JUVENILE HYALINE FIBRAMATOSIS

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A 1½-year old female baby had pearly papules and crusting over the nasolabial folds, earlobes, nape of the neck and scalp. She had gingival hypertrophy and showed osteolysis of tibiae on roentgenography. Biopsy was characteristic of juvenile hyaline fibramatosis.

Key Words : Juvenile hyaline fibramatois

Introduction

Juvenile hyaline fibramatosis (JHF) is a rare form of fibromatous proliferation occurring in children. Iwata et al[1] suggested that it is a connective tissue disease characterised by increased synthesis of glycosaminoglycans by fibroblasts. We report one of the youngest case of JHF.

Case Report

A female child of 1½ years age, born to non-consanguineous parents reported for symmetrical skin lesions of 1 year duration over the earlobes, nape of neck and scaly crusted lesions over the sides of the nose and nasolabial folds.

On examination, she had plenty of closely set papules and few nodules over both earlobes and especially over the margins of pinnae (Fig 1). She also had a few papules and a small plaque over the nape of neck and occipital region. A solitary nodule was present over the tip of left index finger. Minimal hypertrophy of gingiva was present.

Along the nasolabial folds in-addition to the skin coloured papules, she had crusted lesions mimicking seborrhoeic dermatitis (Fig 2) which showed good remission with topical corticosteroid cream. Her physical and mental development were normal. She had no flexural deformities or systemic abnormalities.

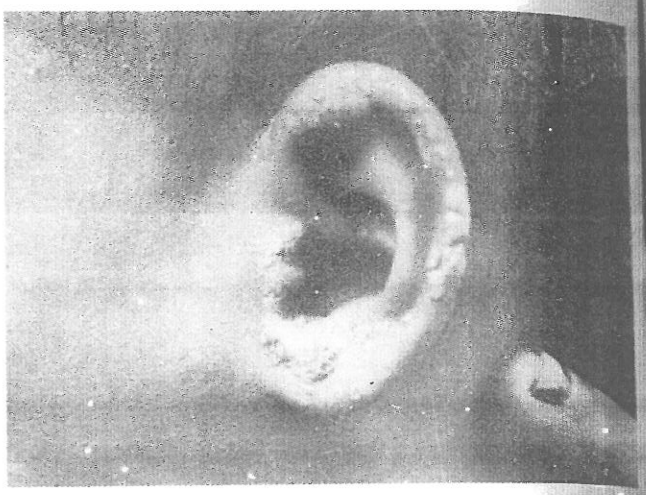

Fig. 1. Papules and crusting over the earlobe.

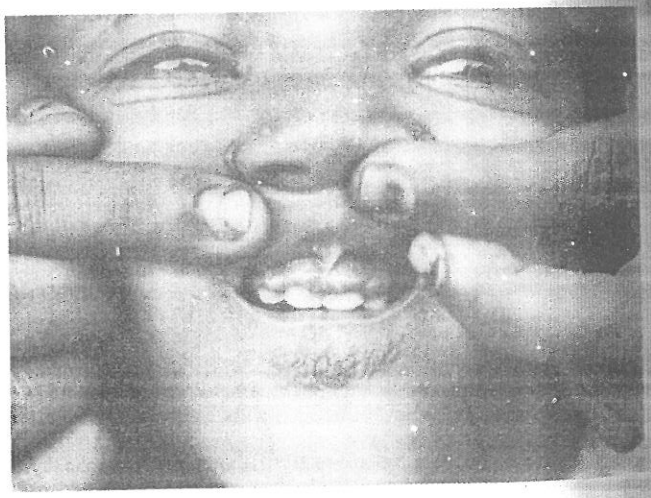

Fig. 2. Papules and crusting seen over the nasolabial folds and chin with gingival hypertrophy.

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Routine investigation and skeletal survey showed bilateral, symmetrical osteolytic lesions in the proximal ends of both tibiae. Histopathological examination of the papule showed homogenous eosinophilic band of amorphous substance in the dermis which was strongly PAS positive and diastase resistant (Fig 3).

Discussion

Quintal, et al[2] described the clinical picture of JHF presenting as 3 types of lesions.

1. Small fleshy pearly papules in nasolabial folds, mastoid area and nose.
2. Translucent nodules on pulp of fingers, external portion of the ears and,
3. Large subcutaneous tumors over the scalp, trunk and extremities.

In addition they have gingival enlargement, osteolytic lesions and joint contracture. Lesions start early in life between 3 months to 4th year. Along with the clinical picture, the presence of PAS positive amorphous hyaline material in the dermis enables the confirmation of diagnosis.

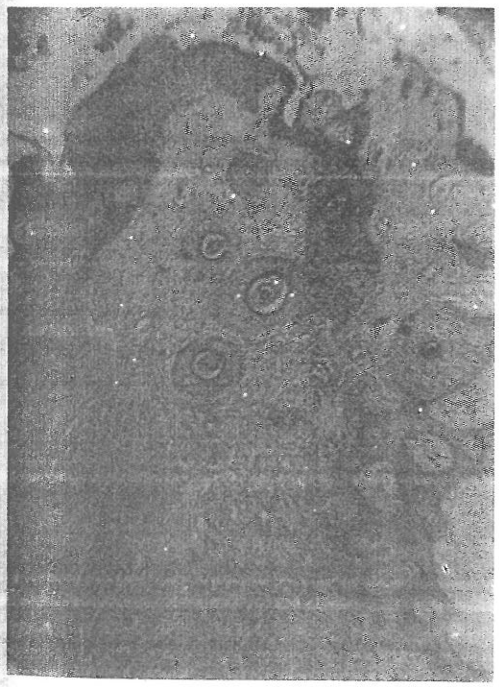

Fig. 3. Biopsy specimen showing homogenous amorphous material in the fibrous dermis (H&E, x 50).

The presence of small pearly papules over both ear lobes, nasolabial folds and the occipital region, few nodules over the scalp along with gingival hypertrophy and osteolytic lesions over the upper ends of tibiae in our case commencing as early as from the 6th month suggested the possibility of JHF. There were no large tumors over the scalp. However, the contracture of the joints reported in the litreature were not seen, as it be too early for contractures to develop. The histopathology of the papule form the nape of the neck was characteristic.

The longest surviving case reported is about 51 years, the onset of the lesion being from the 6th year.[3] The youngest case reported was at the age of 18 months by Finlay, et al in 1983.[4] Our patient is most probably the youngest child with the commencement of lesions from the age of 6 months to be reported. The first case reported form India by Yesudian, et al[5] was a 2 year old child. Another interesting feature was the close similarity of the lesions over the nose and earlobes covered with yellowish crusts to histiocytosis X. But biopsy of the papule and skeletal survey clinched the diagnosis.

Our case also showed bilateral symmertical osteolytic lesions in the proximal ends of both tibiae similar to the case reported by Finlay et al.[4]

In the paediateric age group its resemblance to Letterer-Siwe disease is worth nothing.

References

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