LETTERS TO THE EDITOR

PIGMENTARY STAGE OF INCONTINENTIA PIGMENTI: IS IT A DISTINCT ENTITY ?

To the Editor,

A question of diagnosis of incontinentia pigmenti (IP) sometimes arises when strange pigmented dermatoses occur in older children, teenagers and even in adults. Most of these patients may not give past history of having any vesiculo-bullous lesions or warty lesions. Histopathology of lesions shows mostly incontinence of pigments in the upper dermis within melanophages.

IP is an X-linked dominant disease, lethal to males. Vesiculo-bullous and warty stages of IP are followed weeks to months later by the pigmentary stage. In 96.4% of the cases, all these three stages are manifested by 6 weeks of age.[1]

As many as 40% of the patients with IP manifest with pigmentary stage without preceding inflammatory stages.[2] There is no proof for the hypothesis that patients who manifest only pigmentary stage either at birth or later had erythema, vesicles or warty lesions in utero. Pigmentation is more common on trunk,while inflammatory stages are present on limbs. It seems reasonable that inflammatory stages may result in post inflammatory hyperpigmentation. However, it is difficult to believe on that basis alone that the pigmentation should last for long time rather than several months, as in the usual case with post inflammatory hyperpigmentation. The noninflammatory pigmentation is more common on trunk and has characteristic whorls and streeks pattern. It tends to last, atleast in part, for a number of years. The reports which appeared before 1995 postulated that melanin would have travelled via perivascular spaces or lymphatics to the distant sites. However, these have not been confirmed subsequently.

Carney[1] and other authors suggested that this noninflammatory pigmentation must represent some sort of naevoid anomaly, the pathogenesis of which is as yet unclear. This suggestion was sidelined by some authors using electron-micrographic evidences. They demonstrated phagocytosis of melanin by dermal macrophages[4] and the presence of dyskeratotic keratinocytes in the epidermis during all three stages of the disease.[5] However, these electron micrographic findings need not be specific for IP. They are also seen in lichen planus, fixed drug eruption and lupus erythematosus. On light microscopy, various conditions with incontinence of pigment can be differentiated, so also the three stages of IP.

Are we justified in making the diagnosis of IP when adults present with bizarre hyperpigmented lesions in trunk and extremities ?

Narahari SR
Kasaragod

References

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