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Pang, YZ Koh, WL Ang, CC

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Photo vignette

Idiopathic eruptive macular pigmentation with papillomatosis (IEMPP): A controversial entity.

YZ Pang MD MRCP, WL Koh MBBS MRCP, CC Ang MBBS MRCP

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Department of Dermatology, Changi General Hospital, Singapore

Correspondence:

Dr. Yu Zhi Pang

Email address: pangyuzhi@hotmail.com

Address: Department of Dermatology, Changi General Hospital, 2 Simei Street 3, Singapore 52889

Telephone number: +6583661378

Fax number: +6567816202

Abstract

A 19-year-old man with a 6-month history of progressive development of hyperpigmented, velvety plaques on the face and body. A diagnosis of idiopathic eruptive macular pigmentation with papillomatosis (IEMPP) was determined. This entity is discussed.

Keyword: Idiopathic eruptive macular pigmentation with papillomatosis; IEMPP

Case synopsis

A 19 year-old man with Fitzpatrick skin type V presented to our clinic with multiple round dark brown macules and thin plaques over his face and body (Figure 1). Closer examination of the plaques revealed a velvety surface reminiscent of acanthosis nigricans (Figure 2). Darier sign was absent. These lesions gradually appeared over 6 months. The plaques were neither itchy nor tender and were not preceded by erythema and induration. He did not have acanthosis nigricans involving the skin folds. He was otherwise healthy and did not report any intake of oral medications or supplements and he had not applied any topical medicaments prior to the onset of his eruption. He did not recall his family members having a similar eruption. Histopathology from lesional skin revealed basketweave orthokeratosis, irregular acanthosis, and basal keratinocyte hyperpigmentation with a mild perivascular lymphocytic infiltrate in the upper dermis (Figure 3). His fasting glucose and lipid panel tests were normal. His clinical presentation was suggestive of idiopathic eruptive macular pigmentation with papillomatosis (IEMPP), although the histology did not show papillomatosis. Close monitoring over the next 1 year did not reveal any worsening or development of new lesions. The existing lesions remained unchanged.

Figure 1. Multiple brownish macules and thin plaques on the anterior trunk and proximal upper limbs. Note the lack of acanthosis nigricans on the neck, as well as extensive striae alba at the shoulder region from the pubertal growth spurt.



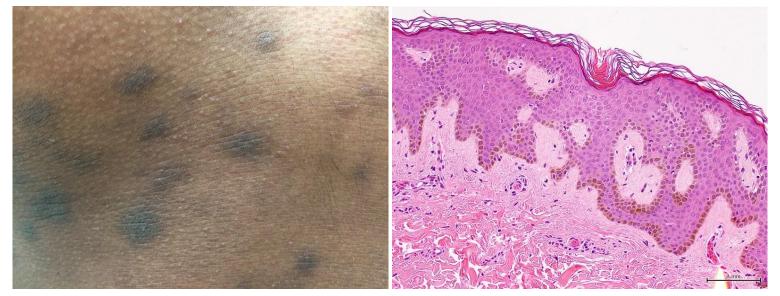


Figure 2. Close up view of the lesions reveal a slightly raised velvety surface with prominence of the skin markings compared to the surrounding skin lesions. **Figure 3.** Histology from the lesional skin (H&E, x 200 magnification) showing basketweave orthokeratosis, irregular acanthosis and basal keratinocyte hyperpigmentation, as well as a mild superficial dermal perivascular lymphocytic infiltrate.

IEMPP is a rare skin disorder characterized by asymptomatic brown macules and plaques with a velvety surface involving the neck, trunk, and proximal extremities, clinically reminiscent of lesions of acanthosis nigricans [1,2,3,4]. Typical histopathologic findings reported include orthokeratosis, acanthosis, papillomatosis, increased pigmentation in the basal layer of the epidermis and a sparse dermal perivascular lymphocytic infiltrate [1,2,3,4]. The etiology and pathogenesis remain unknown. Most cases of IEMPP reported in the literature involved Indian and Asian patients with Fitzpatrick skin type III or greater, with ages reported from 6 to 21 years and involvement of both genders [1,2,3,4]. Most lesions were reported to exhibit spontaneous resolution, some taking up to 5 years [1]. The differential diagnoses to be considered include urticaria pigmentosa, lichen planus pigmentosus, erythema dyschromicum perstans, fixed drug eruption, post inflammatory pigmentation, confluent and reticulate papillomatosis, and tinea vesicolor.

There is much controversy in the literature regarding the nosology of this condition. Most of the reports highlight this entity as a subset of idiopathic eruptive macular pigmentation (IEMP) [1,2,3,4], although there was a suggestion to consider this entity as a variant of acanthosis nigricans [1,2]. Our patient's history and clinical presentation is most suggestive of IEMPP, although the histology failed to show the presence of prominent papillomatosis. This could be attributed to sampling issues, or may point to the fact that IEMPP and IEMP lie within a clinic-pathological spectrum.

Another interesting point is that the majority of cases reported involved young patients of Indian descent (our case, as well as references 1,2,4), suggesting that this condition may have a genetic basis. A database of such rare patients looking at the natural history of the clinical lesions as well as for possible development of frank acanthosis nigricans or metabolic syndrome associated with acanthosis nigricans will be useful to answer some of the nosological issues raised.

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