Lymphomatoid Papulosis: A Case Report

Abstract

Lymphomatoid papulosis (LYP) is a chronic CD 30 + lymphoproliferative disorder (LPD) which is characterized by chronic, recurrent, and self-healing papulonecrotic or papulonodular skin eruptions, which are clinically benign and histopathologically malignant. It can resolve spontaneously; however, long-term follow-up is essential as it can progress to malignant lymphoma in 10–20% of the patients. We hereby report a case of a 42-year-old male presenting with recurrent papulonecrotic lesions over the face, trunk, and extremities from the last 3 years which heal with post-inflammatory hyperpigmentation and atrophic scars with a history of treated pulmonary tuberculosis one year back. There was no systemic involvement. LYP, involving cosmetically sensitive area like face, is an infrequent finding.

Keywords: *Lymphomatoid papulosis, lymphoproliferative disorders, papulonecrotic lesions*

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Introduction

Lymphomatoid papulosis (LYP) is a chronic, recurrent, self-healing papulonecrotic or papulonodular skin disease. It is a part of the group of cutaneous CD30+ lymphoproliferative disorders (LPDs). It is associated with the increased risk of secondary lymphomas, such as mycosis fungoides and CD 30+ large cell lymphoma (LTCL) in approximately 10–20% of the patients.[1] The most intriguing feature of this disorder is its benign course and spontaneous resolution combined with aggressive histological characteristics closely resembling lymphoma. Follow-up is essential as it can progress to malignant lymphoma in a subset of patients. We are reporting a case of a 42-year-old presenting with recurring papulonecrotic lesions over face, trunk, and extremities.

Case Report

A 42-year-old male presented to the dermatology outpatient department with the complaint of recurrent mildly itchy raised lesions over the face and trunk from 3 years. It was gradual in onset and progressive. Few lesions were associated with mild pain and pus discharge. It was associated with fever. Each lesion would last for a few weeks, later healing by post-inflammatory pigmentation and minimal scarring. The lesions are used to heal spontaneously. The patient was diagnosed with pulmonary tuberculosis 1 year back for which he received 6 months of anti-tuberculoid treatment (ATT). The patient was otherwise healthy, and his family had no similar illness. On examination, there were multiple discrete skin colored to hyperpigmented papules and nodules over the face, trunk, and bilateral thigh [Figure 1a-c]. Few of the papules had necrotic crusting at center. The patient also had some residual hyperpigmented macules from previous lesions, with minimal scarring over the trunk. Left cervical lymph nodes were tender and matted. The rest of the clinical examination was unremarkable. Based on history and clinical examination, differential diagnosis of LYP, lupus miliaris disseminated faciei, papulonecrotic tuberculid, and insect bite hypersensitivity was kept.

Laboratory investigations were all within normal limits, and there were no atypical cells in the peripheral blood smear. Histopathological evaluation with hematoxylin and eosin stain revealed a focally thinned epidermis with dense pan-dermal lympho-histiocytic infiltrate creeping in between the collagen bundles and adnexa. Minimal focal lymphocytic epidermotropism is seen [Figure 2a and b].

