INCONTINENTIA PIGMENTI ACHROMIANS (Hypomelanosis of Ito)

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A case of incontinentia pigmenti achromians is reported in a 20-year-old female. She had associated hyperkeratosis palmaris et plantaris and a high arched palate. Spontaneous repigmentation developed in some patches after the age of 12 years. Differentiating features of this dermatosis from classical incontinentia pigmenti, achromic nevus, piebaldism and vitiligo are briefly discussed.

Key Words : Incontinentia pigmenti achromians, Ito's hypomelanosis, Systematized hypochromic nevus.

In 1952, Ito described the occurrence of a bilateral systematized depigmented nevus in a 22-year-old woman. He used the term incontinentia pigmenti achromians (IPA) for this disorder because the configuration of the leucoderma appeared as the negative picture of classical incontinentia pigmenti (Bloch-Sulzberger type). Since then, a number of cases have been reported from Japan, Europe and United States.[1] A case of incontinentia pigmenti achromians in a 20-year-old female seen by us is reported.

Case Report

A 20-year-old female nursing student, born to non-consanguinous parents was seen with multiple, asymptomatic hypopigmented patches of various sizes and shapes on the trunk and limbs distributed in an irregular bizzare pattern. She developed these lesions at the age of two and they gradually increased in size and number till the age of twelve, after which some of the patches on the limbs got repigmented spontaneously. There was no history of any vesicular, verrucous or inflammatory changes preceding the loss of pigment in the involved areas. She had three siblings and none in her family suffered from similar disease. General physical and systemic examinations did not reveal any abnormality except for a high arched palate and diffuse hyperkeratosis of the palms and soles.

Dermatologic examination showed multiple and bilateral but asymmetrical hypopigmented and depigmented patches of various sizes and shapes on the buttocks, trunk and limbs. Hypopigmented discontinuous bands and streaks on the upper limbs extended from the shoulder to the wrist on each side. On the legs, the lesions extended from the knees to the feet. Multiple depigmented patches and spotty macules were seen distributed irregularly in splashes and in whorls on the trunk and some of them crossed the midline of the body. A large, irregular, bizzare, depigmented patch was noted on the left side of her buttocks. A few hyperpigmented macules which developed since 6-7 years were also seen among the depigmented patches on the arms and buttock. There was no atrophy and the sensations and sweat responses in the patches were normal. Hairs and nails also appeared normal.

Routine laboratory investigations on blood and urine did not show any abnormality. Histopathology of the skin lesion taken from the leg revealed slight thinning of the epidermis. There was no incontinence of melanin and the basal cell layer was intact. Dermis also appeared normal. A dopa staining could not be done.

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Comments

The term incontinentia pigmenti achromians was coined for this rare dermatosis by Ito because the pattern of hypopigmentation in these patches resembled the pattern of hyperpigmentation seen in incontinentia pigmenti (Bloch-Sulzberger) and it affected predominantly the females. It begins early in life without any preceding inflammatory changes or bullae and persists for many years. A variety of ectodermal and mesodermal anomalies have been reported in association with IPA. Among the 21 cases reviewed by Jelenik et al,[2] ten had associated non-cutaneous anomalies including anomalies of the eyes, musculo-skeletal system, CNS, blood vessels, sweat glands, hair, breasts and teeth. Verrucous epidermal nevi, thick lips and congenital hydronephrosis were also noted. Mittal et al[3] reported a case in a girl who developed both incontinentia pigmenti and IPA before the age 2 with nail dystrophy, delayed dentition and epilepsy. The case reported by Maize et al[1] was a 9-year-old mentally retarded girl. She had a high-arched palate and the teeth were irregularly placed. Histologically, the term IPA is a misnomer since there is no basal cell destruction and incontinence of melanin pigment into the dermis. IPA has been satisfactorily differentiated from piebaldism and depigmented nevi, in both of which the occurrence of depigmented areas is noted at birth, the size of the patches is constant and the lesions continue throughout life.[4] A white fore-lock is present in 80-90% of the patients with piebaldism.[5] Bilateral systematized depigmented nevi would be difficult to be differentiated from IPA. Most authors now prefer to consider IPA as a variant of systematized achromic nevus even as Ito did 32 years ago in describing the original case. Bleehen and Ebling[6] also described IPA under the title 'nevus depigmentosus.' Histologically also, IPA seems more closely related to systematized achromic nevi than the classical incontinentia pigmenti.[1] But some consider IPA as a separate entity because the onset of this dermatosis is usually during childhood rather than at birth and the lesions tend to repigment as age advances. This does not happen usually in achromic nevi. Though there is lack of incontinence of melanin pigment histologically in IPA, there are certain features which suggest a clinical relationship between IPA and incontinentia pigmenti. Both are inherited diseases and are associated with other ectodermal defects and show whorled patterns of pigmentary changes. Hyperkeratosis palmaris et plantaris and a high-arched palate were the additional abnormalities noted in the present case. There is no effective treatment for IPA. Eventual repigmentation is the rule.[7] This was quite evident in our case also who developed spontaneous repigmentation in some of the patches, after the age of 12 years.

References

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