Original Research Paper

Volume - 14 | Issue - 09 | September - 2024 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Dermatology

UNVEILING THE UNIQUE: A COMPREHENSIVE CASE STUDY OF URTICARIA PIGMENTOSA

Iswarya Shree Nee PA*

3rd year Postgraduate, M.D. (DVL), S.V. Medical College, Tirupati. *Corresponding Author

ABSTRACT Cutaneous mastocytosis, characterized by mast cell hyperplasia in the skin, commonly manifests as urticaria pigmentosa. Over 50% of children are diagnosed prior to two years of age. This condition primarily affects the skin and often resolves spontaneously by adolescence. Despite its clinical significance, the precise etiology of cutaneous mastocytosis remains elusive, although current understanding implicates cytokine-mediated mast cell proliferation. We present a case of a 1.5-year-old female with multiple brownish macules across her body, accompanied by a history of erythema and wheal formation upon gentle stroking or toweling. Clinical examination revealed hyperpigmented macules and papules on the trunk and limbs, with wheal and erythema formation upon lesion stimulation (darier sign). Histopathological examination confirmed mast cell infiltration and edema, consistent with cutaneous mastocytosis. Laboratory investigations were unremarkable. Treatment with desonide lotion and chlorpheniramine maleate, along with parental education on skin care practices, was provided. After 4 weeks, the child exhibited only residual pigmentation and was kept on regular follow-up.

KEYWORDS : *Cutaneous mastocytosis*, urticaria pigmentosa,mast cell proliferation, darier sign

INTRODUCTION

Mastocytosis, a hematopoietic disorder, typically occurs sporadically and is characterized by an increased number and accumulation of mast cells in one or more organs. It can manifest as either cutaneous mastocytosis or systemic mastocytosis.

Cutaneous mastocytosis encompasses four clinical subtypes: urticaria pigmentosa, mastocytoma, diffuse cutaneous mastocytosis, and telangiectasia macularis eruptiva perstans (TMEP). Among these, urticaria pigmentosa is the most prevalent in children. Clinically, the disease is marked by multiple erythematous and pigmented macules, papules, and plaques, often accompanied by localized blistering of varying sizes.[1]

Over 50% of cases in children are diagnosed before the age of two, and the condition generally remains confined to the skin. Diagnosis is typically confirmed through histopathological examination, considered the gold standard. The presence of a positive Darier's sign aids clinical diagnosis. Most children experience spontaneous resolution by adolescence.[2]

Symptomatic treatment involving antihistamines, mast cell stabilizers, and topical steroids is effective in managing symptoms. Herein, we present a typical case of this dermatosis.[3]

CASE REPORT

A one and half year old female child presented with multiple brownish macules all over body for the past one year. Lesions initially started over trunk and further progressed to involve the upper limb and face. There was history of erythema and wheal formation on toweling and massaging the child. There were no associated systemic symptoms. The personal and family history was not contributory.

On examination, there were multiple hyperpigmented macules and a few papules on the face, neck, trunk, and limbs. Gentle stroking of individual lesions induced wheal and erythema, indicative of Darier's sign. These reactions were accompanied by itching and typically subsided within 15-20 minutes. There was no hepatosplenomegaly or lymphadenopathy.

Laboratory investigations, including a complete blood count, liver and renal function tests, urine analysis, and chest radiography, revealed results within normal limits.

Histopathological examination of lesional skin showed a dense superficial and mild perivascular infiltrate of lymphocytes and mast cells with moderate epidermal hyperplasia and hyperpigmentation with marked upper dermal edema. Extracellular mast cell granules are also seen. Giemsa stain confirmed the presence of mast cells, consistent with a diagnosis of urticaria pigmentosa.

The child was treated with desonide applied twice daily application and chlorpheniramine maleate. Necessary advice was given to parents regarding avoidance of excessive scrubbing and massaging of the skin. After 4 weeks, the child exhibited only residual pigmentation and remains under regular follow-up.

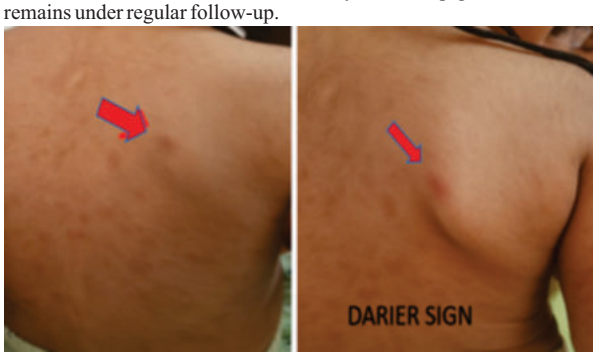

Figure1: Positive Darier sign observed after stroking a hyperpigmented macule.