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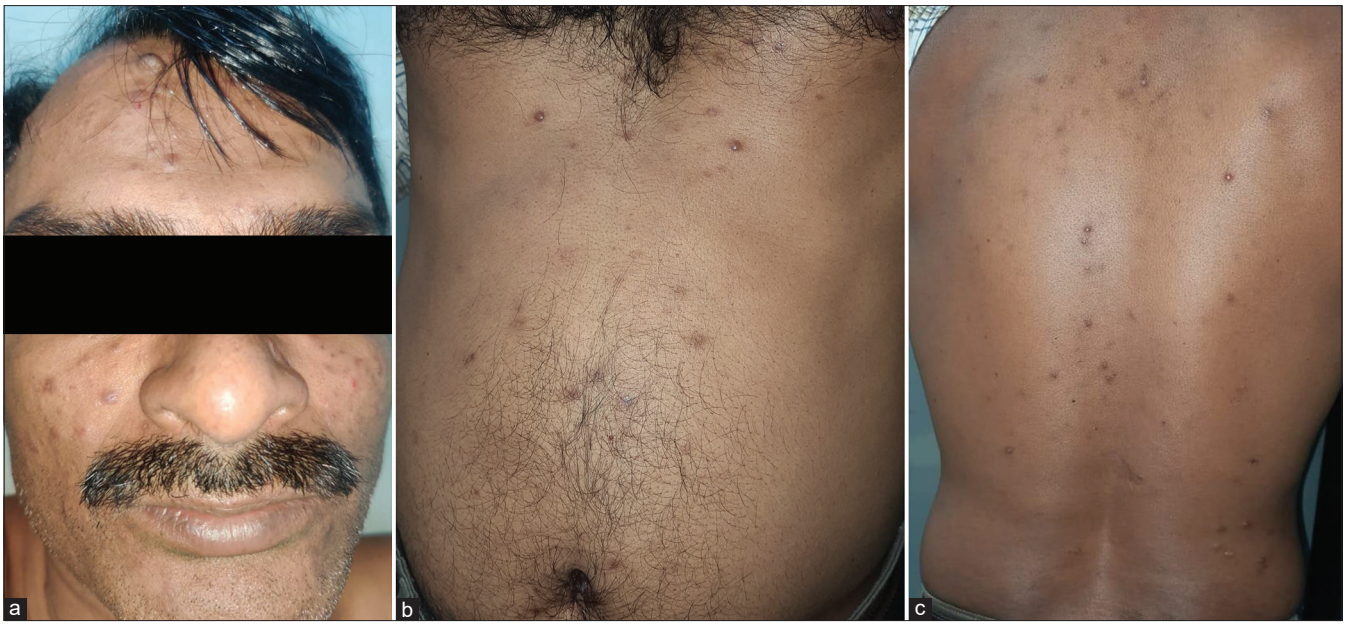

Figure 1: Multiple, discrete skin colored to hyperpigmented papules and few nodules over face (a) and trunk (b and c). Few papules over face have erosions and crusting present over it. Post-inflammatory pigmentation is present over the trunk

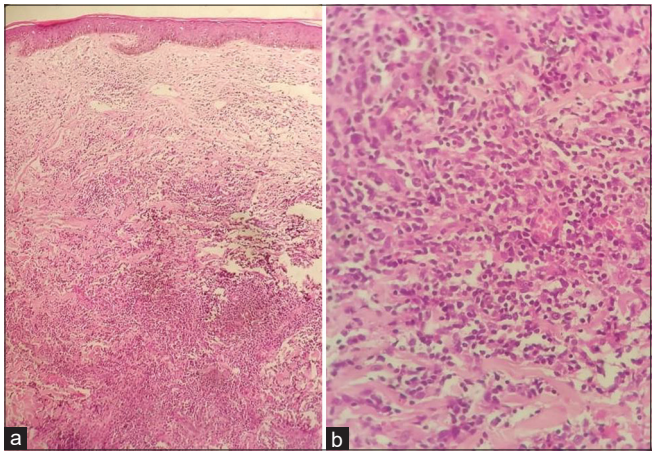

Figure 2: (a) Focally thinned epidermis with dense pan-dermal lympho-histiocytic infiltrate creeping in between the collagen bundles and adnexa (H&E, 10x). (b) Lymphohistiocytic infiltrates in the dermis (H&E, 40x)

On immunohistochemical evaluation, there is an admixture of CD 3 and CD 20 positive cells along with the presence of few CD 30 positive cells and numerous CD 68 positive histiocytes [Figure 3a-d]. The diagnosis of LYP type A was kept, and the patient was planned for low-dose methotrexate. The patient is currently under follow-up and is improving with few flare-ups. He was explained the possible risk of malignancy and the importance of lifelong follow-up.

