

> THE PATIENT

3-year-old girl

> SIGNS & SYMPTOMS

- Generalized, well-dispersed rash
- Wheal development after tactile irritation
- Normal vital signs

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> THE CASE

A 3-year-old girl was brought to our clinic with a generalized rash over her scalp, face, neck, chest, abdomen, back, perianal area, extremities, and the plantar surface of her right foot. On physical examination, we noted many round, hyperpigmented, brown and reddish pink, well-circumscribed macules on her body (FIGURE). Only a few of these macules had appeared on the girl's trunk within the first 3 months of her life, but since then they'd increased in number and spread to other parts of her body as she'd aged. The lesions became edematous and erythematous with tactile irritation. Darier's sign (the development of a hive or wheal when a lesion is stroked) was present.

The patient's vitals at the time of examination included a temperature of 98.2°F, respiratory rate of 17 breaths/min, heart rate of 92 beats/min, blood pressure of 100/66 mm Hg, and oxygen saturation level of 100% on room air. The girl's parents said they hadn't traveled. There was no mucosal involvement and no systemic involvement. The patient had no past surgical or medical history, was not taking any medications, and had no significant birth history. A skin biopsy was performed.

THE DIAGNOSIS

Based on the presence of a positive Darier's sign and the results of the skin biopsy (which showed increased mast cells), we diagnosed urticaria pigmentosa (UP), which is the most common form of cutaneous mastocytosis.¹

The diagnosis had been delayed for almost 3 years because of several factors. For one thing, there had been few lesions present early in the child's life, and as a result, the parents chalked them up to "beauty marks." Then, as the lesions started to increase in number, the parents thought bed bugs were to blame.

As time went on, the parents attempted to treat their daughter's hives with homeopathic remedies suggested by family members. When the lesions didn't resolve with homeopathic remedies, the parents tried over-the-counter H1 and H2 antihistamines such as diphenhydramine, loratadine, and ranitidine. When these treatments failed, the parents brought their child to our office for medical evaluation.

DISCUSSION

UP is a chronic skin disorder in which there is an abnormal proliferation of mast cells in the dermis of the skin. It is consid-

FIGURE

Hyperpigmented macules on child's back



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Urticaria pigmentosa lesions are often self-limited and completely resolve in about half of patients by puberty.

ered an orphan disease.¹ UP that presents in children is most often benign. Approximately 50% of cases occur before 6 months of age and 25% occur before puberty.² The lesions are often self-limited and completely resolve in approximately 50% of patients by puberty.³ By adulthood, the lesions either resolve or some lightly colored non-urticating macules remain; however, some patients will continue to have a positive Darier's sign.⁴

■ Dermatologic symptoms. A patient with UP may present with brown or reddish maculopapules, papules, nodules, pruritus, and flushing of the face. Darier's sign is usually seen in cases of UP; in a study of mastocytosis in children, Darier's sign was present in 94% of cases.⁵ Lesions are more prominent in areas where clothes can rub the skin, and they often vary in size and appearance. The presence of lesions can vary from localized and scant to hundreds located over the entire body. UP can be difficult to identify when the lesions are limited, which can lead to delayed diagnosis and treatment.⁶

■ Systemic involvement. UP also can affect the skeleton, bone marrow, liver, spleen, lymph nodes, gastrointestinal tract, kidneys, cardiovascular system, and/or central nervous system. Skeletal involvement may manifest as osteoporosis or bone pain in 10% to 20% of patients with UP.⁷ Bone marrow involvement may progress to anemia or mast cell leukemia. UP can result in hepatomegaly or enlarged lymph nodes. Patients may experience nausea, diarrhea, or abdominal pain if the gastrointestinal tract is affected. Cardiovascular involvement may manifest as tachycardia and shock.⁸

Diagnosis of UP is made based on the physical exam findings noted earlier, as well as skin biopsy and laboratory results. Skin biopsy will reveal increased mast cells. In up to two-thirds of patients who have systemic involvement, laboratory testing will show elevated urine histamine levels, as well as elevated serum concentrations of tryptase.⁹

UP can appear similar to many other skin conditions

The differential diagnosis of UP can include urticaria, atopic dermatitis, contact derma-

titis, pityriasis rosea, an allergic reaction/drug eruption, Henoch-Schonlein purpura, erythema multiforme, fifth disease, folliculitis, guttate psoriasis, miliaria rubra, insect bites, viral exanthem, lichen planus, and scabies.^{10,11} In addition to the clinical appearance of the rash, these conditions can be distinguished from UP by skin biopsy and other relevant tests, as well as a thorough history.

Treatment options include antihistamines, corticosteroids, PUVA

Patients with UP should be instructed to avoid precipitating factors such as temperature changes, friction, alcohol ingestion, aspirin, physical exertion, or opiates. Treatment options include H1 and H2 antihistamines, cromolyn sodium, topical corticosteroids, and PUVA (psoralen plus ultraviolet A photochemotherapy).³ PUVA is normally avoided in pediatric patients because it is associated with an increased risk of skin cancer later in life.¹²

■ Our patient. We prescribed a topical corticosteroid, 0.05% betamethasone dipropionate cream, and oral cromolyn sodium 100 mg qid for our patient, but this failed to significantly improve the macules. The patient and parents grew increasingly anxious. Ultimately, the parents decided to have their daughter treated with PUVA in limited amounts. Topical psoralen was also used. After 2 months of treatment, the patient's lesions substantially improved and many of them disappeared. In addition, the parents were educated on the importance of sunscreen and limiting their daughter's exposure to the sun, when possible.

THE TAKEAWAY

UP can be diagnosed by taking a thorough history and conducting a physical examination; a skin biopsy that reveals increased mast cells will confirm the diagnosis. UP is usually self-limited and resolves in about one-half of patients by puberty. Treatment options include H1 and H2 antihistamines, cromolyn sodium, topical corticosteroids, and PUVA. Patients should be referred to a specialist

if their symptoms become severe, systemic UP is suspected, or they do not respond to therapy. **JFP**

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