Letters to the Editor

Idiopathic Eruptive Macular Pigmentation in an Indian Male

Sir,

We laud Subhadarshini et al.\(^1\) for publication of their interesting case of idiopathic eruptive macular pigmentation in an Indian male. As they have pointed out this case differs from others described earlier in two aspects, namely, the persistence of lesions unchanged for 16 years and an aggravation thereafter for one year and association of acanthosis nigricans in the axillae, a finding that has not been described earlier in idiopathic eruptive macular pigmentation (IEMP).

The authors, however, have not elaborated on the nature of the aggravation, as to whether there were new lesions developing or the old quiescent lesions started getting darker and/or thicker with velvety appearance. Furthermore, the duration of acanthosis nigricans has not been mentioned, particularly if it also developed at the same time as the aggravation of the lesions. This is important because it has in the past been suggested that IEMP is actually an eruptive form of acanthosis nigricans where metabolic changes are not evident and may represent an eruptive phase without associated metabolic changes.\(^2,3\) The finding of acanthosis nigricans of the axillae without metabolic changes in this case may add credence to the above suggestion if the aggravation of skin lesions had occurred in conjunction with the appearance of axillary acanthosis nigricans.

Since the publication of the paper on IEMP\(^4\) by Sanz de Galdeano et al. in 1996, most authors have used diagnostic criteria of IEMP suggested by them namely: (1) Eruption of brownish-black discrete nonconfluent asymptomatic macules involving the neck, trunk, and proximal extremities in children and adolescents. (2) Absence of any preceding inflammatory lesions. (3) No previous drug exposure. (4) Basal cell hyperpigmentation of the epidermis with dermal melanophages without any basal cell damage or lichenoid infiltrate. (5) Normal mast cell counts.

In a recent review of published literature on IEMP\(^5\) it was suggested by the authors that a revision of diagnostic criteria is due because finding of significant melanophages in the papillary dermis or an inflammatory infiltrate or interface changes suggests a dermal melanotic condition such as lichen planus pigmentosus, Ashy dermatosis or Riehl’s melanosis and should be considered as a negative criterion in the diagnosis of IEMP. Some authors, however, have expressed reservations on accepting the presence of significant dermal melanophages as being against the diagnosis of IEMP.\(^6\)

Idiopathic eruptive macular pigmentation is predominantly an epidermal hypermelanotic condition with papillomatosis (pigmented papillomatosis), a finding that it shares with acanthosis nigricans, confluent and reticulate papillomatosis of Gougerot–Carteaud, and some epidermal nevi. Histopathological examination is essential to differentiate IEMP from the more commonly occurring dermal melanotic conditions such as lichen planus pigmentosus and Ashy dermatosis, all of which show numerous dermal melanophages with variable inflammation and interface dermatitis.

Finding of melanophages in the upper dermis in pigmented skin of Fitzpatrick types 4 and 5 is not unusual even when there is no evident pathology. How then does one evaluate and report on the presence of melanophages to determine if their presence is significant? Is finding of a few small melanophages indicative of a dermal melanosis?

We use a simple semi-quantitative method to analyze the presence of melanophages in the dermis to evaluate their significance in the histopathological diagnosis of melanoses.

The criteria used are magnification at which they can be seen, their size and composition, and their number and distribution.

A. Melanophages seen clearly at low power (40×) are assigned a grade of 3; those seen only at intermediate power (100×) are grade 2 and grade 1 when one needs high power (400×) to appreciate the melanophages in the tissue sections.

B. Large size of melanophages that appear uniformly dark brown or brownish-black and are stuffed with melanin are seen in conditions of dermal melanoses such as lichen planus pigmentosus, pigmented contact dermatitis, Riehl’s melanosis, and FDE. On the other hand, small melanophages that appear to have fine granular light brown melanin are not significant as they are seen even in conditions that have primarily epidermal melanin like lentigines and Becker’s melanosis and even in normal pigmented skin.

C. Presence of large heavily pigmented melanophages over a large area of the upper dermis is indicative of dermal melanosis. Finding of occasional small poorly pigmented melanophages in one or two foci is incidental.

Applying these criteria to the current case reveals that no melanophages are evident at 40× magnification, as seen in Figure 2 in the case report, and are only seen as three small lightly pigmented melanophages in a single focus at 400× magnification in Figure 3 in the case report. In contrast, the uniform hyperpigmentation of the basal layer is seen clearly even at 40× magnification. Thus, the finding of these small focally located melanophages is not significant but causes confusion because one of the criteria...
mentioned in the diagnosis of IEMP by Sanz de Galdeano is dermal melanophages.

IEMP is an epidermal hypermelanosis and does not show significant melanophages in the papillary dermis. If it were to do so then differentiation from other dermal melanoses such as lichen planus pigmentosus would not be possible.

In summary, this case report is instructive in highlighting two points, namely, the possible association of IEMP with acanthosis nigricans and the need to accurately assess the significance of finding of melanophages in the upper dermis, which can be done by a simple semi-objective histological scale.

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Conflicts of interest

There are no conflicts of interest.

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Hypotrichosis in a Child with Olmsted Syndrome

Sir,

Olmsted syndrome is a rare and unique keratinizing disorder which presents with bilateral mutilating transgradient palmoplantar keratoderma and periorificial keratotic plaques. Other reported features include leukokeratosis of the tongue, ichthyotic lesions, pain, itching, absent premolar teeth, hearing loss for high frequencies, sclerosing cholangitis, short stature, and laxity of the large joints, linear hyperkeratotic follicular streaks, and acral hyperhidrosis.

Hypotrichosis has rarely been reported in Olmsted syndrome.

A 5‑year‑old male child presented with periorificial keratotic plaques associated with painful fissures and thickening of bilateral palms and soles [Figure 1]. He had flexion contracture in both hands for the past 2 years. The patient was the only child of a second‑degree consanguineous marriage. There was no history of similar complaints in the family. On general examination, there was pallor and grade III IAP (Indian Association of Pediatrics) protein energy malnutrition. On examination, the palms and soles showed keratoderma with flexion contracture of bilateral fingers [Figure 2]. The child was unable to walk because of the associated pain. There were hyperkeratotic plaques with fissuring around the perioral region, intranasal, external auditory canal, and in the intergluteal region [Figure 3]. The intranasal plaques caused difficulty in breathing. Scalp examination showed hypotrichosis with sparse, short, and light‑colored hair [Figure 4]. Light microscopic examination of hair shaft showed reduced pigmentation, reduced hair shaft diameter, and trichoschisis. Similar findings along with folliculocentric papules and empty follicles were seen in trichoscopy [Figure 5]. Ophthalmic examination showed...