Discussion

LYP is a rare cutaneous CD 30+ LPD which is characterized clinically by self-healing, recurrent papulonecrotic, or papulonodular eruptions with a waxing and waning course. They heal either with post-inflammatory hypo- or hyperpigmentation or atrophic scars. The term LYP was originally used by Macaulay in 1968 to describe "a self-healing rhythmical paradoxical eruption which is histologically malignant but clinically benign."[2] CD 30 + LPDs include LYP, primary cutaneous anaplastic large cell lymphoma (pcALCL), and borderline CD30+ lesions.[3]

The prevalence of LYP is 1.2–1.9 cases per million population. It can occur at any age, but the peak incidence is in the fifth decade.[4] The etiology and nature of the LYP are not clear, whether it is a benign or malignant condition. It was previously considered to be a pseudolymphomatous inflammatory process, because of its typical waxing and waning clinical course. However recently, it is regarded as a low-grade cutaneous T-cell lymphoma according to World Health Organization-European Organization for Research and Treatment of Cancer (WHO-EOTC) combined classification due to its association with other malignant LPDs. Although clinically it is a benign condition manifested by recurrent, self-healing lesions, it resembles malignant lymphoma histologically. It involves trunks and extremities commonly. The face, buttocks, and genitalia are less frequently involved. Delle *et al.* reported a case of a 44-year-old male with recurrent self-healing eruption limited to the right cheek. The diagnosis was kept as type A LYP based on histopathological examination. This was the first case report of LYP localized to face.[5] In our patient, the lesions were predominantly present over face which is quite a rare finding. Rarely, plaques and nodules can also be seen, as reported by Khondker *et al.* in a 48-year-old male who presented with the complaint of erythematous, painless, nonpruritic plaques, and subcutaneous nodules.[6] The clinical presentation of LYP in children did not differ greatly from

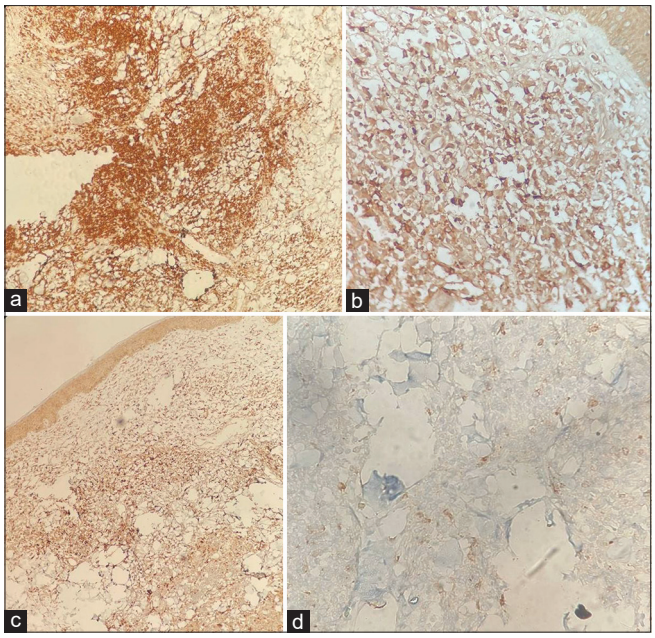

Figure 3: Immunohistochemical evaluation- [a; 10x]- CD 3 positive, [b; 40X] – CD 20 positive; [c; 10X] – CD 68 positive and [d; 40X] – CD 30 positive

the presentation in adults. Satya *et al*. reported a case of a 16-year-old boy with a 4-year history of asymptomatic, recurrent, papulonodular lesions on the arms, right axilla, trunk, groin, and legs, healing with post-inflammatory hyperpigmentation and atrophic scars. Histopathology and immunohistochemistry studies of these lesions are consistent with CD30+ LYP.[7]

Histopathologically, LYP is characterized by wedge-shaped inflammatory infiltrate extending to the deep dermis or superficial subcutaneous tissue and is further divided into subtypes based on morphological features and immunohistochemistry. LYP Type A is characterized by wedge-shaped, superficial infiltrate of CD 30+ atypical lymphoid cells that resemble Reed–Sternberg cells along with small lymphocytes, plasma cells, neutrophils, and eosinophils. No epidermotropism is seen in this subtype. Type B is characterized by perivascular or band-like dermal infiltration of small- to medium-sized lymphocytes with cerebriform nuclei which are CD 30+ or CD 30-, with epidermotropism, resembling mycosis fungoides. LYP type C shows the monotonous population of CD 30+ large atypical cells with fewer inflammatory cells, resembling anaplastic large cell lymphoma (ALCL). Type D is a variant that histologically simulates an aggressive epidermotropic CD8 + cytotoxic T-cell lymphoma. Type E LYP clinically and histologically resembles angiocentric and angiodestructive T-cell lymphoma.[8] The granulomatous and eccrinotropic LYP is a rare histological variant that was reported by Jain *et al.* in a 40-year-old male who presented with self-healing papulonodular lesions over the trunk and extremities.[9] The atypical cells of LYP resemble the Reed–Sternberg cells of Hodgkin's disease (HD) or the Sézary cells of cutaneous T-cell lymphoma and have the immunophenotype of activated T-cells and hence histologically resemble malignant lymphoma. Histopathological differential diagnosis of LYP includes cutaneous lymphomas, insect bite reaction, chronic dermatitis, and lymphomatous drug reactions.

