Idiopathic eruptive macular pigmentation with papillomatosis

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ABSTRACT

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We present a case of an otherwise healthy 10-year-old girl who presented with asymptomatic brown macules over the trunk and proximal extremities, of three months' duration. The cutaneous examination revealed multiple, dark brown, discrete, round to oval macules and a few mildly elevated lesions over the trunk and proximal limbs. The individual lesion was 1-3 cm in diameter and a few showed velvety appearance over the surface. Darier's sign was negative. The histopathological study from the velvety lesion showed acanthosis, papillomatosis and increased melanin in the basal layer. The upper dermis showed sparse perivascular infiltrate of lymphocytes without any dermal melanophages. It fulfilled the criteria for idiopathic eruptive macular pigmentation with additional histological finding of papillomatosis.

Key words: Idiopathic eruptive macular pigmentation, papillomatosis

INTRODUCTION

Idiopathic eruptive macular pigmentation (IEMP) is a rare skin disorder characterized by the presence of asymptomatic, brown pigmented macules that involve the face, trunk and proximal extremities in children and adolescents. The first description of this condition was given by Degos *et al.*,^[1] in 1978. These hyperpigmented macules gradually resolve over months or years without any residual pigmentation or scarring. We report a case of a 10-year-old girl who fulfilled all the criteria for this entity. In addition, a few lesions had a velvety surface and showed papillomatosis histologically.



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A 10-year -old girl presented with asymptomatic brown macules over the trunk and proximal extremities, of three months' duration. The lesions first appeared on the trunk and then gradually spread to the extremities. They progressively increased in number and size over a period of one month and became stable. The lesions were not preceded by any other skin lesions. There was no history of drug intake prior to the eruption. The patient was born of a non-consanguineous marriage. The general physical and systemic examination revealed

no abnormality. The cutaneous examination showed multiple dark brown, discrete, round to oval macules and mildly elevated pigmented lesions over the anterior and posterior trunk and proximal extremities sparing the palms and soles [Figures 1 and 2]. The individual lesion was 1-3 cm in diameter and the elevated lesions had a velvety appearance on the surface [Figure 3]. There was no scaling and Darier's sign was negative. The mucosae, hair and nails were normal. The routine blood, urine and stool examinations, liver function, renal function and thyroid function tests revealed no abnormality. Potassium hydroxide mount for fungal hyphae was negative. Biopsy from elevated lesion on back showed acanthosis, moderate papillomatosis and uniformly prominent melanin in the basal layer of the epidermis with normal number of melanocytes [Figure 4]. The upper dermis showed sparse perivascular infiltrate of lymphocytes without any dermal melanophages. Giemsa staining revealed normal mast cell number. The final diagnosis made was IEMP with papillomatosis. The patient was treated with emollients and topical steroids. No new lesions or change in preexisting lesions was seen at six months of follow-up.

DISCUSSION

IEMP is a rare skin disorder characterized by



Figure 1: Dark brown discrete macules over the anterior trunk and proximal extremities



Figure 3: Velvety appearance of pigmented lesions

asymptomatic, brown macules involving the neck, trunk and proximal extremities. Though the first case was reported around



Figure 2: Characteristic macules on back



Figure 4: Acanthosis, papillomatosis and increased melanin in the basal layer (H and E, $\times 45)$

30 years back, the exact etiology and pathogenesis is still not known. Sanz de Galdeno et al.,[2] in 1996 summarized the criteria for the diagnosis of this condition, namely (a) Eruption of brownish-black, discrete, nonconfluent, asymptomatic macules involving the neck, trunk and proximal extremities in children and adolescents, (b) Absence of any preceding inflammatory lesions, (c) No previous drug exposure, (d) Basal layer hyperpigmentation of the epidermis with dermal melanophages without any basal cell damage or lichenoid infiltrate, and (e) Normal mast cell counts. The youngest and oldest case reported in the literature is that of a one-year-old and a 50-year-old.^[3,4] The largest series of ten cases and nine cases have been described by Jang et al.,[3] from Korea and Rajiv Joshi^[5] from India. The differential diagnosis of IEMP includes post-inflammatory hyperpigmentation, fixed drug eruption, urticaria pigmentosa, lichen planus pigmentosus and erythema dyschromicum perstans. IEMP can be differentiated from these conditions by taking proper history and doing skin biopsy study. Histopathologically, IEMP shows acanthosis, basal layer hyperpigmentation of the epidermis with dermal melanophages without any basal cell damage or lichenoid infiltrate and normal mast cell count. However, the histological finding is not really specific.^[1,5] Papillomatosis as a histopathological finding has been only mentioned by Joshi^[5] and Grover.^[6] Joshi^[5] reported presence of dermal melanophages in two out of nine cases. Our case showed papillomatosis but no dermal melanophages in histopathology. Gougerot and Carteaud^[7] in 1932 described several variants of cutaneous papillomatosis. Some authors^[5,8] believe that IEMP may be related nosologically to confluent and reticulate papillomatosis (CRP) and eruptive acanthosis nigricans because both these conditions show histological findings of pigmented papillomatosis similar to our case. However, the clinical features and etiopathogenesis of these conditions are quite different.

It is important to consider IEMP in the differential diagnosis of pigmentary lesions as IEMP is a self-resolving condition. The treatment of this condition is not required as spontaneous resolution of IEMP is expected in a few weeks to few years. In our case, topical steroid was started for the first two weeks to speed up thinning of elevated velvety lesions but without any benefit. To include papillomatosis as one of the diagnostic criteria, more reports of IEMP with papillomatosis are awaited.

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