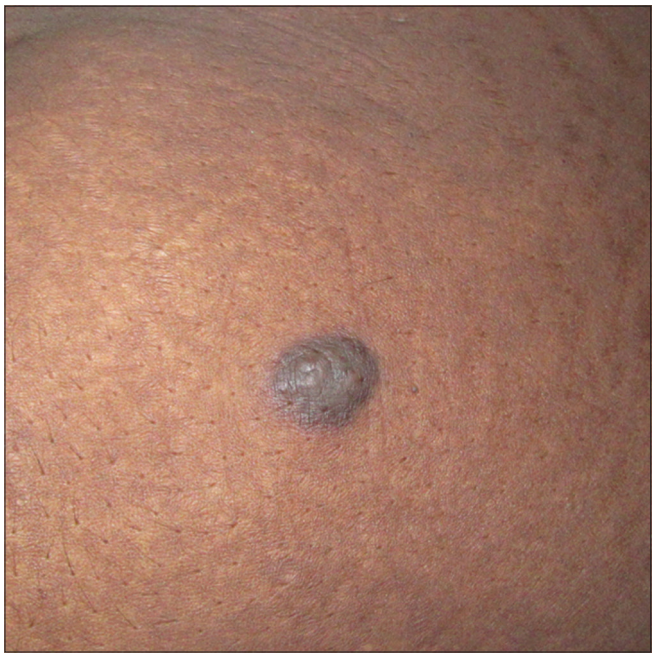

Figure 1a: Solitary firm pigmented plaque on the abdomen

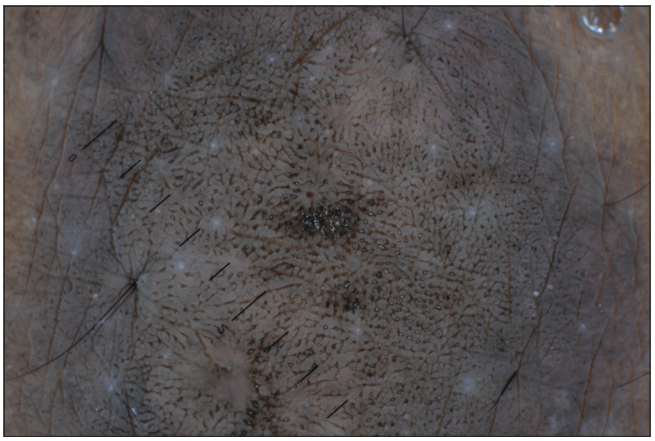

Figure 1b: Dermoscopic examination (under nonpolarized contact mode, HEINE DELTA20® Dermatoscope, Germany, ×10) demonstrating a cerebriform pattern, a gray background, comedo-like openings, linear irregular crypts and keratotic plugs

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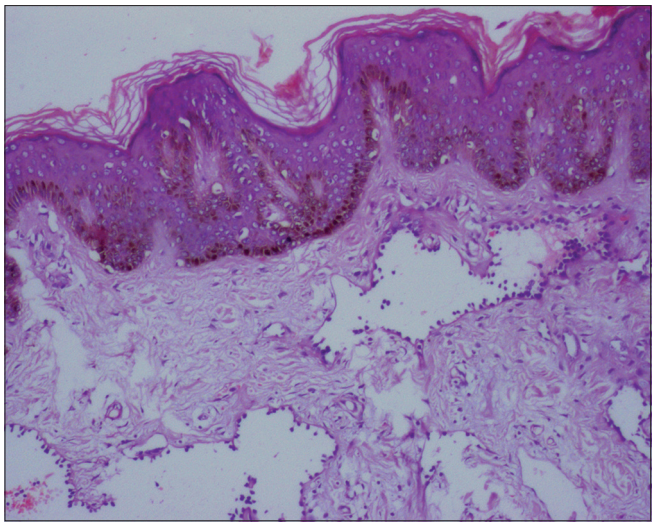

Figure 2a: Multiple lymphangioma-like dilated spaces in the upper dermis lined by hobnail endothelial cells with an overlying hypertrophic and hyperpigmented epidermis (H and E, ×100)