A dense pan-dermal lympho-histiocytic infiltrate creeping in between the collagen bundles and adnexa with minimal focal lymphocytic epidermotropism was noted in our patient. There were CD3 and CD 20 positive cells along with the presence of a few CD 30 positive cells and numerous CD 68 positive histiocytes. Pai *et al.* reported a case of LYP in a child in which the lymphoid cells were predominantly CD 3 + with admixed CD 20 + positive cells, while the large atypical cells stained positive for CD3 and CD30 which was quite similar to our case; however, the lymphoid cells were angiocentrically distributed which was not present in our case.[8]

LYP is often diagnosed as a more common entity that presents as the recurrent papulonodular lesions [Table 1]. The overall prognosis is good, and the disease follows a benign course. The available treatment modalities include topical steroids, oral methotrexate, targeted phototherapy, photodynamic therapy, oral or topical retinoids, and anti-CD 30 monoclonal antibody-drug conjugate. They may hasten the healing of the lesions or prevent the eruptions of new lesions but one of them reduces the natural course of the disease or reduces the risk of developing an associated lymphoma.[10] Wait and see strategy is recommended in case of limited or asymptomatic disease. In terms of extensive, refractory disease, or disease involving cosmetically sensitive areas (e.g., face) as in our case, low-dose methotrexate between 10 and 25 mg weekly is recommended as the treatment of choice.[11]

Conclusion

LYP is a rare disease with a benign course and excellent prognosis. This case highlights the indolent course of this entity although the histology would suggest a more aggressive disease. Clinicopathological correlation is mandatory for the correct diagnosis of these diseases. Some cases may resolve spontaneously. However, some cases may have an aggressive course and transformation to malignant lymphoma can occur variably with increasing duration of the disease, and hence, long-term follow is warranted.

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

Table 1: Differential diagnosis of lymphomatoid papulosis

Differential diagnosis	Cutaneous findings	Histopathological features	Differentiating features
Pityriasis lichenoides et varioliformis acuta (PLEVA), Mucha-Habermann disease and pityriasis lichenoides chronica (PLC)	• Recurrent crops of spontaneously regressing erythematous papules • Lesions of PLEVA are crusted and occasionally vesiculopustular, while in PLC they are scaly	• Interface dermatitis with necrotic keratinocytes • Infiltrate is predominantly T-cell that is often monoclonal	• Often short-lived and more often in younger individuals as compared to LYP. • Does not develop nodular lesions and rarely, if ever progress to a lymphoma
Cutaneous T-cell lymphoma	• Polymorphic persistent patches and plaques in limb/girdle, breasts, and buttocks • Can progress to erythroderma with severe pruritis	• Large number of atypical mononuclear cells in dermis with epidermotropism • Sezary like cells	• Persistent polymorphic lesions and progression to erythroderma are quite common • Comparatively more epidermotropism, pautrier microabscess, atypical lymphocytes with large nuclei, and frequent abnormal mitosis • Commonly in elderly men and aggressive course
Pseudolymphoma	• Variable presentation ranging from solitary or multiple, smooth-surfaced or excoriated, asymptomatic or itchy, papules and nodules to exfoliative erythroderma • Presence of predisposing factors such as trauma, insect bite, infections, or drug intake.	• Mixed cellular infiltrate, including eosinophils and plasma cells • Minimal cellular atypia and polyclonality	• Benign course with spontaneous resolution on removal of predisposing factors. • Do not heal with scarring

Other differential diagnosis—Arthropod bite, folliculitis, miliaria, milia, scabies, and leukemia cutis–can be differentiated based on history and examination

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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