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Case report

# Congenital self-healing reticulohistiocytosis (Hashimoto–Pritzker syndrome)

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**Abstract**

A case of congenital self-healing reticulohistiocytosis in an otherwise healthy newborn boy is presented. Erythematous papules covered by scales on the groin and single nodule on the left side of the forehead, which disappeared spontaneously within four months. Clinical features, histological and immunohistochemical findings are described. Up to our knowledge this is the first case described in Saudi Arabia.



*Keywords:* Congenital; Reticulohistiocytosis; Erythematous papules; Scales; Nodule; Immunohistochemical

## 1. Case report

Full term male newborn, a product of Normal Vaginal Delivery, primigravida and consanguineous marriage presented with erythematous papules covered by scales on the groin (Fig. 1) and single nodule on the left side of the forehead (Fig. 2). Skin biopsy examination revealed large mononuclear cells and multinucleated giant cells with ground-glass or foamy cytoplasm present in the dermis and epidermis (Figs. 3 and 4). Immunoperoxidase staining was positive for CD1 and S-100. Electron microscopic study showed Langerhans cell granules (Fig. 5). Physical examination revealed no lymphadenopathy, no organomegaly, complete blood count, liver function tests, and bone survey. Liver-spleen scan and bone marrow biopsy were done without any evidence of systemic involvement. The skin lesions had disappeared spontaneously within four months. Follow-up for the baby for six month period revealed no evidence of any relapse. The diagnosis of congenital self-healing reticulohistiocytosis (Hashimoto–Pritzker syndrome) was considered based upon histopathogical findings along with clinical correlation.

## 2. Discussion

Congenital self-healing reticulohistiocytosis (CSHLCH) or (Hashimoto–Pritzker syndrome) represents a special form of purely cutaneous Langerhans cell histiocytosis, first described in 1973. This clinical entity is usually present at birth or appears very soon thereafter. Affected infants are otherwise healthy and skin lesions tend to involute spontaneously within weeks to months. The diagnosis of LCH is based on histopathology which is indistinguishable for all forms of LCH. Characteristic histopathology and absence of other system involvement permit differentiation

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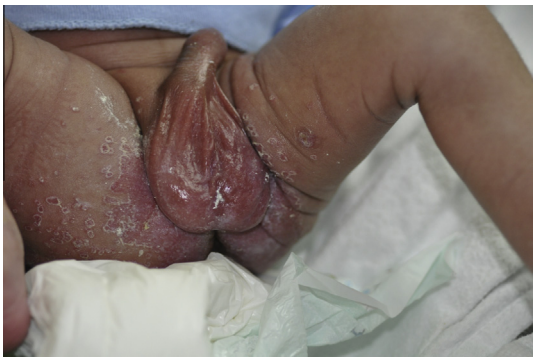

Figure 1. Erythematous papules covered by scales on the groin.

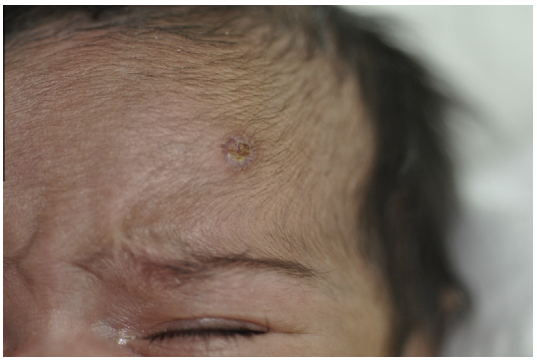

Figure 2. Single nodule on the left side of the forehead.

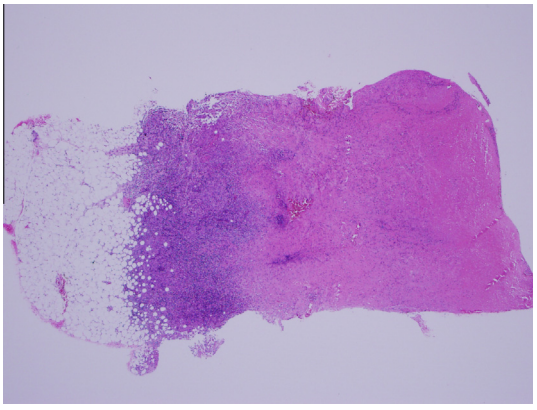

Figure 3. Low power large mononuclear cells and multinucleated giant cells with ground-glass or foamy cytoplasm present in the dermis and epidermis.

of benign forms of LCH. The diagnosis of CSHLCH should be made only retrospectively, after exclusion of involvement of other organs/systems (Popadic et al., 2012). It has been described in two forms: a solitary and a multinodular variant. Solitary or generalized lesions can affect any part of the cutaneous surface. Lesions range from 0.2 to 2.5 cm in diameter. Lesions may grow postnatally. Exceptionally large tumors up to 8 cm can occur (Alexis et al., 1991). At presentation the lesions can be papules or nodules with or without erosion or ulceration.

