INCONTINENTIA PIGMENTI

We have recently seen two cases of incontinentia pigmenti. The first case, a three-month-old female child, born to non-consanguinous parents presented with vesicular lesions in linear streaks and bizzare patterns on the trunk since birth. The lower lip and genitalia also were affected. A detailed physical and systemic examination did not reveal any abnormality. Blister fluid from a skin lesion as well as lip lesion showed plenty of cosinophils. Histopathology of a skin lesion showed intra-epidermal blisters filled with cosinophils. One year follow up of the child showed that the vesicles were replaced by verrucous lesions and later by hyperpigmented linear patches.

The second case, a one-year-old boy born to non-consanguinous parents, presented with bizzare whorls and splashes of slate-blue-black, discrete and confluent macules and patches on the lower limbs and trunk since birth. There was no history of preceding vesicular skin lesions. A detailed examination showed no abnormality except a delayed dentition. Only the lower central incisors had erupted. X-ray chest and ECG showed no abnormality. Parents denied consent for biopsy of the skin lesion. Two months after discharge from the hospital, the child died suddenly at home.

Incontinentia pigmenti is a rare genodermatosis. Only a few cases have been reported from India. Our first case presented in the early vesicular stage, whereas the second case showed features of the late pigmentary stage of the disease. Though conjunctival lesions have been reported before, we have not come across any report of involvement of the lips in this disease. Males are only rarely affected by this dermatosis. Our second case was a male. He had delayed dentition also. Occurrence of early death in this child, with the least severe form of skin lesions, once again demonstrates the lethal nature of this disease in males even though the cause and nature of death in this child could not be ascertained.

Dr. K. Pavithran,
Dr. P. Ramachandran Nair; and
Dr. J. Zachariah.
Department of Dermatology and Venereology, Medical College, Kottayam, India.