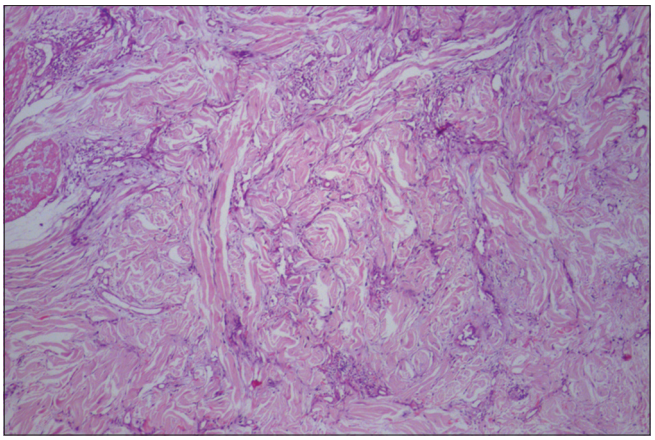

Figure 2b: Slit-like vascular spaces dissecting the collagen bundles in the lower dermis (H and E, ×100)

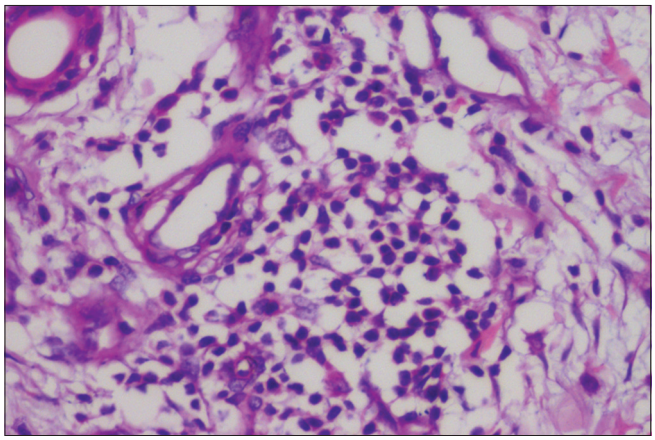

Figure 2c: Perivascular lymphocytic infiltration (H and E, ×400)

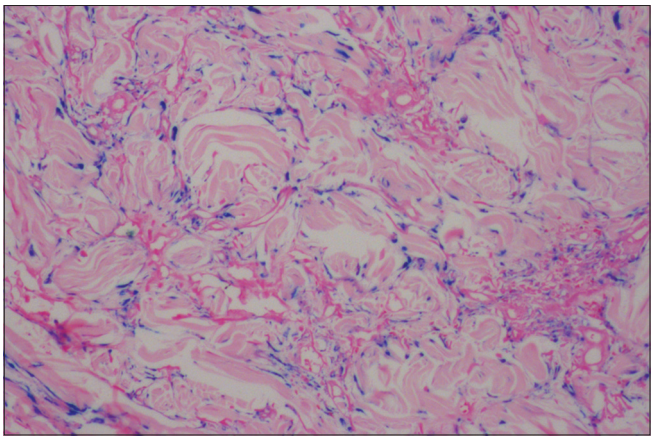

Figure 2d: Special stain highlighting hemosiderin (Perl's Prussian blue stain, ×100)

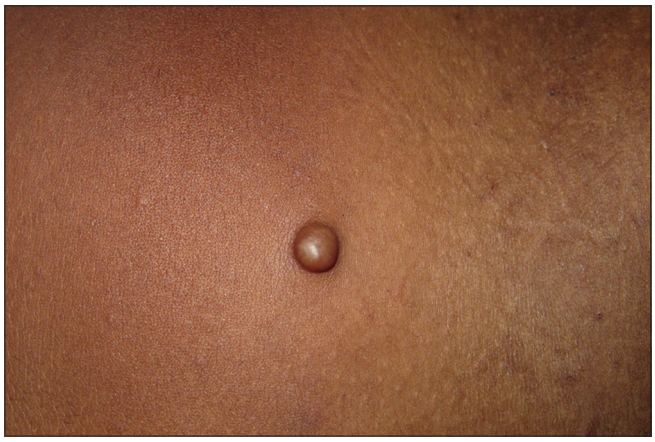

Figure 3a: Solitary firm non-tender dome-shaped nodule on the abdomen

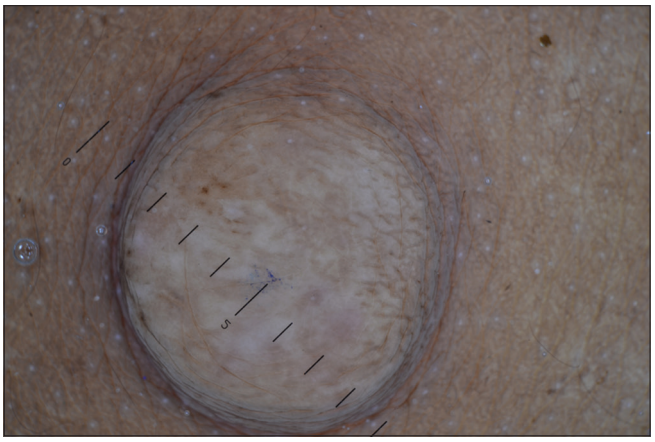

Figure 3b: Dermoscopic examination (under nonpolarized contact mode, HEINE DELTA20® Dermatoscope, Germany, ×10x) demonstrating a gray homogenous area and brown peppering

