Localized Grain-Leather Plaque in Urticaria Pigmentosa – An Unusual Coexistence of Dual Morphology

Abstract

Urticaria pigmentosa (UP), the most common variant of cutaneous mastocytosis (CM), is characterized by hyperpigmented, brownish macules and patches, with positive Darier's sign. Diffuse cutaneous mastocytosis (DCM), is a rare variant of CM, characterized by diffuse infiltration of the skin by mast cells (MCs), resulting in a typical grain-leather appearance. Blistering is commonly seen in infants with CM but it is not specific for any variant of CM. Herein, we report a case of a 5-month-old male infant who presented with recurrent bullous lesions and hyperpigmented, brownish patches with intervening normal skin suggestive of UP, as well as a single, localized plaque with grain-leather appearance, thereby exhibiting an interesting dual morphology.

Keywords: *Cutaneous mastocytosis, grain-leather appearance, urticaria pigmentosa*

Sanjiv Choudhary, Ankita Srivastava, Deepti Joshi[1], Santosh Tummudi[2]

Department of Dermatology, All India Institute of Medical Sciences, Nagpur, Maharashtra, [1]Department of Pathology, All India Institute of Medical Sciences, Bhopal, Madhya Pradesh, [2]Department of Pathology, All India Institute of Medical Sciences, Mangalagiri, Andhra Pradesh, India

Introduction

Mastocytosis is a rare acquired condition characterized by pathological increase and accumulation of mast cells in the skin and other tissues. Broadly, it is classified into cutaneous and systemic forms. Urticaria pigmentosa (UP) is regarded as the commonest form of cutaneous mastocytosis (CM) while diffuse cutaneous mastocytosis (DCM) is very rare.[1] Herein, we report a case of urticaria pigmentosa in an infant, who also had a localised lesion morphologically similar to DCM, thereby exhibiting dual morphology.

Case Report

A 5-month-old male infant born out of nonconsanguineous marriage presented with clear fluid-filled blisters, on and off, all over the body since birth. The baby was born in a hospital through normal vaginal delivery and weighed 2.9 kg at birth. The mother was primigravida and the antenatal, perinatal, and postnatal period was uneventful. He was being exclusively breastfed and had received all vaccines as per the immunization schedule. There was no delay in developmental milestones. There was no history of flushing, diarrhea, vomiting, or respiratory complaints. On examination baby appeared healthy, vitals were within normal limits and body weight was 5.6 kg. There was a tense bulla about 4 × 3 cm in size over an infiltrated plaque on the medial border of left foot [Figure 1b]. Multiple well-defined round-to-oval hyperpigmented patches were present over the trunk with intervening normal skin [Figure 1a]. A well-defined plaque with a grain-leather appearance was present on the dorsal aspect of the left hand [Figure 2]. Palms, soles, and mucous membranes were spared. Darier's sign was positive over the pigmented lesion of back and on the plaque over the dorsum of the left hand [Figure 1a]. All routine investigations including hemogram, liver,

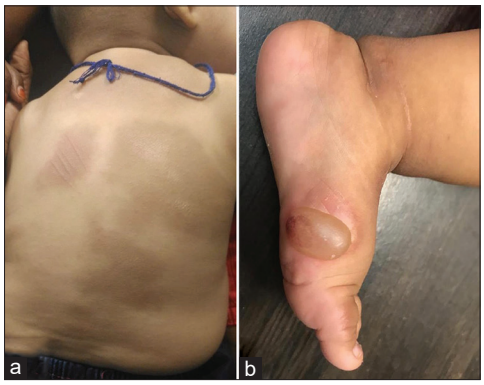

Figure 1: (a) Multiple hyperpigmented patches over back with positive Darier's sign. (b) Tense bulla over the medial border of the left foot

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Address for correspondence:
Dr. Sanjiv Choudhary, Department of Dermatology, All India Institute of Medical Sciences, Nagpur, Plot No. 2, Sec- 20, MIHAN, Nagpur - 441 108, Maharashtra, India.
E-mail: sanjiv.derma26@gmail.com

