

Large Hyperpigmented Plaques on the Trunk of a Newborn

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A 4-day-old girl with no notable medical history presented with 2 pink lesions on the right side of the back and left side of the flank. Both lesions were present at birth and had not changed in size, shape, or color in the first 4 days of life. She had no constitutional symptoms. The child was a full-term newborn, and her mother experienced no pregnancy or delivery complications. She had no family history of similar skin findings.

What's the diagnosis?

- a. aplasia cutis congenita
- b. connective tissue nevus
- c. cutaneous mastocytoma
- d. epidermal nevus (epidermolytic type)
- e. epidermolysis bullosa

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The authors report no conflict of interest.

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The Diagnosis: Cutaneous Mastocytoma

Physical examination revealed a 58×51-mm hyperpigmented plaque with central pink coloration and scale on the right side of the back as well as a 39×33-mm pink plaque with a hyperpigmented border on the left side of the flank (Figure 1). At follow-up 2 weeks later, the patient's parents reported that blisters formed within both of the plaques. The blisters ruptured a few hours after forming and drained clear fluid with scant blood. Both plaques contained erosions from the ruptured bullae but remained the same size with no surrounding erythema or warmth. A 4-mm punch biopsy was performed of intact skin from the back lesion (Figure 2A). Histologic examination revealed a cellular infiltrate of monotonous bland cells that completely filled the dermis without epidermal involvement, along with occasional intermixed eosinophils. The morphology of these infiltrating cells was compatible with mast cells confirmed by strongly positive Leder staining (Figure 2B).

Mastocytosis encompasses a rare group of disorders characterized by abnormal mast cell accumulation or mast cell mediator release in various tissues. These disorders can be classified as either systemic mastocytosis with mast cell infiltration into bone marrow or other extracutaneous organs, or cutaneous mastocytosis with disease limited to the skin.¹ Mutations involving activation of the c-Kit receptor in stimulating mast cell growth and development have been implicated in both systemic and cutaneous forms of the disease.^{2,3}

Cutaneous mastocytosis is most often diagnosed in childhood and typically is characterized by spontaneous regression before puberty in a majority of cases.^{1,4} Under the World Health Organization classification system, cutaneous mastocytosis can be further subdivided into 3 disorders (listed in order of most to least common): urticaria pigmentosa (also known as maculopapular cutaneous mastocytosis) with typical, plaque, and nodular forms; cutaneous mastocytoma (as seen in this



Figure 1. Cutaneous mastocytoma. Lesion on the left side of the flank 1 month after birth during a follow-up visit.

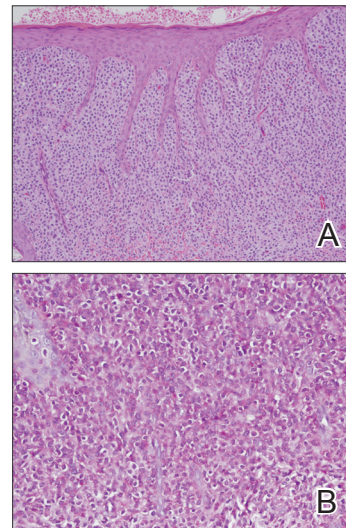


Figure 2. A 4-mm punch biopsy of the lesion on the right side of the back revealed an infiltrate of monotonous bland cells that filled the dermis with occasional eosinophils (A)(H&E, original magnification ×10). Leder stain of the biopsy specimen was strongly positive for mast cell infiltrate (B)(original magnification ×20).

patient); and diffuse cutaneous mastocytosis.⁵ Compared to the widespread distribution of small macules and papules in urticaria pigmentosa, the cutaneous mastocytoma subtype presents with 1 to 6 brown to orange-yellow plaques or nodules measuring more than 1 cm in diameter. Cutaneous mastocytoma typically presents in infancy and is located most commonly on the trunk and extremities, though it may be found on the face or scalp. The plaques of mastocytoma often have well-defined margins, and these lesions may become bullous or demonstrate Darier sign of urtication and erythema on physical stimulation. Patients most commonly experience pruritus from mast cell degranulation and rarely exhibit systemic symptoms of mast cell mediator release; however, generalized flushing, hypotension, headaches, and gastrointestinal symptoms may occur, particularly if the lesion is vigorously rubbed.^{6,7} Conditions in the differential include aplasia cutis congenita, connective tissue nevus, epidermal nevus, and epidermolysis bullosa. They should not elicit a blister if rubbed, except for epidermolysis bullosa, which can easily be differentiated based on histology.

The workup for cutaneous mastocytosis in the pediatric population may include a biopsy of lesional skin, though in many cases the characteristic cutaneous manifestations are sufficient to make a diagnosis.

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Histologically, biopsy results often reveal abundant diffuse dermal infiltration of mast cells, which are characterized by their large pink granular cytoplasm and round dense central nuclei. In pediatric patients, mast cells typically are restricted to the dermis, and there is a low risk for hematologic abnormalities, thereby precluding the need for bone marrow examination in the absence of organomegaly or notable peripheral blood abnormalities such as severe cytopenia.^{5,6}

Management of cutaneous mastocytosis consists of avoidance of mast cell degranulation triggers and symptomatic treatment of histamine release. Triggers include certain medications (eg, narcotic analgesics, aspirin, nonsteroidal anti-inflammatory drugs, iodinated contrast agents, antibiotics, muscle relaxants), mechanical irritation, insect stings, spicy foods, stress, or extreme temperature changes.⁸ Symptomatic treatment can be achieved through topical corticosteroid or oral antihistamine use. Along with decreasing pruritus, topical corticosteroids also may be helpful in decreasing time to spontaneous resolution and healing.⁷ The patient in this case was treated with desonide ointment 0.05% daily to both lesions as well as mupirocin ointment 2% as needed for erosions. These treatments helped reduce the patient's symptoms, but her lesions persisted over a follow-up period of 4 months.

REFERENCES

1. Valent P, Sperr WR, Schwartz LB, et al. Diagnosis and classification of mast cell proliferative disorders: delineation from immunologic diseases and non-mast cell hematopoietic neoplasms. *J Allergy Clin Immunol*. 2004;114:3-11.
2. Bibi S, Langenfeld F, Jeanningros S, et al. Molecular defects in mastocytosis: KIT and beyond KIT. *Immunol Allergy Clin North Am*. 2014;34:239-262.
3. Yavuz AS, Lipsky PE, Yavuz S, et al. Evidence for the involvement of a hematopoietic progenitor cell in systemic mastocytosis from single-cell analysis of mutations in the *c-kit* gene. *Blood*. 2002;100:661-665.
4. Méni C, Bruneau J, Georjin-Lavialle S, et al. Paediatric mastocytosis: a systematic review of 1747 cases. *Br J Dermatol*. 2015;172:642-651.
5. Valent P, Horny HP, Escribano L, et al. Diagnostic criteria and classification of mastocytosis: a consensus proposal. *Leuk Res*. 2001;25:603-625.
6. Wolff K, Komar M, Petzelbauer P. Clinical and histopathological aspects of cutaneous mastocytosis. *Leuk Res*. 2001;25:519-528.
7. Patrizi A, Tabanelli M, Neri I, et al. Topical corticosteroids versus "wait and see" in the management of solitary mastocytoma in pediatric patients: a long-term follow-up. *Dermatol Ther*. 2015;28:57-61.
8. Bonadonna P, Lombardo C. Drug allergy in mastocytosis. *Immunol Allergy Clin North Am*. 2014;34:397-405.