Langerhans cell histiocytosis with bilateral parotiditis

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Dear editor,

A 9-month-old male child presented with itching skin rashes, bilateral ear pain and unilateral ear discharge, bilateral swelling in the mandibular angle, excessive water drinking and thirst since 6 months of age. Review of systems showed neither clues indicating of multi-system involvement nor constitutional symptoms. The parents denied any history of fever. There were no further complaints indicative of possible gastrointestinal (GI) and bone involvement. On physical examination, the patient was well developed, well nourished, and in no acute distress. He was afebrile, and his vital signs were within normal limits. Skin lesions were crusted erythematous papules over the scalp, face, ears, around the oral orifice, neck, axilla and inguinal region (Figure 1-3). Alopecia was also present at areas of scalp lesions (Figure 1). Bilaterally enlarged firm parotid glands and enlarged palpable cervical lymph nodes were detected. No hepatosplenomegaly was present. Routine laboratory findings were within normal limits except for a high erythrocyte sedimentation rate. Parotid ultrasonography was performed and bilateral chronic parotiditis (sustained for nearly 3 months) was confirmed by otolaryngologists. Mumps and other infectious causes of parotiditis were ruled out by pediatricians. In plain radiographic studies, no bony lesions were detected. A few histiocytes were seen on bone marrow aspiration and biopsy.

Biopsy of the skin lesions was performed in which aggregations of large clear histiocytic cells with enlarged lobulated nuclei and eosinophilic cytoplasms were detected. Invasion by these atypical histiocytes was also noticeable. The diagnosis of Langerhans cell histiocytosis was confirmed by immunohistochemistry study which showed positive results for CD1a glycoprotein (Figure 4).

Langerhans Cell Histiocytosis (LCH), previously known as histiocytosis X, is a rare proliferative disorder in which the accumulation of pathologic Langerhans cells leads to local tissue infiltration and destruction [1]. Langerhans cell histiocytosis mainly

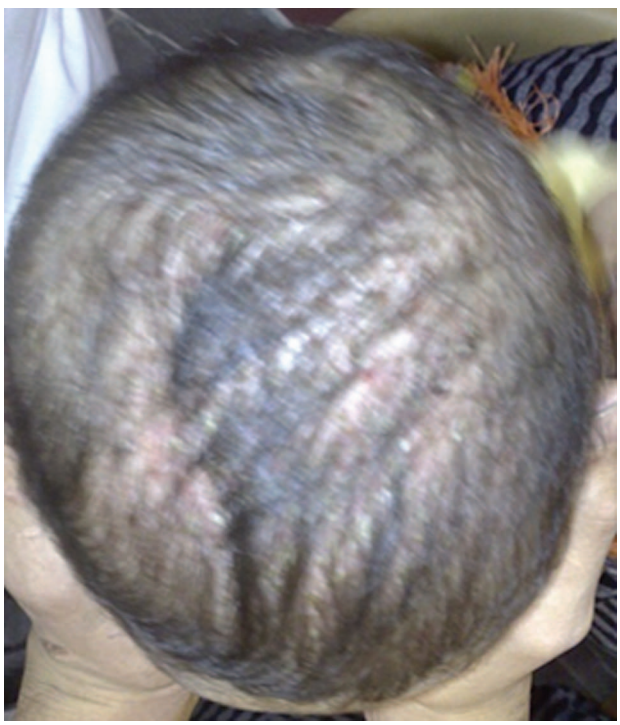

Figure 1. Skin lesions of the scalp with concomitant alopecia.

