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Congenital Self-healing Reticulohistiocytosis
-Report of a Case of the Solitary Type and Review of the Literature-

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A 2-month-old female infant presented with a single hemorrhagic crusted papule on the chin present since birth. No visceral involvement could be demonstrated. The lesion involuted spontaneously with scarring in 3 months. Mononuclear cells in the cutaneous infiltrate were Langerhans cells with typical Birbeck granules which positively stained with S-100 protein. This case is the solitary type of congenital self-healing reticulohistiocytosis.

Key Word: Congenital self-healing reticulohistiocytosis

Congenital self-healing reticulohistiocytosis (CSHR) reported first by Hashimoto and Pritzker (1973), is a rare primary histiocytic skin disorder. Before Berger *et al.* (1986) described the solitary type of this entity, all reported cases of CSHR had multiple cutaneous lesions; only 33 cases had been described in the literature, including one solitary type (Kanitakis *et al.* 1988). Since Berger *et al.* (1986) first reported four more cases of the solitary variant, a total of 7 cases of this type have been reported (Taieb *et al.* 1986; Jordaan *et al.* 1986; Ofuji *et al.* 1987).

We report herein a solitary type of CSHR and review the previous seven reported cases of solitary type of CSHR.

CASE REPORT

A 2-month-old female infant came to our department with her mother because of a lesion on the chin since birth. She was delivered vaginally at term as a first daughter at a private clinic. The past and family history were non-contributory. Physical examination revealed an apparently healthy female infant without hepatosplenomegaly or lymphadenopathy. She had a solitary, pea-sized, erythematous papule with hemorrhagic crusts on the center of her chin (Fig. 1). A partial skin biopsy was performed.

The histopathology of the specimen showed a well-defined, dense, nodular infiltration of mononuclear cells into the entire dermis (Fig. 2); the infiltration extended into the hair follicle, which was dilated and destroyed. In the epidermis, there was parakeratosis, crusting, and thinning. The dermoepidermal junction was indistinct (Fig. 3). The majority of

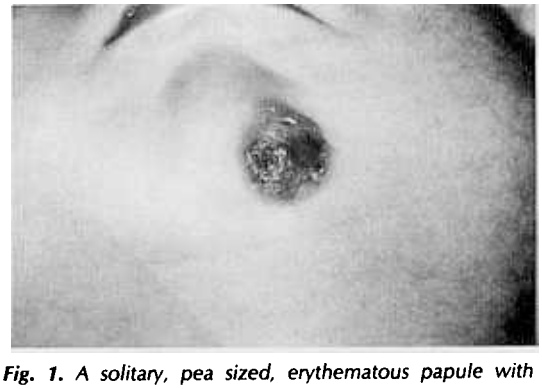

Fig. 1. *A solitary, pea sized, erythematous papule with hemorrhagic crusts on the chin.*

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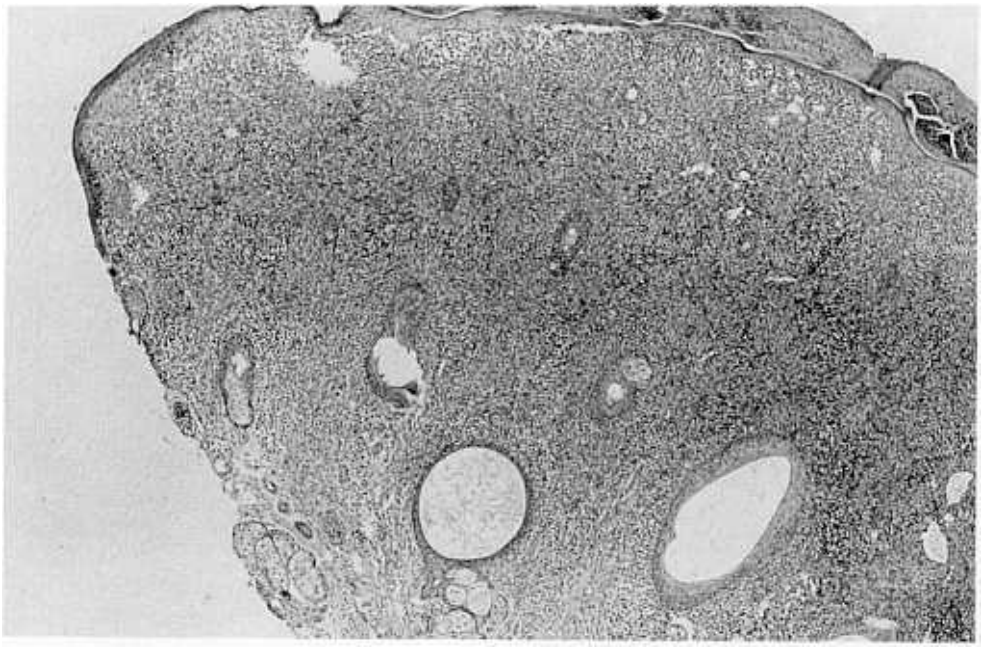

Fig. 2. A well-defined, dense, nodular infiltration of mononuclear cells into the entire dermis (Hematoxylin-eosin, ×40).

