

## Chronic, Brown-Red, Macular and Papular Rash in a Child

A 15-month-old female child presented with brown-red macules and papules all over her body. The child's mother reported that the macules, which first appeared 6 months ago, became red and swollen when scratched or rubbed, and decreased in size as the child grew. Over time, more lesions spread to the rest of the trunk and extremities. Her past medical history was unremarkable and no significant finding was detected in her family history.

Physical examination of the skin revealed multiple, widespread, oval/round brown-red macules and papules on the posterior trunk and extremities except the soles, palms, and mucous membranes (**Figure 1**). The lesion blanched with diascopy and urticated with stroking. The child's growth and development parameters were normal for her age. There was no dermatographism, or pain on palpation of the abdomen, bones, and joints.

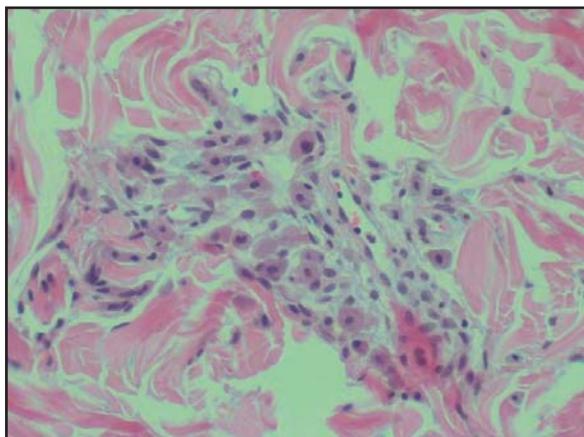
Laboratory investigations showed leukocytosis (eosinophilia). Skull X-ray, chest X-ray, abdominal ultrasound and liver - renal function tests, serum immunoglobulin levels and complement levels were normal.



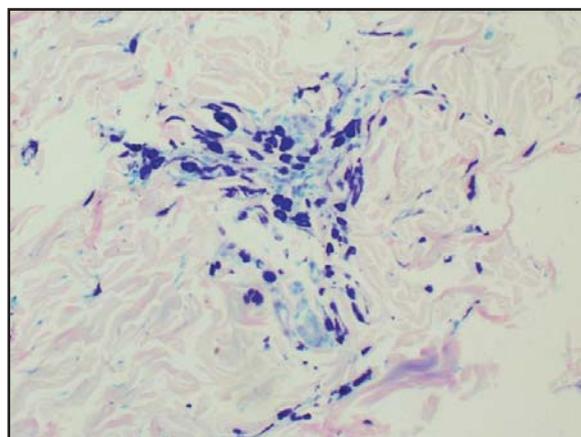
**Figure 1.** Widespread brown-red macules and papules on the trunk

A skin biopsy taken from one of the brown-red papules showed infiltration of fusiform cells with rounded nuclei and eosinophilic cytoplasm in perivascular location in the upper dermis. Giemsa and toluidin blue stainings showed abundant cytoplasmic metachromatic granules (**Figure 2 a, b**).

What is your diagnosis?



**Figure 2a.** Skin biopsy shows infiltration of mast cells around blood vessels (HE x 100).



**Figure 2b.** Skin biopsy shows infiltration of mast cells around blood vessels (Giemsa stain x100)

## Cutaneous Mastocytosis (Urticaria Pigmentosa)

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### Abstract

**Observations:** A 15-month-old girl presented with brown-red macules and papules all over her body. The child's mother reported that the macules, which first appeared 6 months ago, became red and swollen when scratched or rubbed, and decreased in size as the child grew. Dermatologic examination of the skin revealed multiple, widespread, oval/round brown-red macules and papules on the posterior trunk and extremities except the soles, palms, and mucous membranes. The diagnosis of mastocytosis was confirmed with histopathological examination.

