

A Picture Says a Thousand Words

A 6-month-old with urticaria when crawling

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Keywords: Allergy; Mastocytosis; Mastocytoma; Paediatrics; Urticaria.

CASE

A previously well 6-month-old term girl presented to the emergency department with a large, single urticarial lesion on her abdomen and flushing over her torso. There were no respiratory, cardiac, or gastrointestinal symptoms. Prior to onset of her cutaneous symptoms, she had been attempting to crawl. Similar symptoms occurred twice before, again with attempts to crawl.

No new foods were introduced. Parents denied any insect stings or infectious prodrome. She was not on any medications.

On physical examination, she appeared well, and vital signs were normal. She had mild flushing of her cheeks, chest and abdomen. She also had an isolated large urticarial lesion on her abdomen, measuring 20 by 10 mm (Figures 1–2), overlying a large hyperpigmented patch (Figure 3), which parents state was



Figure 1. Urticaria and flushing overlying the patient's hyperpigmented macule.



Figure 2. Urticaria and flushing overlying the patient's hyperpigmented macule.

Received: August 24, 2018; Accepted: December 4, 2018

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Figure 3. The patient's hyperpigmented macule at baseline.

a 'birthmark'. There was no lymphadenopathy or hepatosplenomegaly. She was observed without treatment, and her urticaria and flushing resolved within 2 hours.

She was referred to the allergy clinic for further assessment. In the allergy clinic, after a thorough history and physical examination, the hyperpigmented macule on her abdomen was scratched with a tongue depressor, inducing an urticarial lesion. This physical exam finding is known as Darier's sign, and is seen in cases of cutaneous mastocytosis.

DIAGNOSIS: CUTANEOUS MASTOCYTOSIS; MASTOCYTOMA

Mastocytosis is a heterogeneous group of disorders characterized by the abnormal proliferation of mast cells (1–6). Mastocytosis can be divided into cutaneous mastocytosis (CM; mast cell proliferation limited to the skin) and systemic mastocytosis (SM; extracutaneous mast cell proliferation involving multiple organs, most commonly the bone marrow) (1–6). CM is more common in children, whereas SM is more common in adults (1,2). 60–80% of cases of CM are associated with a gain-of-function mutation in *KIT* in cutaneous mast cells (3,4). CM typically presents within the first six months of life, and lesions can be present at birth (1,2). CM can be subdivided into urticaria pigmentosa (UP), mastocytomas, diffuse cutaneous mastocytosis (DCM), and telangiectasia macularis eruptive perstans (TMEP).

UP accounts for 70 to 90% of cases of CM in paediatrics (2). Lesions in UP are typically reddish-brown in colour, measuring a few millimeters to 1 to 2 cm in diameter (2,5). Patients classically present with multiple lesions in the form of macules, plaques, or nodules (2).

Mastocytomas account for 10 to 35% of cases of CM (2). The diagnosis of cutaneous mastocytoma is made clinically. Lesions are typically brownish-red and larger than those in UP, up to several centimeters in diameter (2,3). Patients may have one or several lesions. Solitary lesions are usually present at birth or appear in the early neonatal period (2). In both UP and mastocytomas, Darier's sign is typically positive. Flushing, pruritus, and swelling around the lesion can occur secondary to mast cell degranulation (2,3).

DCM is rare, accounting for 1 to 3% of cases, but can present with severe symptoms (2). Lesions are typically diffuse, with spontaneous blistering, erythroderma, and prominent dermographism (2,5). Due to the extent of the lesions, systemic symptoms including whole body flushing, hypotension, and shock can be seen (2). TMEP is the least common form of CM, and presents with red, telangiectatic macules on a brown background (2).

If the diagnosis is unclear, a biopsy for pathology can be sent. Laboratory investigations, including a tryptase level, liver function tests (LFTs), and a complete blood count (CBC) should be sent to rule out SM (2,3). The tryptase level should be normal in CM ($<11.4 \mu\text{g/L}$), and a level of $>20 \mu\text{g/L}$ can be suggestive of SM, and should prompt further investigations, including a bone marrow biopsy (2,3).

The cornerstones of treatment for solitary mastocytomas include trigger avoidance (see Table 1 for common triggers) and symptomatic management (2–4). Antihistamines (H1-antagonists) are the first-line therapy for pharmacotherapy, and can be titrated up to four times the recommended dose (5). Colloid bandages to prevent friction, and corticosteroid creams under an occlusive dressing can also be effective (2). As

Table 1. Common triggering factors in pediatric mastocytosis (2)

Factor	Examples
Physical factors	Heat (e.g., hot baths) Sudden changes in temperature Friction or rubbing of lesions
Emotional factors	Stress Anxiety Sleep deprivation
Infectious diseases	Febrile illnesses
Medications	Nonsteroidal anti-inflammatory drugs Opioids Cough medication: dextromethorphan

anaphylaxis is exceedingly rare, epinephrine autoinjectors are not routinely prescribed (5). The majority of mastocytomas resolve by early adolescence or puberty (2,4).

Given the size of our patient's hyperpigmented macule, absence of other lesions, and a positive Darier's sign, a clinical diagnosis of solitary cutaneous mastocytoma was made. Laboratory investigations including a tryptase level, CBC, and LFTs were normal (tryptase $6.4 \mu\text{g/L}$). The family was advised to consider applying a colloid bandage prior to crawling, and to use antihistamines as needed.

Cutaneous mastocytosis should be included on the differential diagnosis of children presenting with urticaria or flushing. A thorough history of physical examination, assessing in particular for cutaneous lesions, can help make this diagnosis in the primary care or emergency department setting.

Informed consent was obtained for this case.

Funding information: No funding was secured for this report.

Financial disclosure: The authors have no financial disclosures relevant to this article to disclose.

Potential Conflicts of Interest: All authors: No reported conflicts of interest. All authors have submitted the ICMJE Form for Disclosure of Potential Conflicts of Interest. Conflicts that the editors consider relevant to the content of the manuscript have been disclosed.

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