Idiopathic Eruptive Macular Pigmentation - Uncommon Presentation of an Uncommon Condition

Resham Jayvant Vasani

Abstract
Idiopathic eruptive macular pigmentation (IEMP) is an asymptomatic condition with dark brown, nonconfluent lesions chiefly occurring in children and adolescents. The usual sites involved are face, trunk, extremities, and the lesions resolve over few months to years. We report an unusual presentation of this uncommon condition. A 22-year-old male presented with multiple dark-colored asymptomatic lesions on the scalp and forehead for the past 3 years. There was no history of preceding symptoms, drug, or applications, past or family history of such lesions. Examination revealed multiple hyperpigmented macules and barely elevated nonscale plaques with a velvety feel akin to acanthosis nigricans (AN). Dermoscopy was similar to AN. Blood sugar, thyroid, and lipid profiles were normal. Histology showed “pigmented papillomatosis” concluding the final diagnosis of IEMP. Confluence of lesions is a rarely described phenomenon, with the scalp being a hitherto unreported site. The velvety feel of lesions, the dermoscopic, and histopathological findings further substantiate the hypothesis of this entity being an eruptive variant of AN.

Key Words: Eruptive acanthosis nigricans, idiopathic eruptive macular pigmentation, Pigmented papillomatosis

Introduction
Idiopathic eruptive macular pigmentation (IEMP) is an uncommon pigmentary disorder of an unknown etiology, characterized by the presence of asymptomatic pigmented macules that involve the face, trunk, and proximal extremities in children and adolescents. Herein, we report an unusual presentation of IEMP, with lesions limited to the scalp and forehead with a velvety feel on touch, coupled with dermoscopic and histologic features similar to acanthosis nigricans (AN) reverberating the question of whether IEMP is an eruptive variant of AN.

Case Report
A 22-year-old male presented with multiple dark-colored lesions over the scalp and forehead for the past 3 years. There was an insidious progression of lesions with increase in the number and tendency toward coalescence on the forehead, for the past 3 months which prompted him to seek medical attention. The lesions were essentially asymptomatic and appeared de novo without any preceding event. The patient denied any associated comorbidity or medication. The past, personal, and family histories were not contributory.

Cutaneous examination revealed multiple ill-defined, dark brown to black-colored macules and minimally elevated plaques with a velvety feel on touch. The smaller 0.5 cm sized plaques located on the scalp were discrete [Figure 1a and b] but coalesced to form a larger irregular 3 cm×6 cm plaque extending from the frontal aspect of the scalp to the forehead [Figure 1c and d]. The plaque had a velvety feel. Darier’s sign was negative. Rest of the cutaneous examination was normal. The palms, soles, mucosae, hair, and nails were normal. Systemic examination was unremarkable. On the basis of history and clinical examination, a differential diagnosis of IEMP, urticaria pigmentosa, and lichen planus pigmentosus were considered.

Hematological examination including the complete blood count, serum insulin level, blood sugar and lipid
profile was normal. Dermoscopy of the affected areas showed linear cristae cutis and sulci cutis in addition to an accentuation of the reticular pattern similar to that noticed in AN [Figure 2]. Histopathological examination of hyperpigmented lesion from forehead showed an increase in the basal layer melanization without any increase in number of melanocytes. There was prominent papillomatosis with elongated dermal papillae [Figure 3a]. No significant dermal infiltrate and melanin incontinence were seen, and no interface changes were evident [Figure 3b]. Fontana Masson stain for melanin was positive with negative staining of toluidine blue for mast cells.

The clinical picture of multiple insidiously progressing asymptomatic hyperpigmented minimally raised plaques with the distinct velvety feel coupled with the histopathological finding of pigmented papillomatosis and a normal basement membrane ruled out other differentials such as urticaria pigmentosa, lichen planus pigmentosus, and fixed drug eruption and hence the final diagnosis was IEMP. The patient was counseled about the possibility of spontaneous regression of lesions and was prescribed topical tretinoin 0.025% cream as a night time application on the forehead lesions. The patient had been applying tretinoin for 3 months till reporting with minimal decrease in the thickness of the plaques.

