

A rare ichthyosiform variant of large plaque parapsoriasis : a report in a young female

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ABSTRACT

Large plaque parapsoriasis is a chronic papulosquamous dermatoses of unknown etiology affecting middle aged individuals of all races and geographic area. It is considered as benign end of mycoses fungoides disease spectrum. Poikilodermatous and retiform are the two known types of LPP closely resembling the patch stage of mycosis fungoides. Herein, we report a young female with ichthyosiform variant of LPP which is a rare entity.

Key words : Ichthyosiform, young female, large palque parapsoriasis, mycosis fungoides.

Introduction :

Parapsoriasis is a group of uncommon papulosquamous disorder characterized by persistent scaly eruptions. The mean age of onset is in the fifth decade. Parapsoriasis comprises of 2 major types, large plaque parapsoriasis (LPP) and small plaque parapsoriasis. LPP is characterized by asymptomatic or mildly pruritic well defined oval or irregular plaques, usually more than 5 cm in diameter with scaling. The two variants of LPP commonly seen in clinical practice are poikilodermatous and retiform. However ichthyosiform LPP is another variant rarely reported in all age groups. There are very few case reports of ichthyosiform variant of LPP in the age group lesser than 20 years of age. We hereby report a case of rare ichthyosiform variant of LPP in a 16 year old.

Case Report :

A 16 year old female presented with multiple dark scaly raised asymptomatic lesions over the back, shoulders and axillae since 4 years. Patient has been treated by many dermatologist in the form of topical corticosteroids, topical vitamin D analogue and moisturisers without any relief. There was no history of any topical application prior to onset of lesions. There was no lymphadenopathy and organomegaly

on systemic examination. Cutaneous examination revealed multiple well defined hyperpigmented thymotic patches measuring 5 X 5 to 10 X 12 cm over lower back, bilateral scapular region and bilateral anterior axillary folds. On nerve, sensory and motor examination no abnormality was detected. Routine haematological investigations were within normal limits. Haematoxylin and Eosin staining of the skin biopsy specimen showed hyperkeratotic epidermis, focal parakeratosis, lymphocytic exocytosis and increased basal layer pigmentation. Dermis shows focal interface change with mild perivascular lymphohistiocytic infiltrate (*Figure 2 a, b, c*). No cellular atypia was seen. Fite Faroco stain was negative for Acid fast bacilli. Patient was advised immunohistochemistry to rule out early stage of mycosis fungoides but she denied due to high cost. Based on the history, clinical and histopathological examination a diagnosis of ichthyosiform large plaque parapsoriasis was made. Patient was started on topical treatment in the form of emollient and planned for phototherapy. Two months later, patient showed only mild improvement with reduction in scaling but patches persisted. She is under regular follow-up to observe possible conversion into malignant disease.

Discussion :

Broc, in 1902 coined the term parapsoriasis which includes inflammatory dermatoses with characteristic common features such as unknown etiology, middle to late age of onset, clinically asymptomatic lesions and resistant to treatment¹. It

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Legends of Figures :

Fig. 1 : Multiple hyperpigmented ichthyosiform patches over lower back, scapular region and anterior axillary lines.

includes three entities : large plaque parapsoriasis (LPP), small plaque parapsoriasis (SPP), and pityriasis lichenoides (PL). LPP has a special importance as it is considered to be the benign end of the Mycosis Fungoides (MF) disease spectrum. Clinical presentation of LPP includes large, more than 5 cm scaly patches which are oval or irregular in shape. They are asymptomatic or mildly pruritic¹. It affects middle aged people in fifth decade with slight male predominance¹. Clinically LPP presents as : poikilodermatous and retiform pattern. There is a single case report of ichthyosiform pattern of large plaque parapsoriasis in a 36 year old male⁴. Our case also represents ichthyosiform variant of LPP that occurred at a very young age and hence mandates constant and regular follow-up in future. Das et al, in 2005, has reported an atypical presentation of LPP with generalised asymptomatic hypopigmented macules all over the body in a five-year-old child². This implies that there are changing patterns of LPP presentation in recent years which needs vigilant attitude to prevent missing accurate diagnosis. Immunohistochemistry shows similar feature in early MF lesion and LPP which includes epidermal

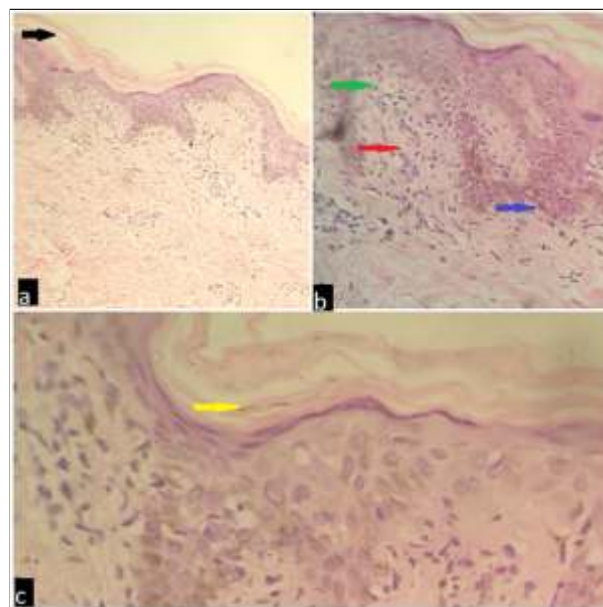


Fig. 2 : (a) scanner view, showing hyperkeratosis (black arrow) (H and E, X4). (b) lymphocytic exocytosis (green arrow), increased basal layer pigmentation (blue arrow), papillary dermis showing perivascular lymphocytic infiltrate. (H and E, X10). (c) epidermis with focal parakeratosis (yellow arrow) (H and E, X40) .

expression of human leukocyte antigen (HLA-DR), predominant CD4+ T cell subsets and CD7 antigen deficiency¹. Skin associated lymphoid tissue (SALT) includes T cells mediating inflammatory skin conditions. There is a continuous trafficking of these T cells between the skin and the T cell domains of peripheral lymph nodes through lymphatics and bloodstream and express cutaneous lymphocyte associated antigen (CLA)^{1,3}. Mycosis fungoides is a proven SALT T cell neoplasm. The SALT nature of MF tumor clones have been proved by PCR based tumor clonality assays. Trafficking of mycosis fungoides tumor cells have been detected in patients with early stage of disease with lesions clinically-pathologically consistent with LPP. Therefore in some cases LPP is a monoclonal proliferation of SALT T cells having the capacity to traffic between skin and extracutaneous sites^{1,3}. The chances of progression from LPP to definite MF varies from

10% to 35% per decade as observed in previous studies^{5,6}. It takes many years for this progression and hence a prolonged and watchful follow-up is needed in all cases of LPP. Histopathology of early lesion of LPP show only a superficial perivascular lymphocytic infiltrate⁷. As the lesion progresses, epidermis develops mild irregular acanthosis with mounds of parakeratosis with dense band of lymphocyte, basal vacuolar change, pigment incontinence and lymphoid epidermotropism. In all stages, the lymphocytic infiltrate composed of monoclonal T-cells⁷. First-line treatment options for LPP includes emollients, topical corticosteroids, sunbathing, broad and narrow band UVB phototherapy. Second-line treatments are topical bexarotene, imiquimod, mechlorethamine, carmustine, PUVA phototherapy excimer laser etc⁸. To conclude, LPP is an indolent disease and hence needs prolonged follow-up. We want to highlight ichthyosiform variant of LPP of which very few cases are reported. Early age of onset makes this case report a rare occurrence.

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