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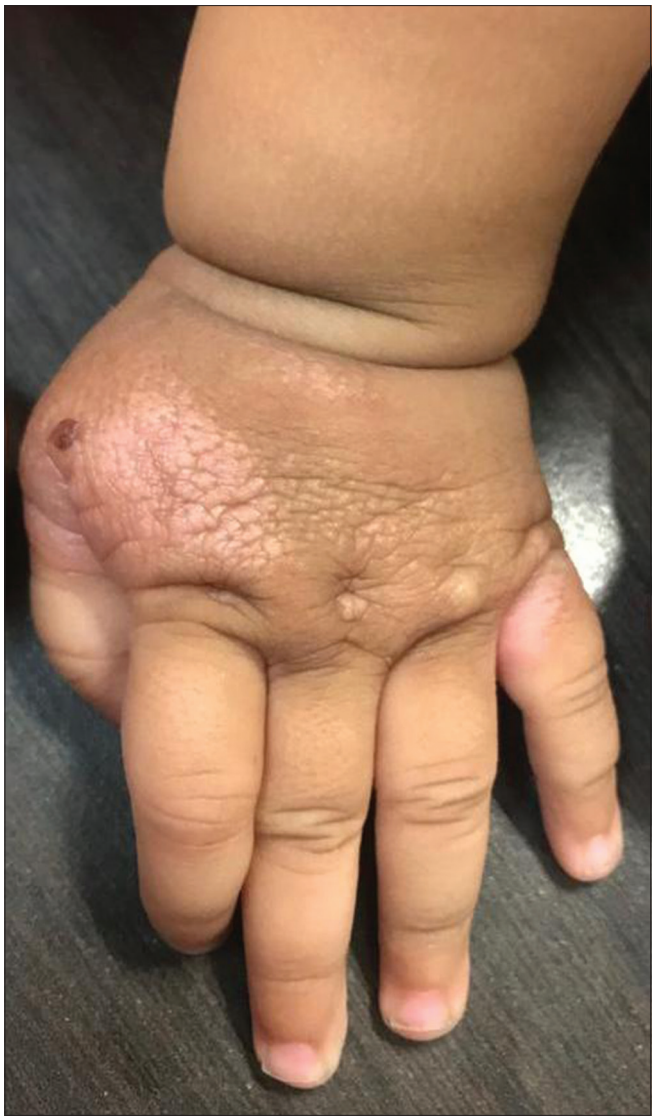

Figure 2: Single, well-demarcated plaque with a grain-leather appearance on the dorsal aspect of the left hand

