

A Case of Idiopathic Eruptive Macular Pigmentation Limited to Flexural Areas

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Idiopathic eruptive macular pigmentation is a rare condition characterized by asymptomatic pigmented macules involving the neck, trunk, and proximal portions of the extremities. On histopathologic examination, there was increased pigmentation of the basal layer in otherwise normal epidermis and scattered melanophages in the papillary dermis. We report a case of a 26-year-old woman with idiopathic eruptive macular pigmentation involving only the flexural areas of the body. This condition should be considered in the differential diagnosis of flexural hyperpigmented skin lesions.

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INTRODUCTION

Idiopathic eruptive macular pigmentation (IEMP) is a rare disease, characterized by asymptomatic pigmented macules involving the neck, trunk, and proximal extremities¹. IEMP was first described by Degos, et al². in 1978 and there have been approximately 30 cases reported since³. For the diagnosis of IEMP, Sanz de Galdeano, et al.¹ listed the following identifying characteristics; [1] eruption of brownish, nonconfluent, asymptomatic macules involving the trunk, neck, and proximal portions of the extremities in children and adolescents, [2] absence of preceding inflammatory lesions, [3] no previous drug exposure, [4] basal cell layer hyperpigmentation of the epidermis, [5] prominent dermal melanophages without visible basal layer damage or lichenoid inflammatory infiltrate, and [6] normal mast cell count on the histopathologic finding. Although IEMP is known to involve the neck, trunk,

and proximal extremities, this report describes an unusual case of a 26-year-old Korean woman with IEMP involving only the flexural areas of the body. This condition should be considered in the differential diagnosis of flexural hyperpigmented skin lesions.

CASE REPORT

A 26-year-old Korean woman was evaluated for asymptomatic, hyperpigmented spots on the axillae, antecubital fossae, and inguinal areas that had developed 3 years before. There was no preceding erythema, pruritus, vesicles, nor scaling before the appearance of hyperpigmentation. She had lost 6 kg in 6 months, prior to the development of the skin lesions. However, there was neither previous history of medication, trauma, nor a family history of pigmentary disorders. The laboratory test results including the thyroid function test were normal. Physical examination revealed multiple, brown, nonscaly macules and patches on axillae, popliteal fossae and inguinal areas (Fig. 1). Skin biopsy specimens were obtained from the axillae. On histopathologic examination, there was increased pigmentation of the basal layer in an otherwise normal epidermis and scattered melanophages in the papillary dermis (Fig. 2A). The number of melanocytes in hypopig-

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Fig. 1. Multiple, brown, non-scaly macules and patches. (A) Axilla, (B) Inguinal areas, (C) Popliteal fossa.

