Langerhans Cell Histiocytosis with Characteristic Skin Involvement

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Langerhans cell histiocytosis (LCH) is a reactive condition in which Langerhans cells accumulate in various tissues and cause damage due, in part, to cytokine production. The etiology has varied from tuberculosis, viruses, lipid abnormality, and immunological abnormality(l). LCH may involve bone, skin, lymph nodes, liver and spleen with hematologic changes and cause fever, malaise and failure to thrive. The skin lesions are characteristically erythematous with greasy scales, scaly papules, nodular and crusted erosions(2). A presumptive diagnosis is possible when histological appearance of biopsy is consistent with diagnosis of LCH. Marker studies involving S100 protein, peanut agglutinin, expression of CD1 complex and electron microscopic demonstration of Birbeck granules further confirm the diagnosis(2,3).

Case Report

A $1^{1/2}$-year-old boy was brought to us with rashes over scalp and trunk of four months duration. Examination of the skin showed diffuse erythema with greasy scales involving scalp and multiple, discrete, scaly red-brown purpuric papules invoving predominantly the seborrheic areas of the trunk *(Fig. 1)*. The child was irritable and had marked anemia, thrombocytopenia, bilateral cervical and axillary lymphadenopathy with mild hepatosplenomegaly. His peripheral blood film showed no abnormal cells, swabs from skin lesions were negative for micro-organisms and pus cells. X-ray studies for skull, long bones and chest were normal. The morphology and distribution of the characteristic skin lesions led us to consider a clinical diagnosis of LCH. The diagnosis

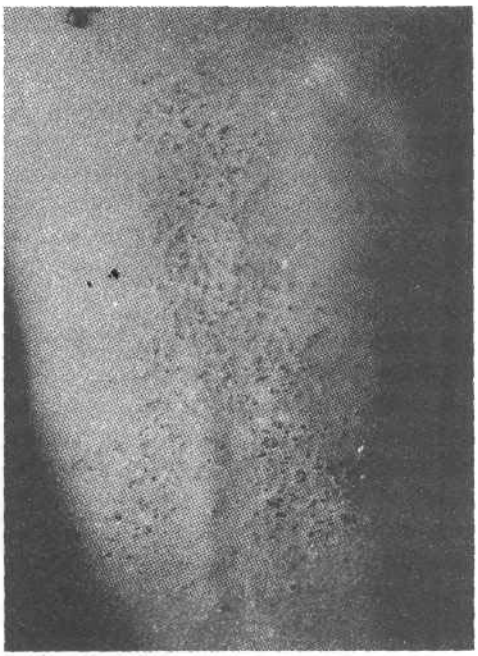

Fig. 1. Photograph of the skin lesions on back showing multiple red-brown, purpuric, scaly papules.

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was substantiated by histopathological examination of the skin. It showed upper dermal and junctional dense histiocytic cell accumulation having homogeneous eosinophilic abundant cytoplasm, with lobulated bean and boat shaped and folded nuclei mostly placed eccentrically. There was no phagocytosis. These cells showed faint positivity with PAS stain, a variable lymphocytic cellular aggregate was discernible in dermis *(Fig. 2)*. The lymph node sections revealed deposits of similar histiocytic cells in the paracortical zones. A presumptive diagnosis of LCH was established on the basis of characteristic histopathological appearances in skin and lymph node. The child was put on oral prednisolone in the dose of 2 mg/kg/day. After 2 weeks a pulse of IV cyclophosphamide 100 mg and vincristine 0.5 mg was given. One month after commoncement of therapy, there was marked improvement in general condition. His appetite improved. The size of lymph nodes, liver and spleen decreased markedly and cutaneous lesions regressed by more than 50%. In between he received a unit of blood and supportive therapy in the form of antibiotics, zinc and B complex. Because of a death in child's family his father got him discharged. After that he failed to turn up for followup and we were informed that he died 3 months after discharge.

Discussion

LCH can affect many organs. The exact incidence is not known because of the heterogeneity of the clinical expression of disease. In a series of 58 patients with LCH, 14 had single system disease (13 bone and 1 skin lesion) and 44 had multisystem disease of whom 50% had vital organ dysfunction(3). Another study showed common bone, lymph node and skin lesions with 50% showing liver disease and 23% lung

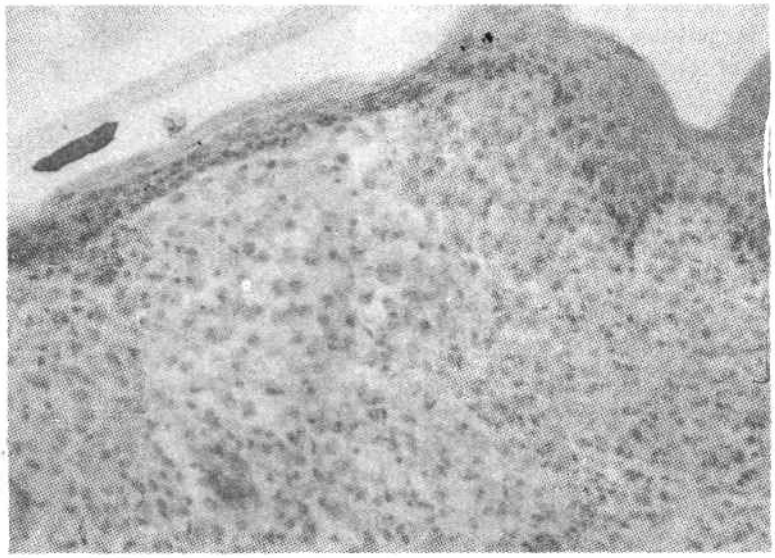

Fig. 2. Photomicrograph showing dense accumulation of large histiocytic cells in the papillary dermis with lymphoid cellularity (H & E x 80).

disease with frequent hematological changes(2). There is a report in Indian literature of a case of congenital histiocytosis with skin and lymph node involvement. In another report of Letterer Siwe disease in an infant with acute presentation, hematalogical abnormalities with hepatosplenomegaly without lymphadenopathy or bone disease were reported(6). The role of skin scrapping in the diagnosis of Letterer-Siwe disease has also been highlighted(7). The present case presented with characteristic cutaneous lesions, lymphadenopathy with hepatosplenomegaly without bone involvement. The skin and lymph node biopsy confirmed the diagonsis and the case was treated with oral prednisolone, vincristine and cyclophosphamide alongwith supportive therapy.

Histopathological diagnosis is most important, as many diseases clinically mimic LCH. S-100 staining or CD1 staining is insufficient to establish a diagnosis of LCH if the histological picture is not consistant. The differential diagnosis in cutaneous LCH includes seborrheic dermatitis, juvenile xanthogranuloma, xanthoma disseminatum and benign cephalic, histiocytosis. In this brief communication we wish to emphasize the important role of lesional biopsy in child with scaly purpuric papules on scalp and trunk with fever, malaise, failure to thrive and stress the rarity of characteristic cutaneous manifestations in LCH.

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