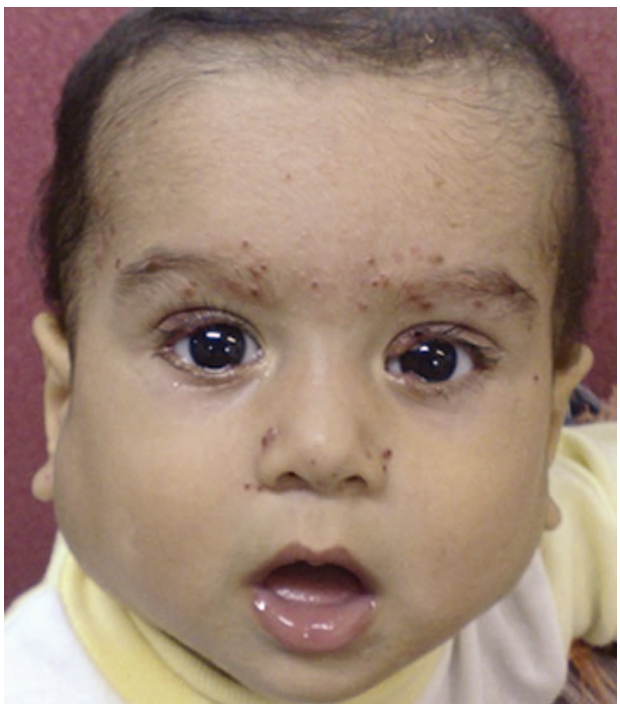

Figure 2. Erythematous skin lesions of face. Bulging of both mandibular angles demonstrates bilateral parotiditis.

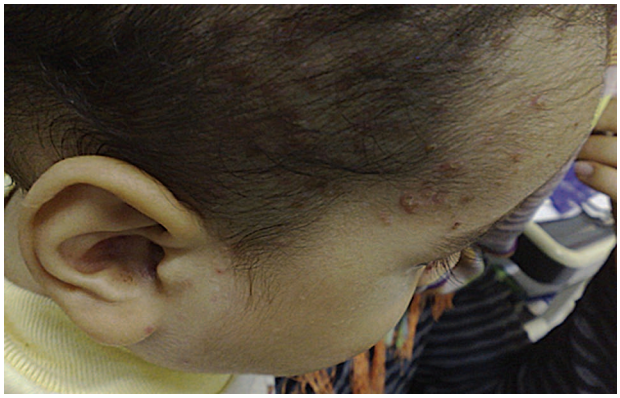

Figure 3. Erythematous papules on the parieto-temporal scalp and forehead. Note the enlarged parotid gland.

affects young children and its clinical presentation differs from single self-healing lesions to a multi-system involvement with organ dysfunction such as bone, liver, spleen, lung, central nervous system, skin, bone marrow and gastrointestinal tract [2]. The most common endocrine involvements are diabetes insipidus and growth hormone deficiency [3]. The inflammatory process of Langerhans histiocytes infiltrating the pituitary stalk leads to a fibrotic intimal proliferatory response which impairs pituitary microvasculuture. This vasculopathy compromises the pituitary functions [4].

The common sites of involvement in Langerhans cell histiocytosis are the bone, lung, skin, liver, spleen, and lymph nodes [5]. In our case, however, only the lymphatic and skin involvement was detected besides bilateral infiltration of the parotid glands as a unique feature.

The determinants of recurrent disease and poor prognosis in LCH include simultaneous bone and mucocutaneous tissue involvement, concomitant infiltration of osseous and extraosseous tissues, hepatosplenomegaly in children less than 3 years old at the onset, pituitary-thalamic axis involvement in the presence of a multi systemic disease, and LCH with features of three or more system involvement [5]. Patients with localized LCH may have a good chance of spontaneous remission and a good prognosis over a period of months to years [6]. The presented case carried none of the poor prognostic criteria and is therefore anticipated to have a favorable outcome.

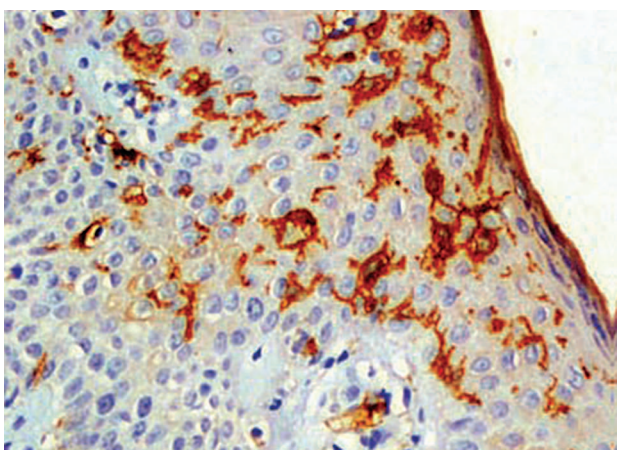

Figure 4. Immunohistochemistray study on skin lesions showing positive reaction to CD1a antigens (IHC counterstained with hematoxilyn*100).

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