### Discussion

Mastocytosis is a disorder characterized by mast cell accumulation in the tissue, most commonly in the skin, and it may also affect the bone marrow, skeletal system, gastrointestinal tract, liver, spleen, and lymph nodes. Mastocytosis is subdivided into two groups of disorders. Cutaneous mastocytosis which is defined by abnormal mast cell accumulation in the skin and systemic mastocytosis which is described by the forms of mastocytosis where mast cells infiltrate extracutaneous organs, with or without skin involvement [1].

Urticaria pigmentosa (UP), the most common type of cutaneous mastocytosis, is characterized by aggregates of mast cells in the dermis, leading to the development of multiple red-brown hyperpigmented macules, papules, or nodules.

There are two types of urticaria pigmentosa. In the classic type (infantile onset), lesions are present at birth or erupt during the first two years of life. Lesions may be macular, nodular, papular, vesicular, or bullous and often have a symmetric distribution. The palms and soles are spared.

In the nonclassical type (adult onset), the lesions are similar to the classic type and may

develop at any time from infancy to adulthood. The lesions do not resolve, new lesions continually develop, and systemic involvement is more common [2].

Childhood disease is usually confined to the skin and is believed to be related to a transient dysregulation in local growth factors. Adult-onset UP involves systemic disease in up to 30% of patients, is less likely to have prominent cutaneous involvement, and is commonly associated with activating c-Kit mutations [3].

Though males and females are reported to be affected equally; the male predominance was pronounced in the UP subgroup. According to the literature, 65% of all cases of mastocytosis begins in childhood, with approximately occurring between the ages of birth and two years of age [4]. UP appears to occur sporadically; however, familial inheritance has been reported in 50 families since the mid-1880s. But no pattern of inheritance has been identified [5].

The clinical features of UP, patients have several to many hundred distinctive red-brown macules, patches and occasionally papules [1]. Initial lesions involve the trunk and then spread centrifugally and symmetrically [3]. The trunk is the most common site; the palms, soles, and face are usually spared [1].

example histamine, leukotrienes, heparin, platelet activating factor, prostaglandins, proteases, tumor necrosis factor eg) [3]. The most frequently signs and symptoms are flushing and pruritus [2]. The association of UP with malignancy has been reported, but it appears to be less frequent in children.

Typically, the diagnosis of pediatric-onset mastocytosis is based on the clinical appearance, characteristic skin lesions and positive Darier sign [4]. Rubbing of a cutaneous mastocytosis lesion within a few minutes results in the formation of a wheal or even a vesicle. This characteristic response is known as Darier's sign and is considered clinically diagnostic [1].

The diagnosis of UP made clinically, it should be confirmed by histopathology. Mast-cell infiltration, predominantly surrounding blood vessels in the papillary and upper reticular dermis, confirms the diagnosis. Mast cells can be identified by various stains including Giemsa, toluidine blue and chloracetate esterase (Leder stain) or monoclonal antibodies to tryptase. Additional investigations, including complete blood count and measurement of serum tryptase, is recommended in disease onset in adolescents or adults or if evidence of systemic disease is present [1].

The disease usually has a benign course and disappears by puberty in about half of the affected children. Although constitutional symptoms as well as several laboratory findings have been reported to be associated with poor prognosis.

Treatment of UP is aimed at relieving symptoms and includes medications and avoidance of the aforementioned triggers that

can lead to mast-cell degranulation. Medications such as topical corticosteroids, H1 and H2 antihistamines and cromolyn sodium, and psoralen ultraviolet A (PUVA) photochemotherapy have been found to improve symptoms [5]. In this patient, general recommendations (avoidance of physical and chemical stimuli such as scratching of the skin, sudden changes in temperature and codeine) were provided and treatment with antihistamines (dexchlorpheniramine maleate, 0.35 mg/kg given in divided doses in every 6 h) was started. Control of the symptoms was obtained two month later.

In our patient, disease occurred in the ninth month of life. Clinical feature is typical for UP, oval/round brown-red macules and papules on the posterior trunk and extremities. Darier's sign was positive. The pathohistological examination confirmed the diagnosis of UP.

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