

Case Reports:

Urticaria Pigmentosa in infant

Sanjay N. Agrawal, Sujata J. Agrawal, Subodh D. Jane, Pranita P. Daware, Anuprita A. Rawal

Corresponding Author: Dr. Subodh D. Jane, Dept of Dermatology, Dr. Panjabrao Deshmukh Memorial Medical College, Amravati. Email – dr.subodhjane86@gmail.com

Received: 11, March, 2019; **Reviewed:** 3 May 2019; **Accepted:** 9, May, 2019.

Citation of article: Sanjay N. Agrawal, Sujata J. Agrawal, Subodh D. Jane, Pranita P. Daware, Anuprita A. Rawal. *Urticaria Pigmentosa in infant*; New Indian Journal of Pediatrics, 2019; 8.3.

Abstract:

A 6 months old male presented with recurrent multiple, diffuse, brownish macula-papular lesions all over the body since 3 months. The lesions on gentle stroking showed appearance of wheal and erythema. Histopathological examination revealed collection of mast cells in upper dermis suggesting a rare condition of cutaneous mastocytosis.

Keywords: *Cutaneous mastocytosis, Urticaria Pigmentosa, Darier's sign*



Figure 1: Anterior view of patient showing multiple diffuse, brownish macules, patches and papules with post-inflammatory hyperpigmentation.

Introduction:

Mastocytosis is an uncommon condition characterized by abnormal growth and accumulation of mast cells. This could be systemic or limited to skin with resultant various manifestations. WHO classifies the mastocytosis as cutaneous mastocytosis, indolent systemic mastocytosis, systemic mastocytosis with an associated clonal hematologic non mast cell lineage disease, aggressive systemic mastocytosis, mast cell leukemia, mast cell sarcoma and extra cutaneous mastocytoma (1) . Urticaria pigmentosa (UP) is the commonest and benign type of cutaneous mastocytosis. Onset is usually before the age of 2 years, but can also appear in later age. Multiple, reddish-brown macula-papular lesions with plaques or nodules appear in symmetrical distribution with predilection for the trunk. The extremities and face are generally involved and palms and soles tend to be spared. The lesions urticate within 2 to 5 minutes on gentle rubbing and stroking which may remain for 30 minutes to few hours this is known as Darier's sign. We present a patient who has the classical history, lesions and positive Darier's sign.

Case report:

A 6 months old male child presented with multiple brownish lesions over trunk, extremities, head and face since 3 months. The lesions started as erythematous macules which at places turned into papules, plaques and sometimes into nodules. The lesions were transient and disappeared on their own after some time leaving pigmentation at some places. The mother also noticed appearance of swelling after massage or rubbing of the lesions. No history of similar complaints in family. On examinations there were multiple, diffuse, brownish macules and papules present over trunk, extremities, head, face and scalp [Fig. 1] [Fig. 2]. On gentle stroking over the lesions wheals and



erythema developed (Darier's sign). At places there were post-inflammatory hyperpigmentation [Fig. 2]. The mucosa, palms and soles were spared. Systemic examination and routine blood investigations were normal. With the characteristic lesions in a particular distribution in this age group a diagnosis of Urticaria Pigmentosa with a differential diagnosis of papular urticaria was made. The patient was investigated for any mast cells in peripheral smear along with the routine blood investigations which was negative. A confirmatory skin biopsy was done which showed focal collection of polygonal cells with round or elongated nuclei and abundant deep eosinophilic cytoplasm suggestive of mast cells in upper dermis confirming cutaneous mastocytosis. Dermal adnexal structures appeared normal. The patient was given oral anti-histamine and the mother was explained the course, the prognosis of the condition and precautions like avoid rubbing and massage.

Figure 2: *Posterior view of patient showing involvement of scalp and post-inflammatory hyperpigmentation.*

Discussion:

Urticaria Pigmentosa is an uncommon disorder of cutaneous mastocytosis usually seen below 2 years of age (2). There is presence of characteristic lesions showing Darier's sign. Systemic involvement is absent in majority of the patient. It can also mimic urticaria or papular urticaria. Papular urticaria is seen on exposed parts while Urticaria Pigmentosa mainly involves trunk. Urticaria is mainly characterized by wheals and will not leave behind any post-inflammatory pigmentation. Prognosis is usually good in children. The presented case is a typical case of Urticaria Pigmentosa and being reported because of rarity. (3)

Declaration:

Contribution of authors: All authors participated in the analysis of the presented case report.

Conflict of Interest: None

Ethical Approval: The study approved by the Institutional Ethics Committee

Funding: Self

References:

1. Valent P: World Health Organization Classification of Tumors: Pathology and Genetics of Tumors of the Hematopoietic and Lymphoid Tissues, edited by Jaffe ES. Lyon, IARC Press, 2001
2. C.E.H. Grattan & A. Kobza Black. Urticaria and Mastocytosis. In: Tony Burns, editor. Rook's Textbook of Dermatology, 8th ed. UK: Wiley Blackwell Publishers; 2010. p. 22.31
3. Ritambhra, Mohan H, Tahlan A. Urticaria pigmentosa. Indian J Dermatol Venereol Leprol 2001;67:33