Images in Clinical Practice

Langerhans Cell Histiocytosis

A 10-month-old male child presented with fever, ear discharge, swellings in the neck since 2 months of age. The child had a skin rash initially on scalp, gradually progressing to involve the trunk and then palms and soles. Over the scalp the skin rashes were scaly and crusty (*Fig.1*). There were discrete hypopigmented macules over the face, trunk (*Fig. 2*), and soles; a few papules were hemorrhagic.

The infant also had blepharitis, bilateral ear discharge, alopecia and oral thrush. There was cervical and occpital lymphadenopathy with a 4 cm hepatomegaly. The skin biopsy and FNAC of lymph node revealed langerhans cells with S 100 positive. The bone marrow and skeletal survey were normal.

Langerhan cell histiocytosis (LCH) is a monoclonal disorder leading to proliferation of cells of the mononuclear phagocyte system and the dendritic cell system. The criteria for definitive diagnosis of LCH is demonstration of Birbeck granules in the cytoplasm of langerhan cells by electron microscopy or CDI antigenic determinants on the surface of lesional cells by immunocytology or immunohistology.

The presumptive diagnosis of LCH can be made when the lesion has characteristic morphology and phenotype to an experienced pathologist and the cells express S100 and at least one of the following ATPase, α-mannosidase and peanut lectin positive in the lesional cells.

The differential diagnosis of the skin lesions to be considered are eczema, miliaria, scabies,

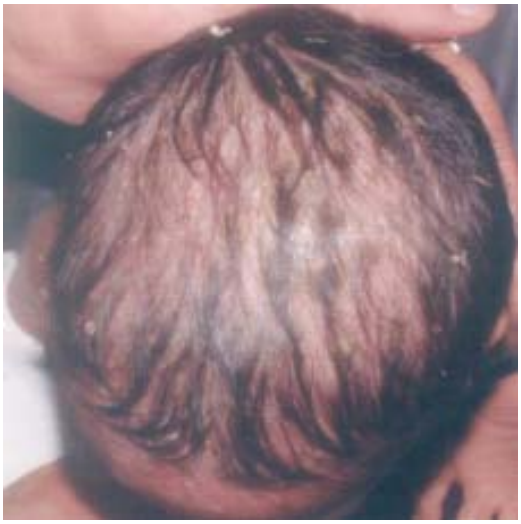

Fig. 1. Sclap showing scaly and crusty rashes.

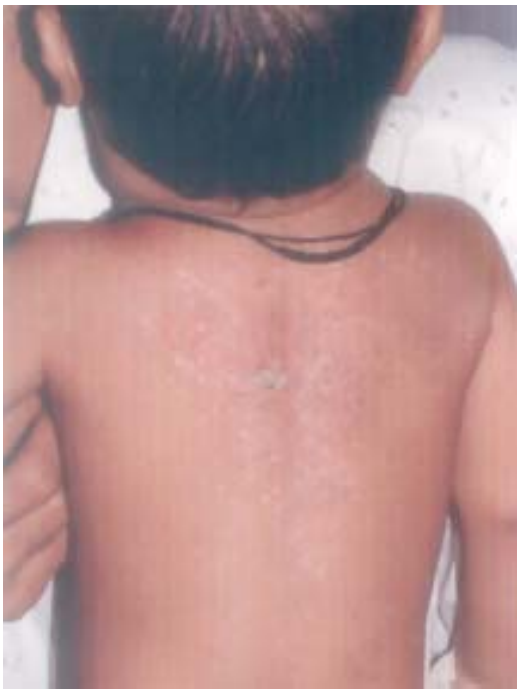

Fig. 2. Discrete hypopigmented macules.

varicella and non-specific dermatitis. In Dariers disease papules are hyperkeratotic. Histopathological examination will differentiate some lesions of juvenile xantho-granuloma and xanthoma disseminatum.

LCH commonly involves the skull, skin and mucous membrane, lungs, liver spleen and lymphnodes, involvement of the pituitary with diabetes insipidus and growth retardation may be seen in 15-20%.

The treatment includes giving chemotherapy using drugs like vinblastin, etoposide and prednisolone given for 6 months in low risk and for high risk methotrexate and 6-Mercaptopurine is added and given for 12 months. Surgical curettage may be done in solitary lesions. Radiotherapy may be given to non-healing skeletal lesions.

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