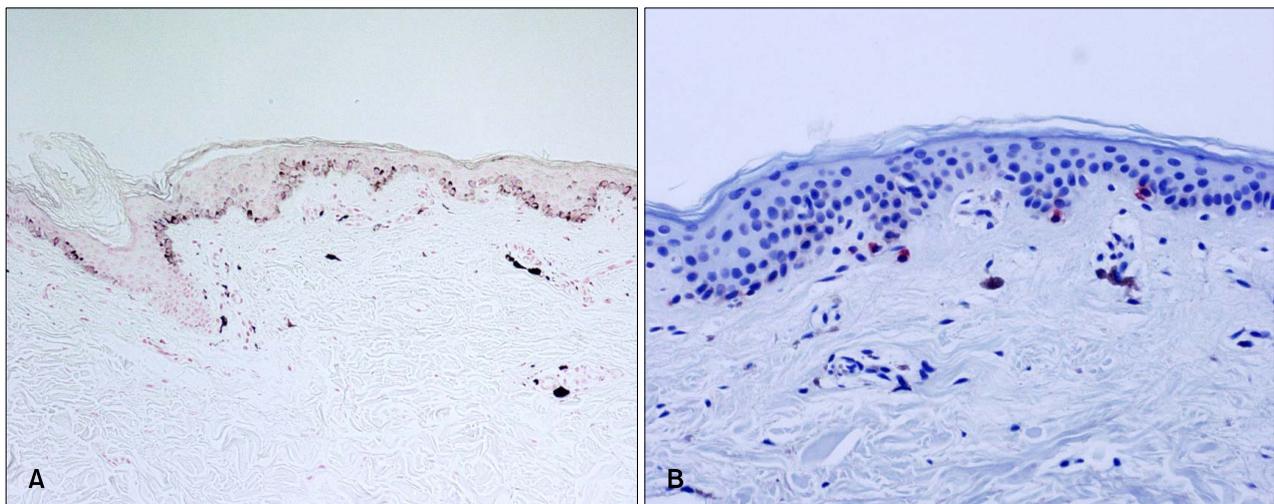


Fig. 2. (A) Basal hyperpigmentation and dermal melanophages (Fontana-Masson staining, $\times 100$), (B) No increase in the number of melanocytes compared to normal lesions (NKI/beteb staining, $\times 200$).

mented skin were not different from perilesional normal skin (Fig. 2B). The patient was diagnosed with IEMP based on clinical and histologic findings.

DISCUSSION

IEMP should be differentiated from erythema dyschromicum perstans (EDP), fixed drug eruptions, friction melanosis or postinflammatory hyperpigmen-

tation⁴. EDP, a common differential diagnosis to IEMP, is characterized by ash-colored macules with a slightly elevated erythematous border. The initial lesions, measuring a few millimetres in diameter, can extend peripherally over months and may form confluent patches. Histology of the inflammatory border shows vacuolar alteration of the basal keratinocytes, colloid bodies and moderate infiltrate of lymphocytes in the papillary dermis mixed with melanophages⁵. EDP can be separated from IEMP

by the different clinical appearance of the macules; grey with an erythematous border and possibly confluent in EDP vs. brownish and nonconfluent in IEMP. The brownish color of IEMP is explained by basal cell layer hyperpigmentation. Vacuolar alterations of the basal layer are seen only in EDP and not in IEMP. The lead-grey color of EDP lesions is reflected by prominent pigmentary incontinence. Friction melanosis is characterized by hyperpigmentation of the skin over the bony regions of the back and limbs occurring after prolonged rubbing with nylon towels and brushes. Histologic examination shows pigmentary incontinence and sporadic deposition of amyloid in the papillary dermis⁶. Thus it can be differentiated from IEMP clinically and histologically.

In our case, pigmentary disorders that involve the flexural areas like Dowling-Degos disease or acanthosis nigricans should also be considered. Dowling-Degos disease shows a reticulate or confluent distribution and has an autosomal dominant mode of inheritance. Pigmented comedone-like lesions, follicular hyperkeratotic papules and perioral pitted acneiform scars are also its features. Thin filiform, pigmented interconnecting epithelial strands are characteristic histopathological findings⁷. Differentiation from acanthosis nigricans was possible because characteristic clinical features in acanthosis nigricans show brown velvety or verrucous plaques. Also, the histopathological findings do not show basal pigmentation but instead show papillomatosis, hyperkeratosis and slight acanthosis⁸. Our patient did not have a family history of pigmentary disorders and the biopsy result was not consistent with Dowling-Degos disease or acanthosis nigricans.

The pathogenesis of IEMP remains unknown. Recently, association with pregnancy and Hashimoto thyroiditis has been reported suggesting the role of hormonal factors and autoimmunity in the etiopathogenesis of IEMP⁹. In our patient, hormonal imbalance due to sudden weight loss could be considered as a possible cause. This could be explained by the fact that hormonal changes had sensitized melanocytes and eventually caused hyperpigmentation^{9,10}. However, the specific reason why the pigmentations were limited to the flexural areas is unknown.

Here, we describe a unique case of IEMP limited to the flexural areas in an adult. IEMP is known to appear on various parts of the body. In previous

cases of IEMP in young children and adolescents, the lesions were usually distributed on the face, trunk and extremities^{1-4,9}. However, there has been no report of IEMP limited to flexural areas. Also, only a few cases have been reported in adults. The alleged rarity of this pigmentary disorder may be because of medical unfamiliarity with this entity, despite its characteristic clinical and histopathologic picture. The knowledge of this entity may be important to avoid unnecessary treatment because spontaneous resolution is to be expected within a period of several months to a few years. This condition should be considered as a differential diagnosis of flexural hyperpigmentation.

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