Case 19

A 7-year-old Thai girl from Lopburi

Chief complaint: Hyperpigmented macules on the neck, trunk and extremities for 2 months

Present illness:

2 month history of asymptomatic hyperpigmented macules, first appeared on the neck then gradually progressed to trunk and proximal extremities. There was no history of preceding erythematous lesion, other skin disorders or medication.

Personal history: She was previously healthy with normal growth and development.

Family history: No family member had similar skin lesions.

Physical examination:

Sharply demarcated, nonscaly hyperpigmented macules involving neck, trunk and proximal extremities. Darier's sign was absent. Mucous membranes, palms and soles were normal.

Histopathology: (S08-9416)

- Mild perivascular inflammatory infiltrate of lymphocytes admixed with some melanophages in the papillary dermis
- Abundant melanin in the epidermis

Diagnosis: Idiopathic eruptive macular pigmentation

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Discussion:

Idiopathic eruptive macular pigmentation (IEMP) is an uncommon disorder of pigmentation characterized by an eruption of asymptomatic brown macules involving the trunk, neck and proximal extremities in children and adolescents. It was first described by Degos et al. in 1978. The pathogenesis and etiology of the disease are still unknown. Some authors believe that hormonal factors may be involved in the pathogenesis because the disease primarily occurs during childhood and adolescent. The age of onset varies from 1 to 20 years. There was a report of 2 cases of IEMP following pityriasis rosea.

According to Sanz de Galdeano et al., the following items are essential for diagnosis of IEMP: (i) an eruption of brownish, nonconfluent macules located at the trunk, neck and proximal extremities in children and young adults; (ii) an absence of preceding inflammatory skin disease; (iii) no previous medication; (iv) basal cell layer hyperpigmentation and occasionally dermal melanophages without visible damage of the basal layer or lichenoid inflammatory infiltrate; and (v) a normal amount of mast cells. Our patient met all defining criteria, so we diagnosed IEMP.

The course of the disease last from months to few years. The greatest time was 21 years duration in a single case. Ashy dermatosis, fixed drug eruption, pityriasis vesicolour, multiple lentigines, postinflammatory hyperpigmentation and mastocytosis must be differentiated. Ashy dermatosis is characterized by ash-coloured macules with an erythema, which can extend peripherally and form confluent patches. Histology of Ashy dermatosis shows...
vacuolar alteration of the basal keratinocytes and an infiltrate of lymphocytes in the papillary dermis mixed with melanophages.

Histopathology of IEMP, there is hyperpigmentation of the basal layer of the epidermis and prominent dermal melanophages. The number of mast cell is normal.

Treatment is unnecessary because spontaneous regression of the lesions can be expected within months to a few years. The recognition of this disease is a prerequisite to avoid nonspecific and time consuming therapeutic approaches and unnecessary expense.

Reference