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Pathology

URTICARIA PIGMENTOSA- A CASE REPORT

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ABSTRACT

INTRODUCTION - Urticaria Pigmentosa is rare cutaneous disorder. It is the prototype of mast cell disease. This name is given to a type of cutaneous mastocytosis, in which there're yellow-brown hyperpigmented patches on skin due to abnormal collection of mast cells. Urticaria pigmentosa has clinically diagnostic Darier Sign positivity and should be confirmed by histopathology.

CASE REPORT - We present a case of 2-year-old boy, who came to the OPD of Department of Dermatology. He presented with yellow brown hyperpigmented macules on whole body. Skin biopsy of a lesion from trunk was taken and specimen stained by Haematoxylin and Eosin and Giemsa stain for mast cells.

CONCLUSION - Skin biopsy demonstrated diffuse infiltration of mast cells, lymphocytes and eosinophils in papillary dermis. Giemsa stain, which stains the mast cells' granules, confirmed the mast cells in dermis.

KEYWORDS : Mast cells, Darier sign, Giemsa stain.

• INTRODUCTION -

Mast-cell disease is a rare disorder, primarily of childhood, that is usually self-resolving. Approximately two-thirds of cases are limited to the skin (1).

Mast cells were first described by Erhlich (2, 3). Mast cells are normally widely distributed in the skin. They contain granules that contain histamine and other mediators of inflamation. When the mast cell is irritated, these chemicals are released into the surrounding skin, causing leaking of the blood vessels, resulting in localized itching, swelling and redness (4).

Mastocytosis is defined as an overproliferation and accumulation of tissue mast cells.

The mastocytosis can present as cutaneous (CM) or systemic lesions (SM).(4)

The salient differentiating points of these lesions are as follows –

CUTANEOUS MASTOCYTOSIS(CM)	SYSTEMIC MASTOCYTOSIS(SM)
The diagnosis of Cutaneous Mastocytosis is based on typical clinical and histological skin lesions and absence of definitive signs (criteria) of systemic involvement.	The diagnosis of Systemic Mastocytosis is based on multifocal histological lesions in the bone marrow (affected almost invariably) or other extracutaneous organs (Major criteria) together with cytological and biochemical signs (Minor criteria) of systemic disease(SM-criteria)
Most patients with Cutaneous Mastocytosis are children and present with maculopapular yellow-brown,hyperpigmented patches(Urticaria pigmentosa-UP).	Most patients with Systemic Mastocytosis are adults. SM is further devided into the following categories-
Other less frequent form of CM is- Diffuse cutaneous mastocytosis(DCM) Mastocytoma of skin	(1) Indolent Systemic Mastocytosis(ISM) (2) SM with an associated clonal hematologic non-mast cell lineage disease(AHNMD) (3) Mast cell leukemia

Urticaria pigmentosa is the prototype of mast cell disease. This name is given to a type of cutaneous mastocytosis, in which there are yellow-tan to reddish-brown patches on the skin due to abnormal collections of mast cells. (5, 6). It is the most common skin manifestation of mastocytosis in both children and adults (7).

Urticaria pigmentosa was first described by Nettleship and Tay in 1869 (8), who recognized the classic cutaneous form of disease now known as urticaria pigmentosa. Urticaria pigmentosa is the most frequent manifestation of cutaneous mastocytosis.

Unna in 1887 (9) demonstrated the relationship between urticaria pigmentosa and the mast cells.

The lesions of urticaria pigmentosa appear as small yellow-tan to reddish-brown macules or slightly raised papules disseminated on the skin of the body. (5). Usually the first skin lesions appear at a few months age. Mild trauma, scratching or rubbing of the lesion may led to urtication and erythema around the lesion which is known a Darier's sign. The condition is accompanied by variable degree of pruritus (5).
Urticaria pigmentosa most often affects infants. In more than 50%, the onset is before 2 years of age (10). Usually the first skin lesions appear at a few months of age. They persist and gradually increase in number for several months or years. They can appear on any part of the body including the scalp, face, trunk and limbs. There is no sex preference (6,7,12,10). Clinical feature consists of reddish or brownish patches on the skin, with dissemination of variable density. Trunk is the most common site of the lesions but they can also present on the other parts of the body. Usually variable amount of pruritus may be present (6,7,12,10).

Over the next few years the urticaria pigmentosa becomes less irritable and eventually the patches fade away. By the teenage years, most patches will resolve spontaneously (6,7,10).

• CASE REPORT:

24-month-old boy came to skin OPD of MDM Hospital Jodhpur. He presented with disseminated yellow-brownish, hyperpigmented patches (1x2 cm in diameter) (fig. 1) all over the body. No subjective symptoms were present. First skin lesion occurred as reddish infiltrates in the sixth month of life on the skin of the trunk and upper extremities. Therapy with topical corticosteroids prescribed by the general practitioner showed no effect. Lesions have spread to the skin of the entire body including the scalp (Fig. 1). Darier's sign was positive & diagnosis of Urticaria Pigmentosa was made. Skin biopsy of a lesion from the trunk was taken. Specimens were stained by haematoxylin-eosin (HE), and Wright Giemsa method for metachromatic granules of mast cells (17). HE staining (fig.-2) showed massive mast cells and few lymphocytic infiltrates in superficial dermis, Section also showed perivascular infiltrate comprising of mast cells and few lymphocytes. The overlying epidermis was hyperpigmented. The mast cells showed metachromatic granules with Giemsa stain (fig-3). Histopathological examination confirmed diagnosis of mastocytosis.

• DISCUSSION

In our patient, disease occurred in the second month of life. Clinical feature is typical for urticaria pigmentosa, including reddish-brown macules and slightly raised plaques on the skin of the entire body. Lesions were present also on the skin of the neck and scalp which is not

so usual. Darier's sign was positive. Blood cell count was within normal values, which is in accordance with cutaneos mastocytosis, while in systemic mastocytosis, it may reveal anaemia, thrombocytopenia, thrombocytosis, leukocytosis, and eosinophilia (7,10).

The histopathologic examination of lesions confirmed the diagnosis of Urticaria Pigmentosa.

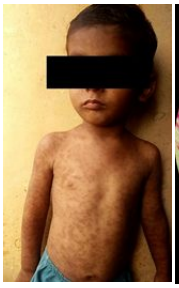
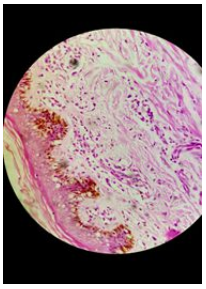
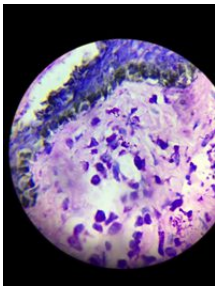

Fig. 1 **Fig. – 2 H&E X 40x** **Fig.3 – Giemsa stain**

In cutaneous mastocytosis, release of histamine from their granules, leads to urticarial edema of the lesions, which results in secondary hyperpigmentation due to melanocytic activity at the dermoepidermal junction (11). This is reflected in the histopathological examination.

Histological features characteristic of secondary syphilis were not seen.

And absence of diffuse infiltration of histiocytes and granulomas ruled out Histiocytosis X and Sarcoidosis.

• CONCLUSION

The clinical features, positive Darier's sign and histopathological examination confirmed the diagnosis of Urticaria pigmentosa. Since this is a form of cutaneous mastocytosis, without systemical involvement, prognosis is good and it can be expected that lesion will resolve during next few years.

This study highlights the important role of Histopathological examination for confirmatory diagnosis of Urticaria Pigmentosa.

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