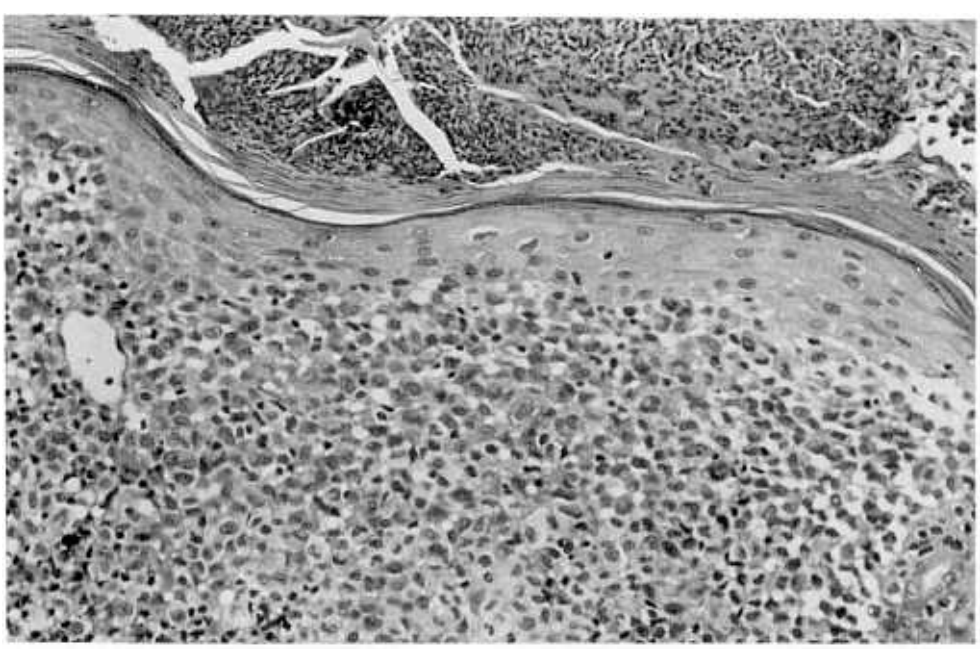

Fig. 3. Parakeratosis, crusting, and thinning of the epidermis and indistinct dermoepidermal junction (Hematoxylin-eosin, × 200).

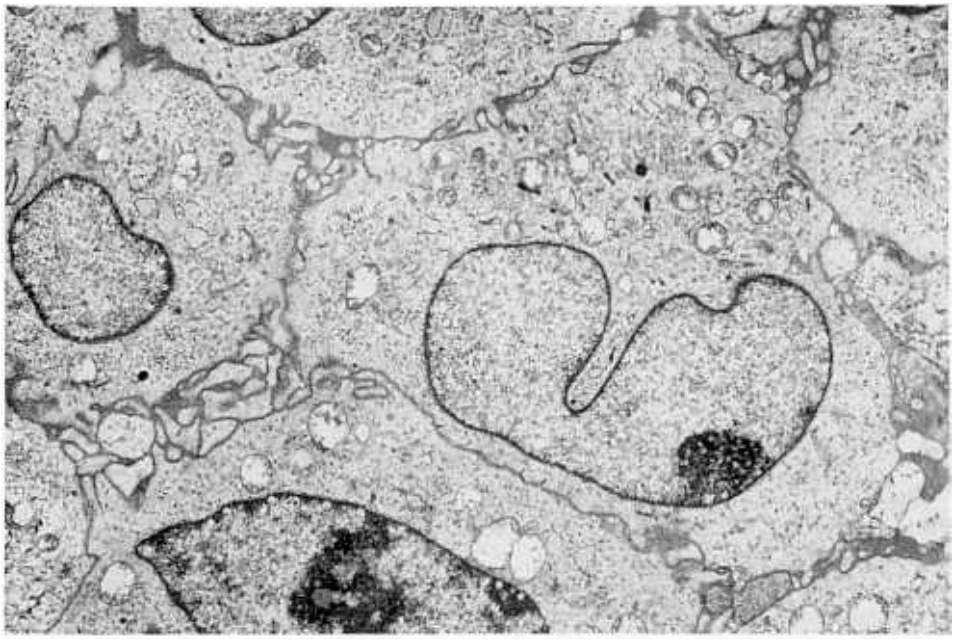

Fig. 4. The infiltrating cells in the dermis showed large, round to polyhedral cells displaying notched and bean shaped nuclei. The cytoplasm was light and abundant and contained various organelles (×8,750).