Hobnail hemangioma, also known as targetoid hemosiderotic hemangioma is a benign cutaneous vascular lesion. It commonly occurs in young to middle-aged persons and has a slight female preponderance. A solitary asymptomatic papule on the extremities and trunk, in a young to middle-aged adult, is the most common presentation of hobnail hemangioma. The size ranges from 0.6 cm to 2 cm. As the name suggests,

a targetoid appearance with a central violaceous to red macule or papule surrounded by a pale rim, and an outermost ecchymotic ring, usually provides a clue to the diagnosis. The targetoid appearance is not universal; cases with nontargetoid appearance have been described.[1,2] In the case of a nontargetoid or atypical presentation, it can be mistaken for other cutaneous vascular (hemangioma, angiokeratoma, Kaposi's sarcoma), melanocytic (melanocytic nevus, dysplastic nevus, melanoma) and fibrohistiocytic (dermatofibroma, fibroma) tumors. Both of the described cases neither had a targetoid pattern nor any clinical clue like reddish-violaceous color, to suggest the vascular nature of the lesions. In the first case, a pigmented firm plaque, we considered differentials of dermatofibroma, dermatomyofibroma and melanocytic nevus. In the second case, a firm skin-colored dome-shaped nodule, we considered differentials of solitary neurofibroma, solitary circumscribed neuroma and fibroepithelial polyp.

The dermoscopic examination is a rapid, noninvasive procedure that provides subtle clues to the diagnosis of a cutaneous lesion. For a vascular malformation/tumor, the presence of lagoons/clods of varying colors during dermoscopic examination is a constant feature. Although not specific to any particular vascular tumor/malformation, their presence guides the clinicians to rule out nonvascular etiology. A recent review of dermoscopic features of hobnail hemangioma described red lacunae, dark lacunae, whitish structures, a central homogeneous area, intermediate yellowish areas, peripheral red-violaceous ring, vascular structures and pigment network.[2]

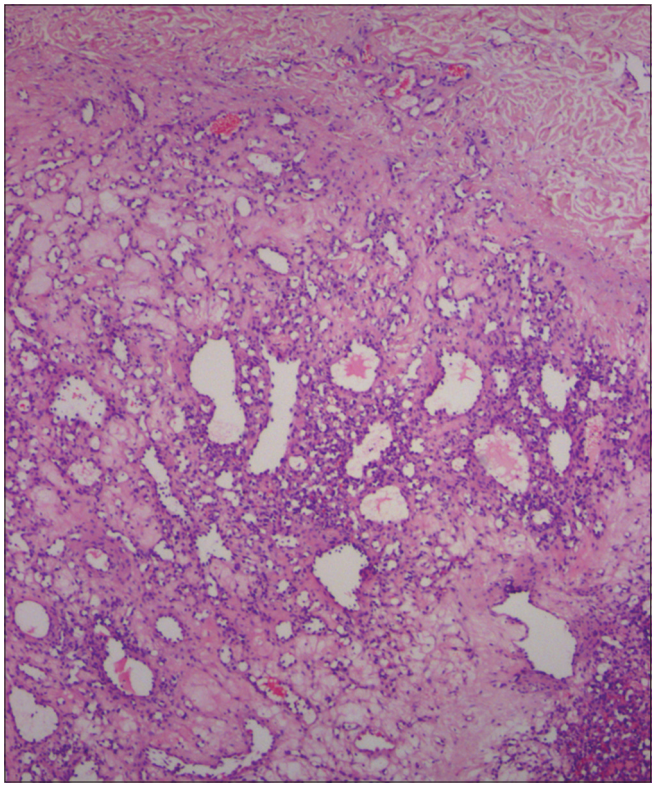

Figure 4a: Proliferating endothelial cells admixed with multiple dilated vascular spaces in the dermis (H and E, ×50)