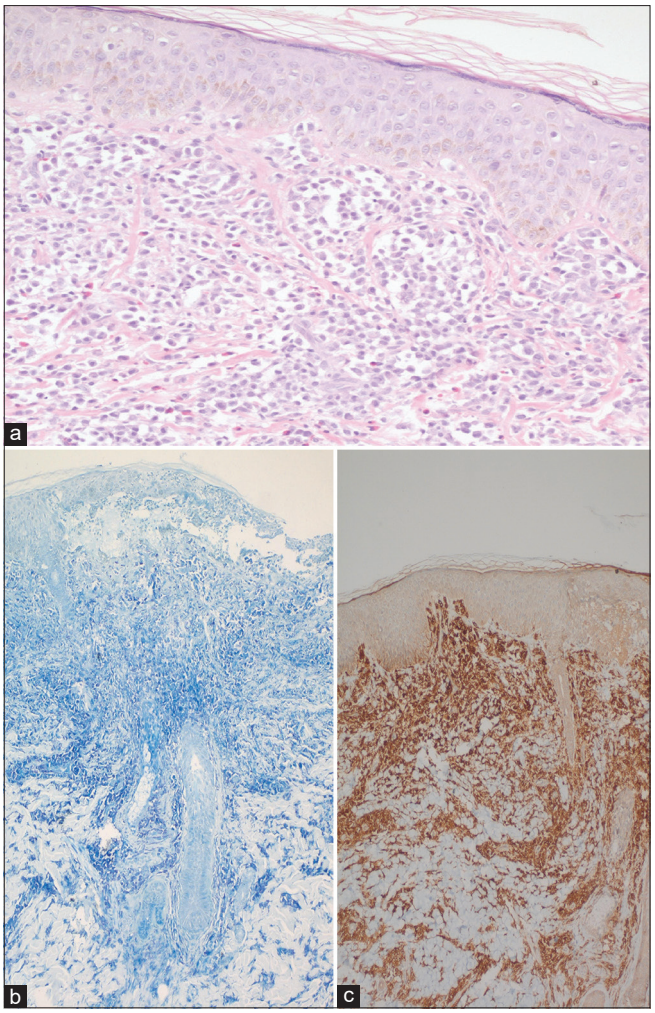

Figure 3: (a) Dermis showing nodules and sheets of mast cells with large, pale nuclei, and eosinophilic granular cytoplasm along with dermal edema, sclerosis, and few blood vessels (H and E, 40×) (b) Mast cells in the dermis (Toluidine blue, 10×) (c) Immunohistochemistry showing CD 117 positivity (10×)

and kidney function tests were normal but serum tryptase level was raised (42 ng/mL). VDRL test of the mother was nonreactive. An ultrasound of the abdomen did not reveal any organomegaly.

Skin punch biopsy from an infiltrated plaque over chest showed an unremarkable epidermis and nodules and sheets of MCs in the dermis with large pale nuclei and eosinophilic granular cytoplasm along with dermal edema, sclerosis, and few blood vessels. Toluidine blue stain for mast cells was positive. Immunohistochemical examination indicated CD117 positivity [Figure 3a-c].

Based on clinical and histopathological findings, a diagnosis of cutaneous mastocytosis was established. The patient had been prescribed hydroxyzine drops, at a dose of 0.6 mg/kg and prednisolone drops, at a dose of 0.5 mg/kg, which was tapered over 3 weeks, however, the patient was later lost to follow-up.

Discussion

DCM is a rare form of mastocytosis characterized by MCs infiltration of the entire skin. Its reported frequency varies from 0.6% to 8%.[2] DCM can present with widespread blistering, erosions, erythroderma, or thickening of the skin. It can be associated with mast cell mediator-related symptoms such as flushing, itching, blistering, diarrhea, abdominal pain, vomiting, hypotension, bronchospasm, headache, and anaphylactic shock.[2,3]

Different morphological variants of DCM include large hemorrhagic bullous variant and infiltrative small vesicular variant in infants.[2,4] Besides, pseudoxanthomatous lesions, large mastocytomas, and pachydermia with extensive skin folding have also been reported in older children and adults.[2]

Our case is unique since the infant presented with multiple hyperpigmented patches with intervening normal skin, with a positive Darier sign that pointed towards the diagnosis of

urticaria pigmentosa. On the other hand, he also had a single, well-demarcated plaque with characteristic grain-leather appearance. Such appearance involving extensive areas of skin is characteristically seen in cases of DCM.[5] Such typical appearance is explained on the basis of infiltration of mast cells and edema resulting from their degranulation products.[5] In DCM, grain-leather appearance is seen later in life while blistering is commonly seen in infancy.[2] In the present case, blistering was seen along with a localized grain-leather appearing plaque, which is an unusual presentation.

Our case did not present with any systemic feature, though serum tryptase level was slightly raised. Serum tryptase level has been noticed to decline after the age of 2 years.[2] We did not go for a bone marrow biopsy at this stage, as the baby seemed to be healthy and serum tryptase was far below the cut-off level (100 ng/mL) recommended for bone marrow examination.[2,6]

Blistering is commonly seen in infants with CM but it is not characteristic of any variant in particular.[7] In our case, the patient had hyperpigmented lesions with intervening normal skin clinically suggestive of urticaria pigmentosa and localized grain-leather plaque, which is usually seen in DCM but with extensive body area involvement. We report this case on account of unusual dual morphology which has not been reported previously.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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