Verrucous eccrine angiomatous hamartoma

Sir,
A 15-year-old girl presented with a lesion on her left thigh for the last 1 year. The lesion was associated with pain and hyperhidrosis which was often provoked by physical activity and emotional stress. Systemic examination was normal.

Cutaneous examination revealed a 9 cm × 6 cm, bluish-black, verrucous, firm plaque on the medial aspect of left thigh [Figure 1a]. The margins of the plaque were irregular. The lesion was warm and tender. On stroking, the surface of the plaque showed oozing beads of a shiny, transparent and colorless fluid [Figure 1b]. The starch-iodide test was positive.

Biopsy from the plaque revealed hyperkeratosis and marked acanthosis with elongated rete ridges and increased epidermal basal layer melanin. The underlying dermis showed dilated eccrine glands and admixed capillaries within and around these glands [Figure 2a]. In addition, tufts and lobules of vascular proliferation with prominent endothelial cells and slit-like capillaries were present in the dermis [Figure 2b]. Accordingly, a diagnosis of eccrine angiomatous hamartoma was rendered. Surgical excision was planned for the patient; however, she was lost to follow-up.

The term eccrine angiomatous hamartoma was coined by Hyman et al. in the year 1968; however, the clinical description was first put forward by Lotzbeck, in 1895 and the earlier name was sudoriparous angioma. Eccrine angiomatous hamartoma is a rare, benign cutaneous proliferation of eccrine glands and thin-walled vascular channels. The lesions arise at birth or during childhood in 77% of cases. Extremities, palms and soles in particular, are the usual sites affected. Feet, face, neck and trunk may seldom be involved. Solitary, flesh-colored, blue-brown or reddish papules, plaques and nodules are characteristic.
Occasionally, eccrine angiomatous hamartoma may manifest multiple lesions. Pain and hyperhidrosis are reported in approximately 42% and 32%, respectively. Interestingly, verrucous changes in the epidermis were noted in the present case which is a rare finding in eccrine angiomatous hamartoma. Unencapsulated, dermal proliferation of mature-appearing eccrine secretory and ductal structures that are intimately associated with thin-walled angiomatous channels usually of a capillary nature but of variable size are the characteristic histopathological features of eccrine angiomatous hamartoma. The exact etiology of this entity is not known. Some authors have proposed a pathophysiological model which considers it a biochemical fault in the interactions between differentiating epithelium and subjacent mesenchyme that gives rise to an abnormal proliferation of adnexal and vascular structures. It might be caused by abnormal induction of heterotypic dependency during organogenesis. Late-onset lesions are related to recurrent trauma.

The differential diagnosis of eccrine angiomatous hamartoma includes tufted angioma, vascular malformations, macular telangiectasia, nevus flammeus, glomus tumor, smooth muscle hamartoma, congenital hamartoma of the eccrine sweat gland, eccrine nevus and single lesion of blue rubber bleb syndrome. These entities can be differentiated by histopathology. Of these, tufted angioma is a close differential as it may also manifest hyperhidrosis as seen in eccrine angiomatous hamartoma; however, histopathologically, tufted angioma shows a pandermal capillary proliferation with characteristic “cannon ball” appearance. Hypertrichosis, hyperhidrosis, pain or itching are valuable diagnostic clues. The natural history of eccrine angiomatous hamartoma is benign and typically slow-growing and hence, aggressive treatment is generally unwarranted. Simple excision is usually curative and is reserved for painful or cosmetically disfiguring lesions. Deep excision with full-thickness grafting or amputation of a finger or toe may be required for symptom control in those with larger lesions on acral parts. Botulinum toxin might be useful in hyperhidrotic cases.

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Conflicts of interest
There are no conflicts of interest.

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Letters to the Editor

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Unilateral monomorphic hypopigmented macules: A variant of Darier disease

Sir,

Darier disease was first described by Morrow in 1886 and independently, by Darier and White in 1889. Usually expressed clinically as hyperkeratotic lesions, primarily on the seborrheic areas, Darier disease has also been reported to have various morphological variants. We describe one such case presenting with monomorphic hypopigmented macules, distributed diffusely on the left half of the body.

A 39-year-old Indian woman presented with multiple non-scaly, non-tender 5 mm to 1 cm sized hypopigmented macules and papules, confined to the left side of the body and nail dystrophy [Figure 1a and b], except for a midline crossover to the right side, in the abdominal area [Figure 2a and b]. The lesions were noticed a decade ago, over the left side of her trunk and arm, later progressed distally to involve the ipsilateral lower limb and have been static since 4–5 years. Some finger and toe nails were dystrophic and showed a V-shaped notch at the free edge of their nail plates [Figure 1b]. Face, palms, soles and mucosae showed no abnormality. These lesions being asymptomatic

Figure 1a: Guttae leukodermic macules localized to the left lower limb with nail dystrophy
Figure 1b: Nails showing dystrophy and V-shaped nicks

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