

5 Cases of Solitary Mastocytoma

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We report five cases of solitary mastocytoma. All patients were under one year of age and diagnosed as solitary mastocytoma based on clinicopathologic findings. Skin lesions were found on the right forearm, left hand, left leg, left elbow and left shoulder. They resolved themselves spontaneously resolution within 2 or 3 years. (Ann Dermatol 9:(2) 155~158, 1997).

Key Words : Solitary mastocytoma

Mastocytosis is a disorder of mast cell proliferation that occurs in both cutaneous and systemic forms. Cutaneous lesions include urticaria pigmentosa, mastocytoma, diffuse and erythrodermic mastocytosis including bullous mastocytosis, and telangiectasia macularis eruptiva perstans. In infancy or childhood, cutaneous mastocytosis usually presents with multiple lesions like urticaria pigmentosa. But mastocytoma is a solitary lesion and comprises of about 10% of cutaneous mastocytosis¹.

We describe five cases of solitary mastocytoma in infancy.

REPORTS OF A CASE

Case 1

A 1-year-old female infant was examined for evaluation of the lesion on the right forearm which had been present for 4 months. Physical examination revealed a 3 X 7 mm sized yellowish spindle-shaped nodule on the right forearm (Fig. 1). Rubbing produced slight swelling. No other lesions suggestive of urticaria pigmentosa were found on physical examination of the entire skin. Histological examination revealed dense aggregates of cells in the upper dermis. The cells were large mononuclear cells with abundant basophilic cy-

toplasm, and scattered eosinophils were also present (Fig. 2). Staining with toluidine blue demonstrated mononuclear cells to have metachromatic granules, confirming the diagnosis of cutaneous mastocytoma (Fig. 3).

Case 2

A 6-month-old female infant was noted by her mother to have a brownish polygonal plaque on the dorsum of the left hand at age 3 months (Fig. 4). The plaque showed a positive Darier's sign. Physical examination revealed no similar skin lesions on other skin surface of the body. Histological examination confirmed mastocytosis.

Case 3

A 1-year-old female infant was noted to have a brownish linear palpable plaque on the left leg at birth. There was no family history of similar skin lesions. No other similar skin lesions were found on physical examination. Histopathological examination confirmed mastocytosis.

Case 4

A 10-month-old male infant was seen for evaluation of a lesion on the left elbow which had occurred 4 days ago (Fig. 5). Similar lesions at the same site had developed 2 months before. Physical examination showed an erythematous well-demarcated firm nodule on the left elbow. There were no similar skin lesions on other skin surfaces. Histopathological examination revealed findings of mastocytosis.

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Table 1. Summary of five cases

| Case | Sex/Age | Age of onset | Site | Darier's sign | Spontaneous regression |
|------|---------|--------------|----------------|---------------|------------------------|
| 1 | F/1yr | 8 mo | forearm, right | + | + |
| 2 | F/6mo | 1 mo | hand, left | + | + |
| 3 | F/1yr | at birth | leg, left | - | + |
| 4 | M/10mo | 8 mo | elbow, left | - | + |
| 5 | F/5mo | at birth | shoulder, left | + | + |

+: positive, -: negative

Fig. 1. Yellowish spindle-shaped nodule on the right forearm (Case 1).

Fig. 2. The mast cells appear cuboidal in shape with abundant basophilic cytoplasm, and lie closely packed in the upper dermis. Scattered eosinophils are also seen in the dermis ($\times 200$, H&E stain) (Case 1).

Fig. 3. Toluidine blue stain demonstrates the mast cell granules to be metachromatic in character ($\times 200$, Toluidine blue stain) (Case 1).

Fig. 4. Brownish polygonal plaque on the dorsum of left hand (Case 2).

