INCONTINENTIA PIGMENTI

RATAN SINGH* & DEVINDER KAUR**

Incontinentia pigmenti is a rare, complex, hereditory syndrome, in which vesicular, verrucous and pigmented cutaneous lesions are often associated with developmental defects of eye, teeth, skeletal and central nervous systems. It was fully described by Sulzberger in 1927, but his case had been previously presented by Bloch in 1925. Siemens in 1929 described it as "Melanosis corridegenerativa". The earlier case reports were limited to description of pigmentary phase and associated developmental abnormalities. It remained for Carney in 1951 to describe first, the initial vesicular state, which of ten preceeds the better known bizzare pigmentary whorls.

Franceschetti and Jadassohn (1954) and Gramer & Schmidt (1955) have reviewed and described the clinical features of Incontinentia pigmenti. Skin changes are often present at birth, have usually developed before the end of first week and rarely after first two months. Inflammatory, clear, tense bullae often grouped in linear pattern develop on the limbs in recurrent crops; crops continue for a few days or for a month or two. The vesicular stage is often associated with eosniphilia. After some time, linear warty lesions may appear on the backs of hands and feet particularly the fingers and toes and eventually the typical pigmentation. This sequence may not be regular, and whorls of pigmentation do not necessarily occur on the sites of early vesicular lesions. The pigmentation ranging in colour from blue-grey or slate to brown, is characteristic of the syndrome and the bizzare "splashed" or "Chinese figure" distribution is diagnostic.

Histologically the vesicles of the first stage are intraepidermal in location and contain numerous eosinophils. The dermis beneath the vesicles show an infilterate and that also contains many eosinophils and lymphocytes. In the second or verrucous stage there is hyperkeratosis and acanthosis with non-specific inflammatory infilterate. The areas of pigmentation show extensive deposits of melanin inside and outside the melanophores in the upper dermis. In some cases melanin in the basal layer is diminished and the basal layer shows degeneration and vaculisation of the cells (Lever, 1967).

The aetiology of the syndrome is not precisely known. The familial incidence of incontinentia pigmenti and its almost exclusive occurance in females has been reviewed by Curth and Warburton (1965). Genetic evidence is incomplete but suggests that the syndrome is determined either by an autosomal dominant gene usually lethal in male, for almost all cases are females (Pfeiffer, 1960) or by a sex linked gene carried on X-chromosome (Lenz, 1961). Viral infec ion of the mother during pregnancy has also been suggested (Haber, 1952).

*Professor & Head of the Department, **Registrar; Department of Dermatology & Venereology, Mauiana Azad Medical College & associated Irwin & G. B. Pant Hospitals New Delhi-1.

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Most of the cases reported are from Europe, Japan and a few from America. British literature did not record this condition till 1948, when Haber, showed the first case of incontinentia pigmenti at the Royal Society of Medicine. Only two cases of incontinentia pigmenti have been reported from the Indian subcontinent by Lahiri (1955) and Marquis et al (1969). A case of Incontinentia pigmenti seen in the Skin & V.D. O.P.D. of Irwin Hospital, New Delhi, is reported here.

Case Report

A 12 days' old female infant was brought to the Skin O. P. D. of the Irwin Hospital on 20-1-69 with chief complaint of vesicular lesions over the right upper and lower limbs and lower part of abdomen since birth.

She is the fifth child of the family and the outcome of sixth pregnancy of the mother. It was a twin pregnancy and the other child was still born and sex was male. The fifth pregnancy of the mother ended in miscarriage; again it was a male child. There was no history of mother having suffered from any viral disease during the entire period of gestation. The other siblings consisting of one brother and three sisters, all are normal.

On examination she presented grouped vesicles arranged in a linear fashion on erythematous base on the anterior aspects of right lower and upper limbs extending upto dorsum of right foot and right hand and lower part of the abdomen. Impetiginisation was present in the right groin. Along with these, hyperpigmented fine maculo-papular lesions were present in linear, grouped and discrete fasion at the same sites. There were no verrucous lesions. Otherwise the child was healthy and systemic examination did not reveal any other finding.

Her hemoglobin was 11 gm%. Total leucocytic count was 4700/cum; with Polymorphs 70%, Lymphocytes 25% and Eosinophils 5%. Sedimentation rate was 12mm/1st hour. Mother's S. T. S. was negative. Culture of fluid from the unruptured vesicles was sterile. X-ray examination of the skeltal system showed no abnoramality.

Biopsy was taken form two different sites. The vesicular lesion, showed intra-epidermal bullae with marked spongiosis. Most of the bullae contained a large number of eosinophils. Large epithelial cells with acidophilic cells were seen in small numbers around the bullae. The pigmented maculo-papular lesions showed hyperkeratosis with acanthosis; superficial dermis had extensive deposits of melanin both inside and outside melanophores. Basal layer was oedematous at places.

Treatment given was only in the form of saline compresses and lotio gention violet 1% to the inflammed vesicular lesions. On the next visit, after 14 days the child showed verrucous lesions at the sites of initial vesiculations, which had almost disappeared. Along with the verrucous lesions, pigmented macules, in streaks and patches had appeared. This time the child was given a bland

ointment. The child was last seen in May, 1969. The verrucous lesions had very much regressed and the pigmentation was coming up more on the trunk. Otherwise the child was normal.

Comments

This case which confirms both by clinical and histological features the diagnosis of incontinentia pigmenti, is the Third one to our knowledge to be reported from India. The first case reported by Lahiri (1965) had only characteristic pigmentary features and no histopathological study could be done. In this case the various stages of incontinentia pigmenti, were very classically seen during the short span of five months of infant's life. Initial inflammatory vesicular lesions were replaced by verrucous ones in 2-3 weeks time. At the age of 4 months, these verrucous lesions were also seen regressing and characteristic pigmentation was seen coming up, particularly on the trunk. So far no other ectodermal or mesodermal abnormalities have been observed. A prolonged follow up of the case is planned.

The sex of both the dead foetuses in fifth and sixth pregnancies of the mother was found to be male. This may support Pfeiffer's (1960) concept that incontinentia pigmenti is caused by an autosomal dominant gene which is lethal in male and hence seen predominantly in female children.

Simple remedies like saline compresses and lotio gention violet 1% were enough to control the impetiginized inflammatory lesions. Corticosteroids and Vit 'C' were not needed.

Japan has reported 21 cases of incontinentia pigmenti (Kitamura et al, 1954) and together with Lahiri (1955) & Marquis (1969) and this case, it makes a total of 24 cases from Asia.

Summary

The third case of incontinentia pigmenti from the Indian subcontinent is reported making a total of 24 cases from Asia.

Various stages of incontinentia pigmenti have been observed very typically in this case and were confirmed by histopathological study.

Literature on incontinentia pigmenti has been briefly reviewed and its etiology discussed.

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