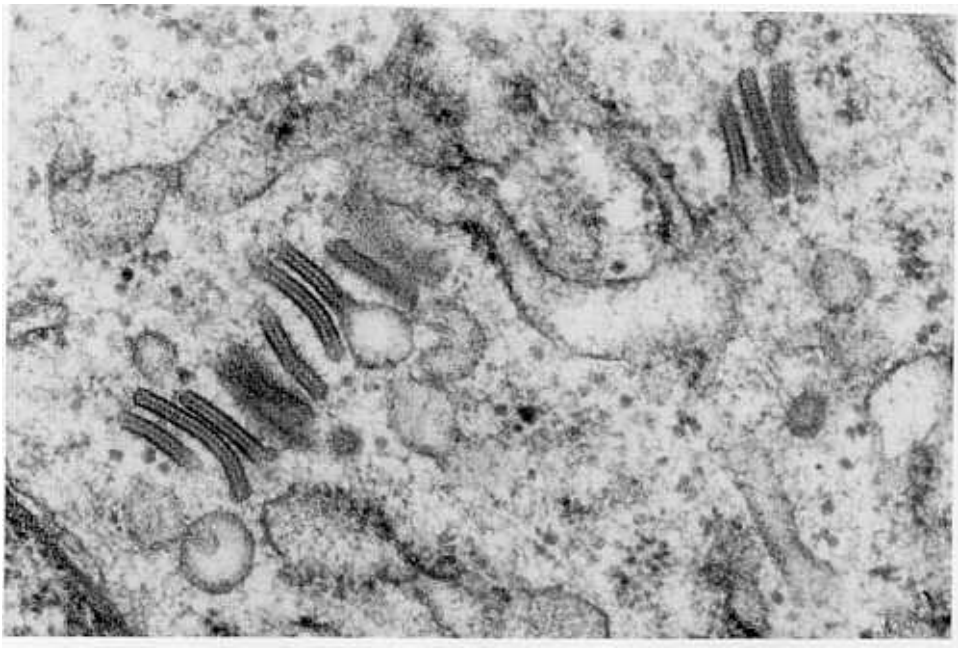

Fig. 5. The cell had Birbeck granules (×90,000)

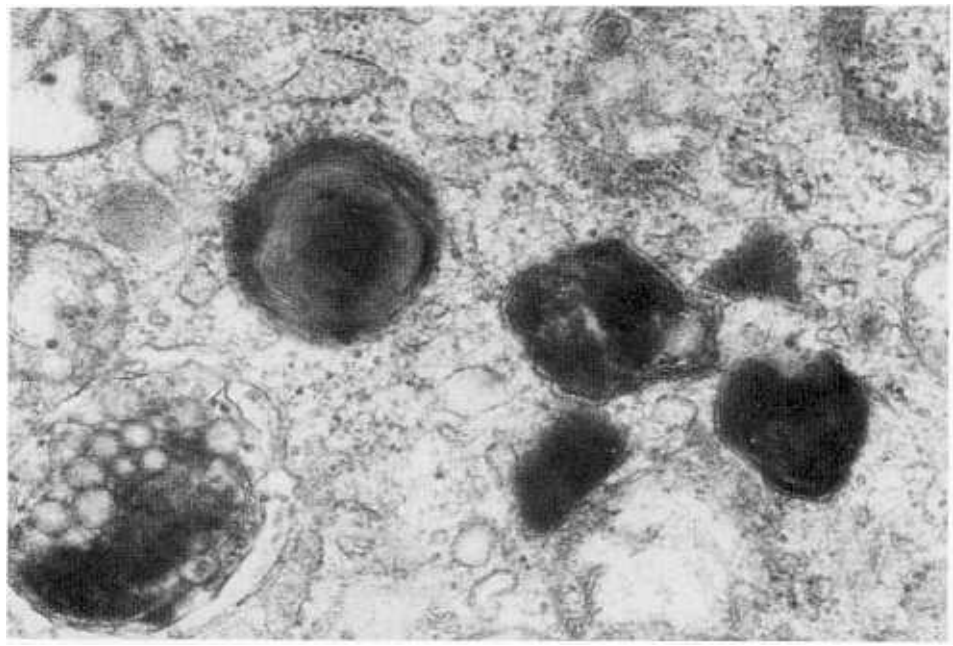

Fig. 6. The cell had laminated bodies (×75,000).

the cells composing the infiltrate were large and round, with abundant eosinophilic cytoplasm. The nuclei were of various shapes and sizes, from irregularly round to elliptic or kidney-shaped. Associated inflammatory infiltrates consisting of lymphocytes and eosinophils were also present. Thus, the histopathologic picture was indistinguishable from that of Letterer-Siwe disease.

