Congenital Self-Healing Histiocytosis
-A Case Report-

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A case of congenital self-healing histiocytosis was studied with S-100 antibody and electron microscopy. Many tumor cells were positive for S-100 protein and a few contained Birbeck's granules and dense bodies. A pathologic fracture was first noticed on the seventh day. The lesions involuted spontaneously by the end of a weeks
(Ann Dermatol 7(2)177~180, 1995)

Key Words : Congenital self-healing histiocysis

Congenital self-healing histiocytosis is a rare primary histiocytic skin disorder which was described firstly in 1973 by Hashimoto and Fritscher. Diagnostic criteria are as follows: 1) congenital or perinatal occurrence of multiple dermal nodular lesions that are brownish pink or dusky red and have a generalized distribution; 2) dermal infiltration of large histiocytic cells with eosinophilic cytoplasm and with diastase-resistant inclusions that are positively stained with periodic acid-Schiff(PAS) reagent; 3) eosinophils that are usually admixed with increased reticulum fibers surrounding large tumor cells, and epidermis that is usually infiltrated and often ulcerated, and 4) ultrastructurally, 10% to 25% of large tumor are characterized by dense bodies. Here we report a case showing the lytic lesion of metacarpal bone with multiple skin lesions. We use immunohistochemical staining for S-100 protein and electron microscopy in our study.

REPORT OF A CASE

A male infant was born with approximately 10 skin lesions distributed over the head, trunk and extremities including the palm (Fig. 1). The lesions were composed of 2 to 12mm, red-violaceous papules and nodules and some showed central ulceration and crusting. Otherwise the infant was on excellent health. There was no hepatosplenomegaly or lymphadenopathy. The past and family history were non-contributory. Laboratory Studies including complete blood cell count, differential count, blood chemistry study, chest x-ray, infantogram, ultrasound examination were normal.

on hematoxylin-cosin stain, the biopsy specimen taken from the papule showed the epidermis was eroded by underlying cellular infiltrate and was covered by a crust. There were diffuse infiltrates of histiocytic cells eccentrically located, and were indented or kidney-shaped viscular nuclei intermingled with lymphoid cells in the entire dermis. The infiltrate invaded the epidermis, hair follicles and subcutis (Fig. 2 & 3). PAP staining for S-100 protein was positive (Fig. 4). An electron microscopic examination revealed that the typical tumor cells had abundant cytoplasm and a deeply invaginated nuclear contour. The cytoplasm contained Birbeck's granule's and dense bodies (Fig. 5). These findings were consistent with Langerhans cells.

On the 7th day, a pathologic fracture developed on the right 2nd metacarpal bone (Fig. 6). An infantogram was done however no abnormal bony lesions were noted elsewhere. The patient's skin lesion spontaneously resolved with postinflammatory hyperpigmentation by the end of a weeks. The osteolytic lesion also healed with new bone

Received May, 16, 1994.
Accepted for publication July 4, 1994.
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DISCUSSION

Histocytic diseases in childhood represent a wide spectrum of entities ranging from reactive non-neoplastic disorders to true tumors. This multiplicity accounts for the variety of clinicopathologic presentations of the diseases. Among these, histiocytosis X (HX) is a relatively well defined subgroup of histiocytosis. Its immunohistochemical and ultrastructural features closely link the proliferating cells to epidermal Langerhans cell (LC). This group includes at least three diseases (Lettterm-Siwe disease, Hand-Schuller-Christian disease, and eosinophilic granuloma) that, despite different clinical severity, share a common pathologic substratum (i.e., the proliferation of abnormal LC).

Congenital self-healing histiocytosis (CSHH) is a rare primary histiocytic skin disorder. The precise nosologic position of the disease is still difficult to establish because CSHH has many features in common with the classical forms of HX. The disease affects both sexes approximately equally with all races being affected. The clinical presentation includes a cutaneous eruption of multiple, red-violaceous or brown, firm, indolent nodules that have a tendency to ulcerate, scattered over the body at birth or shortly after birth. In the case of...
Herman et al., papulovesicles predominated exceptionally. Recently solitary lesions have been reported. Mucous membranes are always spared. Systemic signs are usually absent and the general health is good. The usual laboratory tests are normal. In rare instances hepatomegaly and hematologic abnormalities including bone marrow hyperplasia may be seen. In the case reported by Hawkins and Langston, pulmonary nodule, lytic lesions of and lymph node involvement were observed in addition to hepatosplenomegaly. In our case, an osteolytic bony lesion was noted, however, other systemic abnormalities were not observed. This lesion has a tendency towards spontaneous regression. The cutaneous lesions begin to involute within 2-3 months and the only sequelae are hyperpigmentation or atrophic scars. Even the cases with apparent systemic involvement display a benign clinical course. Our case also showed involution of skin and bony lesions by the end of 6 weeks.

Histologically, the lesions consist of a dense cellular infiltrate occupying the dermis. Epidermal involvement is variable. This is composed of large histiocytes with an abundant, eosinophilic cytoplasm containing PAS(+) granules and a notched, occasionally bean-shaped nucleus. Langerhans cell markers help to differentiate this tumor from other histiocytic proliferation and lymphomas. These include stains for S-100 protein, ATPase, and if fresh frozen sections are available. T6 and HLA-DR antigens. Unfortunately, none of these is a specific LCs or indeterminate cells(LC without
REFERENCES