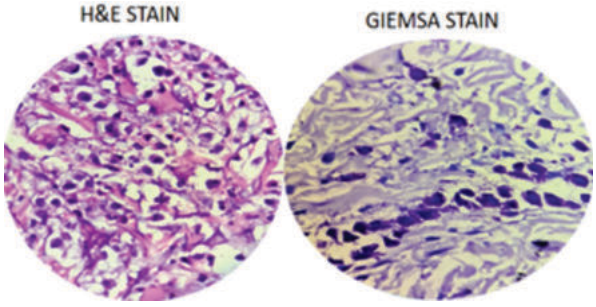

Figure2: Histopathology of the skin lesion showing mast cells with extracellular granules.

DISCUSSION

Urticaria pigmentosa represents the most common form of cutaneous mastocytosis. There is no sex predilection and occurs more often in infants and children compared to adults. Clinical manifestations typically arise within the initial two years of life. Characteristic features include multiple oval or round hyperpigmented macules, papules, or patches, ranging in color from brown-red to yellow and measuring 2-4mm in diameter.[1]

These lesions demonstrate urtication upon manipulation (e.g., rubbing) or may occur spontaneously, a phenomenon known as Darier's sign. Darier's sign may not always be evident, particularly in individuals with a prolonged history of the disorder, and its specificity for mastocytosis is not absolute. Rare occurrences have been documented in conditions such as Juvenile xanthogranuloma and acute lymphoblastic leukemia in neonates. [4]

The sites of predilection are the chest and dorsal areas of the body, while the palms, soles, and face are usually unaffected. Systemic manifestations include flushing, vomiting, diarrhea, tachycardia,

headache, weight loss, and wheezing.[5]

Histologically there is mild to moderate perivascular infiltrate with dendritic mast cells in the papillary dermis; a band like infiltrate or sometimes even nodular infiltrates extending to the subcutis may be seen, especially with special stains like toluidine blue and chloroacetate esterase.

Management of urticaria pigmentosa primarily revolves around symptomatic treatment and avoidance of known mast cell degranulation triggers, including physical stimuli like rubbing, as well as alcohol, morphine, codeine, NSAIDs, radiocontrast media, and scopolamine. Antihistaminics are the first step medications among systemic treatment options.[3]

CONCLUSIONS

In conclusion, this case report underscores the clinical significance and diagnostic sequence of urticaria pigmentosa, the most common form of cutaneous mastocytosis. The presentation of multiple hyperpigmented macules and papules, coupled with the characteristic Darier's sign, highlights the importance of clinical recognition in infants and children, where the condition predominantly manifests.

Successful management, as demonstrated in this case, relies on a combination of symptomatic treatment and avoidance of mast cell degranulation triggers. Early intervention with antihistamines and patient education regarding trigger avoidance are crucial in mitigating symptoms and preventing potential systemic complications. Regular follow-up ensures monitoring of disease progression and response to therapy, leading to favorable outcomes, as evidenced by the significant improvement observed in our patient's condition.

This report contributes to the existing literature by providing insights into the clinical presentation, histopathological findings, and management strategies of urticaria pigmentosa in pediatric patients, thereby enhancing awareness and facilitating optimal care for affected individuals.

REFERENCES:

[1] R, Mohan H, Tahlan A. Urticaria pigmentosa. Indian J Dermatol Venereol Leprol 2001;67:33-34
[2] Caplan RM. The natural history of urticaria pigmentosa. Analysis and follow up of 112 cases. Arch Dermatol 1963; 87: 146- 148.
[3] Keen, M. A. (2016). Urticaria pigmentosa: A case report. Nepal Journal of Dermatology, Venereology &Amp; Leprology, 13(1), 57–60.
[4] Nagayo K, Sakai M, Mizuno N. Juvenile xanthogranuloma with Darier's sign. Int J Dermatol 1983; 10:283-5.
[5] Turchin I, Barankin B, Schloss E. Unusual cutaneous findings of urticaria pigmentosa and telangiectasia macularis eruptiva perstans associated with marked myelofibrosis. Int J Dermatol 2006; 45: 1215-7.