Case 5

A 5-month-old female infant was seen for evaluation of a lesion on the left shoulder which had been present since birth. Physical examination revealed a 5 X 16 mm sized brownish spindle-shaped

nodule on the left shoulder (Fig. 6). Rubbing produced swelling. No other lesions suggestive of urticaria pigmentosa were found on physical examination. Histological examination confirmed mastocytosis.

Fig. 5. Erythematous well-demarcated firm nodule on the left elbow (Case 4).

Fig. 6. Brownish spindle-shaped nodule on the left shoulder (Case 5).

DISCUSSION

Mastocytosis may present during infancy, childhood or adulthood. The disease is characterized by an increased number of tissue mast cells and symptoms are caused by the release of histamine and other mast cell mediators, such as leukotrienes, prostaglandins and platelet-activating factor^{12, 13}. The organ system most frequently involved is the skin; however, mastocytosis may involve the liver, spleen, lymph node, gastrointestinal tract, bone marrow and skeletal system⁴. The occurrence of mastocytosis appears to be sporadic, yet 47 cases of familial mastocytosis have been reported^{5, 11}.

The typical presentation of pediatric-onset mastocytosis consists of cutaneous manifestations of either an urticaria pigmentosa, solitary mastocytoma or less commonly, diffuse cutaneous masto-

cytosis⁶. The commonest presentation of cutaneous mastocytosis in childhood is urticaria pigmentosa appearing as numerous small round or ovoid red or red-brown macules or papules, usually in the first six months to two years of life¹. Solitary mastocytoma has been estimated to be present in 10 to 15% of patients with cutaneous mastocytosis². Almost all of the reported cases of solitary lesion have been in infants or young children. In a review of Korean literature, 5 cases of solitary mastocytoma have been reported¹³⁻¹⁵ and they were all under 2 years of age like the present 5 cases. Although mastocytomas may be present at birth, most appear within the first three months of life; they are rarely described in adults¹⁻³. The usual site is on the trunk or limbs, but not the palms or soles. The solitary lesion is yellow, red-brown or gray in color and is characterized by a 0.5 to 3 cm sized cutaneous nodule or slightly elevated or macular lesion. Lesions of mastocytoma rarely continue to increase in number of additional lesions more than 2 months subsequent to the initial lesion¹. Although some children manifest one form of skin lesion, overlap in classification can occur. In most cases of mastocytosis, the lesions urticate on rubbing, due to mediator release (Darier's sign), but vesiculation or frank blistering is common in lesions in infancy. In our cases, 3 patients were positive in Darier's sign. Pruritus is the primary presenting symptom in children with mastocytosis. The pruritus may be intermittent or unremitting with extensive excoriation of the skin. Additional symptoms include flushing and gastrointestinal complaints, such as vomiting, colicky pain and diarrhea⁷.

In urticaria pigmentosa the number of mast cells in the papillary dermis, particularly around blood vessels, is increased⁸. These cells are characterized by metachromasia by use of basic dyes, such as Giemsa's reagent or toluidine blue. Recently, the use of conjugated avidin and morphometric point counting of mast cells have aided in the diagnosis⁹. The histological alterations in single, multiple and disseminated lesions of urticaria pigmentosa are indistinguishable⁸. If the lesion has been traumatized, edema of the papillary dermis and even subepidermal bullae are present. The infiltrating cells also include eosinophils. In the mastocytomas, mast cells may infiltrate the entire dermis and even extend into the subcutaneous tissue. These tumor-like aggregations of mast cells are often

associated with edema of the papillary dermis and at times, with subepidermal bullae.

The prognosis for children with mast cell disease is variable. Children with mastocytomas have not been reported to develop systemic involvement and their lesions typically exhibit involution during childhood^{2, 10}. In our cases, all underwent spontaneous resolution of the skin lesion in 2 or 3 years. Retrospective studies report that approximately half of the children with urticaria pigmentosa will experience resolution of lesions and symptoms by adolescence, with the remainder exhibiting marked reduction in symptomatic cutaneous lesions and dermatographism⁶.

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