Discussion

Idiopathic eruptive macular hyperpigmentation is a rare entity with less than 60 cases reported worldwide.[2] The term IEMP was first proposed by Degos et al. in 1978 and several cases have been described in French literature.[3]

The etiopathogenesis of this entity is still unclear. All cases reported till date have had a sporadic occurrence.[1] The possible role of hormonal factors in pathogenesis should be considered as lesions appears in puberty and adolescence. In this case, the lesions were noticed at 19 year of age, which could be a possible delay in reporting, on account of the masking of the asymptomatic lesions on the scalp by hair. One report of aggravation of IEMP during pregnancy further supports the hormonal hypothesis.[4]

Sanz de Galdeano et al. proposed the diagnostic criteria. The first criterion was “eruption of brownish black discrete nonconfluent asymptomatic macules involving the neck, trunk and proximal extremities in children and adolescents.”[5] Based on a critical review of the existing literature on the topic, Joshi et al. suggested a modification of this criterion to – “eruption of brownish black discrete, nonconfluent asymptomatic macules, and slightly elevated plaques that resemble AN and involve face, neck, and proximal extremities.”[2] The current patient majorly fulfilled this modified criterion but had lesions on the scalp and the lesions did tend to confluence on the forehead. Lesions on the scalp could have been easily missed in the earlier reports of this disorder because of the masking by hair and confluence of lesions was also mentioned as a feature in the report by de Souza DF et al.[4]
The absence of preceding inflammation and drug intake in this patient rules out postinflammatory and drug-induced causes as fulfillment of the 2nd and 3rd criteria.

The histopathology of this case showing epidermal hypermelanosis with papillomatosis and absence of significant dermal changes reaffirmed the histopathological finding seen in the case series of Indian patients described by Joshi et al. Normal mast cells ruled out urticaria pigmentosa completing the 5th criterion for diagnosis. Clinically, lesions look and feel like AN with dermoscopic picture showing linear cristae cutis and sulci cutis; corresponding to papillomatosis on histology, further substantiate the hypothesis of IEMP being an eruptive variant of AN. A single case described earlier did have mild concomitant AN of bilateral axillae without associated biochemical insulin resistance.

None of the other cases reported till date, nor the one being reported currently had concomitant AN of the other sites or associated obesity. Considering the fact that this condition is commonly seen in adolescence or childhood, associated biochemical hyperinsulinism or malignancy is also thought to be unlikely. Hence, although morphologically and on dermoscopy and histopathology this eruption resembles AN, further studies with larger number of patients and long-term follow-up are needed to further conclusively define this terminology as a variant of AN or eruptive AN as suggested by Joshi et al.

Observing the natural course of IEMP, it is a chronic disorder with spontaneous resolution in months or years. The patient in question had the disease for 3 years and did not show any signs of regression. There exists a single case report outlining the chronic course of IEMP over 21 years with episodic regressions and recurrences.

This case has been reported for its rarity and depicts a hitherto unreported presentation of the disease on the scalp. More dermoscopically and histopathologically backed reports of this entity are needed to know whether this entity represents eruptive AN.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/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.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

**References**
1. Komorowska O, Szczerkowska-Dobosz A, Roziewska D, Czapiewski P. Idiopathic eruptive macular pigmentation – A rare pigmentary disorder. Postepy Dermatol Alergol 2011;28:149-52.
2. Joshi RS, Rohatgi S. Idiopathic eruptive macular pigmentation: A critical review of published literature and suggestions for revision of criteria for diagnosis. Indian J Dermatol Venereol Leprol 2015;81:576-80.
3. Degos R, Civatte J, Bejia S. Idiopathic eruptive macular pigmentation (author’s transl). Ann Dermatol Venereol 1978;105:177-82.
4. de Souza DF, Cunha AC, Piñeiro-Maceira J, Gomes MK, Sodré CT, Ramos-e-Silva M, et al. Idiopathic eruptive macular pigmentation associated with pregnancy. Int J Dermatol 2010;49:810-2.
5. Sanz de Galdeano C, Léauté-Labrèze C, Bioulac-Sage P, Nikolic M, Taïeb A. Idiopathic eruptive macular pigmentation: Report of five patients. Pediatr Dermatol 1996;13:274-7.
6. Joshi R. Idiopathic eruptive macular pigmentation with papillomatosis: Report of nine cases. Indian J Dermatol Venereol Leprol 2007;73:402-5.
7. Subhadarshani S, Singh A, Ramateke PP, Verma KK. Idiopathic eruptive macular pigmentation in an Indian male. Indian Dermatol Online J 2017;8:367-70.
8. Mehta S, Aasi S, Cole R, Chu P, Weinberg JM. Idiopathic eruptive macular pigmentation: A case of 21 years' duration. J Am Acad Dermatol 2003;49:5280-2.