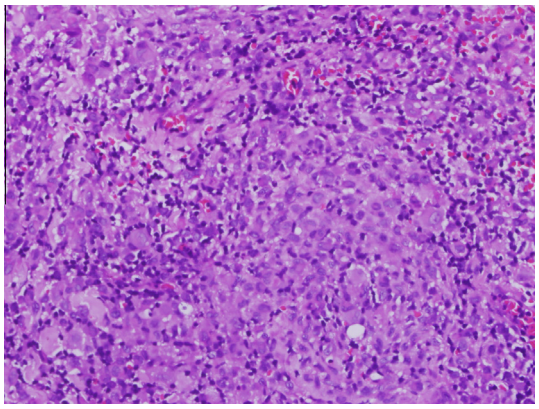

Figure 4. High power large mononuclear cells and multinucleated giant cells with ground-glass or foamy cytoplasm present in the dermis and epidermis.

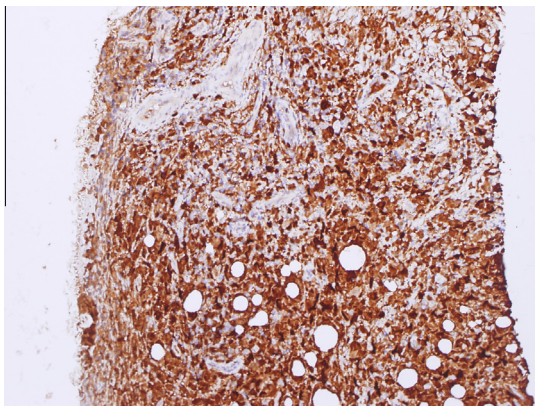

Figure 5. Electron microscopic study showed Langerhans cell granules.

Individual lesions are red, brown, pink, or dusky. Lesions greater than 1 cm characteristically ulcerate as they resolve. Lesions are asymptomatic and spontaneously involute over 8–24 weeks, leaving atrophic scarring from the ulcerated nodules. Internal involvement is not found (Elguezabal et al., 2011).

Histiocytosis has been reported in Saudi Arabia, a retrospective analysis of 21 Saudi children with Histiocytosis-X was reviewed and follow-up from 1 to 10 years revealed bone involvement in 19 patients and three patients died, three had recurrences and eight had other organ dysfunction (Al-Molhim et al., 1991). However in our case there were no systemic involvement and the disease was confined to the skin.

According to the data collected by Belhadjali et al., CSHLCH may be divided into two groups, the first group with skin lesions appeared at birth as observed in our case, and the second group with later onset (Belhadjali et al., 2008). Estimated incidence of LCH is 2.24–8.9 cases/one million/year among European children (Müller et al., 2006; Stalemark et al., 2008). The real incidence of self healing LCH is unknown and may be underestimated due to the spontaneous resolution and failure of clinical recognition (Kapur et al., 2007).

The etiopathogenesis of reticulohistiocytosis is unknown, but the possibility of an immunological etiology has been suggested by the occurrence of a number of immunological abnormalities in this disease. The fact that the lesional cell bears the phenotype of the Langerhans cell, which is a key cell in the immune system also suggests that the immune system is in some way involved in its pathogenesis. None of the immunological abnormalities, however, have been a consistent feature, and most authors now consider them to be epiphenomena (Cunliffe and Simpson, 2010).

On histologic examination large mononuclear cells and multinucleated giant cells with ground-glass or foamy cytoplasm are present in the dermis and epidermis. Immunoperoxidase staining is positive for CD1, HLA-DR, and S-100. By electron microscopy 10%–25% of cells have Langerhans cell granules. This histology is characteristic but cannot distinguish this entity from other forms of Langerhans cell histiocytosis, so a definitive diagnosis cannot be made histologically (Gadner and Grois, 1999).

Systemic evaluation is recommended, including a physical examination, complete blood count, liver function tests, and bone survey. Liver-spleen scan and bone marrow biopsy should be considered to rule out systemic involvement (Gadner and Grois, 1999).

The differential diagnosis includes seborrhoeic dermatitis, juvenile xanthogranuloma, xanthoma disseminatum, and benign cephalic histiocytosis. In disseminated Langerhans cell histiocytosis there may be diagnostic problems with familial hemophagocytic lymphohistiocytosis, sinus histiocytosis with massive lymphadenopathy, and viral-associated hemophagocytic syndrome. Histological examination of tissue biopsy with specific marker studies is usually sufficient to differentiate Langerhans cell histiocytosis from other conditions (Odom et al., 2000).

Treatment depends on the extent and the severity of the disease. Patients with single-system bone or skin disease have a good prognosis and often require no or only limited treatment. In a recent study, McLelland et al showed that in 14 patients with single-system disease, eight required no treatment. In single-system skin disease, topical treatment with 20% nitrogen mustard is effective. Psoralen and UVA (PUVA) therapy may be useful for those patients that do not tolerate topical nitrogen mustard or fail to respond adequately. Recent reports have shown good response of isolated skin disease to thalidomide (McLelland et al., 1990; Shahidi-Dadras et al., 2011), however spontaneous resolution can occur like what happened with our case.

## Conflict of interest

None.

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