The laboratory results of routine CBC, SMA, PT, PTT, and immunoelectrophoresis were normal. Roentgenograms of the chest were normal except for a thickened periosteal new bone on the left mid-clavicle area, suggestive of birth trauma. Roentgenograms of the skull were normal. A whole body bone scan was negative, except for an abnormally increased uptake on the mid-portion of the left clavicular area.

An electron microscopic examination revealed a dense, monomorphous cellular infiltrate throughout entire dermis which also invaded the epidermis. It consisted of large, round or polyhedric cells displayng occasionally notched and bean-shaped nuclei. The cytoplasm was light and abundant, containing various organelles such as mitochondria, vesicles, lysosomes, endoplasmic reticulum, and well developed Golgi apparatus (Fig. 4). The most frequently encountered intracellular inclusions were Birbeck granules (BGs) (Fig. 5). Some cells also had laminated or non-laminated dense bodies (Fig. 6). An immunohistochemical study with S-100 protein revealed that the histiocyte-like cells stained strongly positively in their cytoplasm and nuclei, indicating that these were Langerhans cells (LCs).

The patient's skin lesion spontaneously resolved with scarring during our investigation. Therefore, we diagnosed this patient as having congenital self-healing reticulohistiocytosis.

DISCUSSION

CSHR is characterized by the following: 1) congenital symptomless papulonodule (s), 2) self-healing within a few months without recurrence, 3) no systemic symptoms and no visceral lesions, 4) histopathology showing large mononuclear cells and multinucleated giant cells of histiocytes in the dermis and epidermis, 5) electron microscopy showing BGs and dense bodies (some with myelin-like lamination), and 6) positive immunoperoxidase staining for OKT 6, HLA-DR (Ia), and S-100 (Hashimoto and

Table 1. The solitary type of congenital self-healing reticulohistiocytosis as reported in the literature

Case	Age	Sex	Onset	Site	Time of regression (weeks)	BG	LB	NLB
1.	Newborn	F	At birth	Rt Remple	18	+	+	
2.	Newborn	M	At birth	Rt foot	9	+		
3.	Newborn	M	At birth	Rt hand	?	+		
4.	Newborn	M	At birth	Lt inguinal area	10	not done		
5.	3-month	F	3weeks	Lt buttock	?	+rare	+	
6.	Newborn	M	At birth	Rt iliac fossa	4	+10-15%	+	+
7.	Newborn	M	At birth	Lt shoulder	?	+32/133	+	
8.	2-month	F	At birth	Chin	12	+	+	

Case 1-4 : Berger *et al.* 1986
Case 5 : Taieb *et al.* 1986
Case 6 : Jordaan *et al.* 1986
Cae 7 : Ofuji *et al.* 1987
Case 8 : Our case
BG : Birbeck granule
LB : Laminated bodies
NLB : Non-laminated bodies

Pritzker 1973).

Recently, seven cases of solitary lesions that otherwise fit the clinicopathologic criteria of CSHR have been reported (Hashimoto and Pritzker 1973; Jordann *et al.* 1986; Ofuji *et al.* 1987) (Table 1). Of these cases, 5 were males and 2, females. The age of onset was at birth in 6 cases and at 3 weeks after birth in 1 case. The sites of occurrence of the lesion were temple, foot, hand, inguinal areas, buttock, iliac fossa, and shoulder. Spontaneous regression occurred between 1 month and 18 weeks. On electron microscopic examination, BGs were seen in 6 cases, laminated bodies in 4, and non-laminated bodies in 1. We believe our patient represented a solitary type of CSHR. In our case, the cutaneous lesion was solitary and present since birth. Systemic involvement was not detected from our laboratory results. Histopathologic examination of the lesion showed findings very similar to those of Letterer-Siwe disease. Electron microscopy demonstrated specific markers for LCs in the infiltrating cells, and by the immunoperoxidase technique, the infiltrating cells were positively stained with S-100 protein. The lesion spontaneously resolved within 3 months. The solitary type of CSHR appeared at birth with no predilection areas and regressed spontaneoulsy within 18 weeks. The infiltrating cells were Langerhans cells.

Letterer-Siwe disease and CSHR are the only two diseases currently identified as proliferation of the LCs (Berger *et al.* 1986). Therefore, we suggested that Letterer-Siwe disease is the systemic infiltration of LCs including the skin and CSHR is the only cutaneous infiltration of LCs.

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