Zaballos *et al*. analyzed 35 cases of hobnail hemangioma and reported dermoscopic features such as reddish-violaceous, and brown homogenous areas (85.7%), red and dark lacunae (74.3%), chrysalis (42.8%), vascular structure (40%) and delicate pigment network (14.3%).[3] The various patterns observed by them were; a central lacuna and peripheral homogenous area with or without intermediate skin-colored, yellow or white circular homogeneous area (71.4%), diffuse reddish-violaceous homogenous area and delicate pigment network and red lacuna alone (single report). The other dermoscopic features reported for hobnail hemangioma are enlisted in Table 1.[4-10] From the various reports, it is evident that a lagoon of variable color, with or without a targetoid pattern, is a consistent feature of hobnail hemangioma. In the two of our described cases, dermoscopy neither demonstrated

Table 1: Dermoscopic features described for hobnail hemangioma

Report	Dermoscopic features
Sahin *et al*.[1]	Red lagoons sharply demarcated black macule and violaceous maculae ring A homogenous red area with indistinct violaceous ring Red to reddish-blue lagoon, black macule surrounded by erythema
Morales-Callaghan *et al*.[4]	A central round bright red round structures, smaller round pale-pink structures admixed with a diffuse pink-white pigmentation and peripheral pigment network
Lacarrubba *et al*.[5]	A central round red lagoon-like area with a peripheral thin pigment network
Ghibaudo *et al*.[6]	A central closely packed red-violaceous ovoid structures, whitish-veil and peripheral ecchymotic ring
Ertam *et al*.[7]	Red lagoon-like areas, red homogenous area, linear white structure and a peripheral pigment network
Avci *et al*.[8]	Sharply demarcated blue-red lagoons
Ibrahim and Shwayder[9]	Well-defined red circular blebs
Piccolo *et al*.[10]	A central reddish structureless area with chrysalis-like structures and a peripheral pigment network

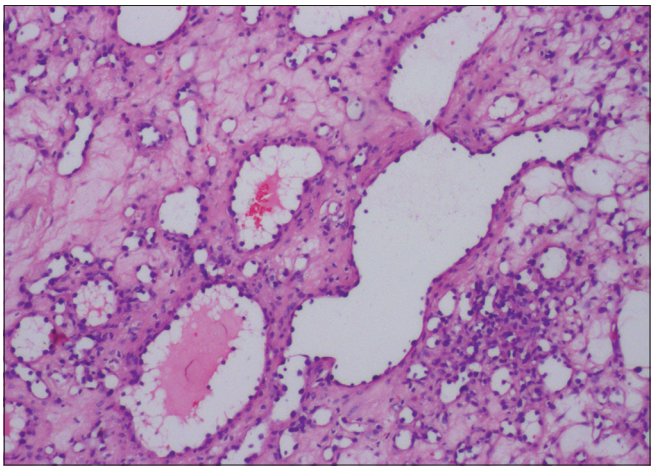

Figure 4b: Dilated vascular spaces lined by hobnail endothelial cells (H and E, ×100)

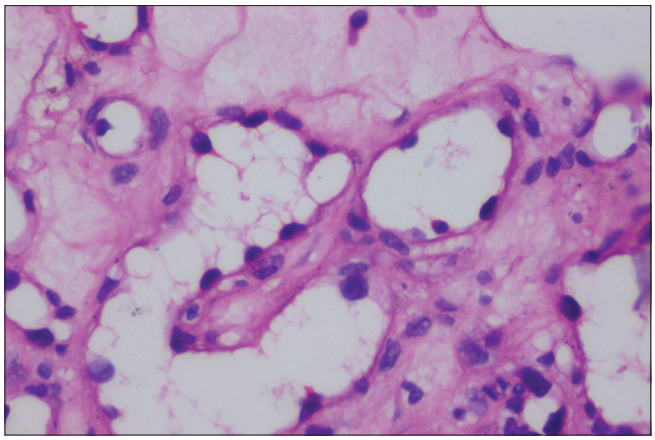

Figure 4c: Hobnail endothelial cells (H and E, ×400)

Table 2: Comparison between dermoscopic features of hobnail hemangioma, dermatofibroma and neurofibroma

