Ind J Dermatol Venereol Leprol 1995; 61 : 295-6

INCONTINENTIA PIGMENTI

S Purohit, M K Singhi, R Khullar, G Kalla

Three cases of incontinentia pigmenti are reported. All the patients were female with bizarre pigmentation and verrucous and nodular lesions on the body. In all the cases there was absence of the vesicular stage and systemic involvement.

Key Word : Incontinentia pigmenti

Introduction

Incontinentia pigmenti is a rare genetic dermatosis. It is inherited by X-linked dominant gene and is lethal in male.[1] Classically it menifests as recurrent linear vesiculo-bullous lesions that are accompanied or followed by nodular and verrcous lesions. These are followed by peculiar bizarre pigmentation which ranges in colour from blue-grey and slatey to brown. The pigmentation lasts for years and may fade slowly.[2]

Very few cases of incontimentia pigments have been reported from India.[3,4] We are reporting three cases of incontinentia pigmenti seen in one year.

Case Reports

Case I : A 3-month-old female child presented with bizarre brown pigmentation and few warty and nodular lesions on the trunk and limbs. She was the first conceived child and was born asymptomatic at full term. On third day she developed bluish-grey pigmented, irregularly linear macular lesions, first on legs and then on trunk and upper limbs. The lesions gradually became brown in colour and a bizarre splashed distribution of pigmentation developed. Two weeks later recurrent linear reddish brown nodular and warty lesions developed on the pigmented areas of the limbs.

Case II : A 45-day-old baby girl was brought with nodules, warty lesions and bizarre pigmentation on the body. At birth smooth red nodules were present in an irregular linear and spiral pattern, on the upper and lower limbs. Later some of them became warty. Gradually a brownish-grey pigmentation developed in a bizarre pattern over both limbs and the trunk. This was a premature birth of 6 months gestation, of a male child who died immediately after birth.

Case III : A 4-month-old female child presented with bluish red pigmentation in a bizarre pattern with bluish red pigmentation in a bizarre pattern and papulo-nodular lesions on the body. The child was asymptomatic at birth. On the sixth day, the parents noted the development of reddish pigmentation in a linear bizarre pattern, first on limbs and then on the trunk. Later papulo-nodular lesions developed, mainly on the pigmented areas. Some of them developed a warty surface. The pigmentation gradually changed in colour to bluish red. This was their third conceived child. The parents had one female child aged 5 years. The second conception ended in abortion of 3 months.

Systemic examination of none of the three cases revealed any abnormality. The

From the Department of Skin, STD and Leprosy, Dr Sampurnanand Medical College, Jodhpur-342003, India.

Address correspondence to : Dr M K Singh, Q No. III/2 Mathura Dass Mathur Hospital Campus, Jodhpur - 342003

developmental milestones were normal for their age in all the cases. The investigations were within normal limits except in case 1 in which eosinophilia of 30% was seen. VDRL test of the parents and the cases was non-reactive. Skin biopsies confimed the diagnosis in all the cases. The parents of all the cases. The parents of all the cases were normal. There was no history of consanguinity in any of the parents.

Discussion

Incontinential pigmenti is a rare disorder. Only a few case have been reported from India.[3,4] The vesicular stage was lacking in all the three cases reported here. Only in one of the three cases, the lesions were present since birth. None of the parents had a live male child. One of parents had a prematurely born male child who died immediately after birth. In another parent, there was abortion of 3 months duration, which could be a male foetus.

References

1. Wiklund DA, William L, Weston MD. Incontinentia Pigment : A four generation study. Arch Dermatol 1980; 116: 701-3.
2. Mosher DB, fitzpatrick TB, Ortome JP, et al. Disorders of melanocytes. In : Dermatology in General medicine (Fitzpat rick TB, Eisen AZ, Wolff K, et al eds). 3rd edn. New York. McGraw-Hill, 1987: 856-8.
3. Jain VK, Kalla G, Bumb RA. Incontinentia Pigmenti. Ind J Dermatol Venereol Leprol 1992; 58: 39-40.
4. Ramalakshmi M, Parthasardhi A. Incontinentia Pigmenti. Ind J Dermatol Venereol Leprol 1987; 53: 244-5.