Tumour	Dermoscopic features
Hobnail hemangioma	A lagoon of varying color (red/reddish-blue/dark) A targetoid pattern: A central lacuna and peripheral homogenous area with or without intermediate skin-colored/yellow/white circular homogeneous area Other features: Diffuse reddish-violaceous homogenous area, chrysalis, pigment network
Dermatofibroma	Typical appearance: A central scar-like area with peripheral delicate pigment network Different dermoscopic patterns: As a result of various combinations of the following features; white scar-like area, pigment network, homogenous pigmentation, white network, brown dots and globules, cerebriform pattern, linear irregular crypts It can vary depending upon histopathological subtypes
Neurofibroma	A yellow, yellow-brown, brown, skin-colored to gray background/homogenous area with exaggerated skin markings Other features: Pigment network-like area, blanchable erythema, brown globules, fingerprint-like structures

a targetoid pattern nor had any dermoscopic clue to the diagnosis of a vascular lesion, thus mislead the diagnosis. The first case dermoscopically mimicked a dermatofibroma or seborrheic keratosis due to the presence of cerebriform pattern, and gray background. Another rare clinical mimic, aneurysmal fibrous histiocytoma, commonly demonstrates a blue homogenous area that may accompany other shades of color like pink, reddish-brown or white. The blue homogenous area corresponds to the dermal hemosiderin deposition. Other features described are pigment network, branched streaks, white crystalline structure, a rainbow pattern and vessels ranging from dotted, polymorphous and linear irregular vessels. The second case dermoscopically mimicked a neurofibroma due to a featureless pattern. The dermoscopic difference between hobnail hemangioma, neurofibroma and dermatofibroma is mentioned in Table 2.[2,3,11-14] In case-1, the hyperplastic and hyperpigmented epidermis masked the dermoscopic vascular structures. In case-2, along with mild hyperplastic and hyperpigmented epidermis, the relatively lower dermal location of the vascular lesion may be responsible for the absence of the dermoscopic vascular feature. The final diagnosis of both cases was revealed by histopathological examination.

In conclusion, we describe two cases of hobnail hemangioma with atypical clinical and dermoscopic features. The vascular nature of the hobnail hemangioma was concealed due to the absence of clinical and dermoscopic vascular features like reddish-violaceous color and vascular lagoons, respectively. At times, the dermoscopic examination can be misleading, especially in cases of atypical clinical presentation, in which case a histopathological examination is essential for the diagnosis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for 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.

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Conflicts of interest

There are no conflicts of interest.

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Antiphospholipid antibodies in a patient of Lucio phenomenon presenting with the gangrene of digits

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Sir,

Lucio leprosy is a diffuse form of lepromatous leprosy, commonly seen in Mexico and Costa Rica, not infrequent in the Gulf Coast, but quite rare in the rest of the world.[1] Lucio leprosy presents as a slowly progressive, diffuse infiltration of the skin of the face and most of the body without any previous evidence of discrete lesions. The disease is often unmasked by a specific severe reaction state, the Lucio phenomenon—presenting clinically as multiple, well-defined, angular jagged purpuric lesions evolving into massive ulcerations. The antiphospholipid antibodies were originally detected and used by Wasserman for the diagnosis of syphilis. It was subsequently found that antiphospholipid antibodies are not only specific for syphilis but are also found in autoimmune diseases such as systemic lupus erythematosus and with other infections.[2] Here we present a case of Lucio phenomenon in a patient of Lucio leprosy with positive antiphospholipid antibodies presenting with digital gangrene.

A 45-year-old woman was referred to the dermatology department for sudden blackening of toes and fingers for 6 days along with fever and arthralgia for the last 8 days. There was no clinically relevant significant past, personal and obstetric history. Menstrual history was unremarkable. Systemic examination was normal except for mild splenomegaly. Bilateral pitting edema was present with tense, dry, xerotic skin of both the lower limbs. Blackish discoloration of the left 3rd, 4th, 5th toes, right great toe and right 3rd,4th 5th toes extending from the tip of toes to metatarsophalangeal joints, with similar blackish discoloration of the right ring finger extending from tip of the finger to distal interphalangeal joint on the palmar aspect of the hand were seen. Affected areas were hard, non-tender and cold on palpation, suggesting established gangrene. Multiple livedo racemosa-like lesions were present involving the bilateral lower limb and trunk. Multiple angulated purpuric necrotic lesions with surrounding ill-defined erythema were present on the left thigh with one lesion showing ulcerative changes. The patient had diffuse erythema of face with difficult to pinch tense shiny skin with complete loss of bilateral eyebrows and eyelashes. There was no infiltration of the ear lobes [Figure 1a-c].

Bilateral infraorbital, left ulnar, bilateral radial, left common peroneal and left posterior tibial nerves were thickened and tender. Temperature sensations were impaired on the face and were completely absent in all limbs. Fine touch sensation was lost in the dorsal aspect of bilateral hands and lower limbs. Arterial pulses were palpable and normal in all the limbs.

Relevant laboratory and radiological investigations are mentioned in Table 1.

Slit-skin smear examination from earlobes, eyebrows, normal looking skin of cheek and scraping from nasal mucosa

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