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Lymphomatoid Papulosis - A Rare Case Report

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Abstract: ***Lymphomatoid papulosis (LyP) ia disease of immune system which presents with recurrent self-healing papulo-necrotic or papulonodular eruption and a histological feature of CD30+lymphoid proliferation of atypical T cells. Thus, it is described as "self-healing rhythmical paradoxical papular eruption, histologically malignant but clinically benign" (1) LyP is characterized by a chronic course, lasting years or decades.***

Keywords: LyP- lymphomatoid papulosis

1. Introduction

Lyp is a rare form of primary cutaneous anaplastic large cell lymphoma. The family of primary cutaneous CD30+ lymphoproliferative disorders include diseases that are non-malignant and full-blown lymphomas. LyP is classified as non-malignant or as a cutaneous T cell lymphoma precursor.

Epidemiology- Incidence LyP is 1.2-1.9 cases per 1 million people. It can affect any age group with average age of onset 35-45 years. Men are more commonly affected than women. (2)

Etiopathogenesis-Nosingle factor has been proven to cause this disease. CD30 signaling is known to affect the growth and survival of lymphoid cell. Several authors suggested a viral etiology like Epstein bar virus, herpes virus but studies failed to prove this association. Thus, exact mechanism is yet unknown.

Clinical features- LyP presents as few to multiple red-brown papules and nodules distributed all over body predominantly over trunk and extremities. Characteristically, skin lesions in different stages of evolution coexist. Lesions are mostly asymptomatic but some patient may experience itch and pain associated with ulceration, crusting and necrosis. Ulcerated lesions heal spontaneously within 3-12 weeks to leave post-inflammatory hypo- or hyper-pigmentation and superficial varioliform scars while non-ulcerated lesions disappear without sequelae.

Complications- The prognosis of LyP is usually excellent but 10-15% of patients with LyP may be diagnosed with T cell lymphoma like mycosis fungoides, cutaneous anaplastic large cell lymphoma or even systemic lymphomas.

Differential diagnosis- Papular Urticaria, Scabies, Pityriasis Lichenoides, Prurigo, Folliculitis, Primary Cutaneous Anaplastic Large Cell Lymphoma.

Histopathological examination- multiple histological patterns have been recognized (type A-F), the most common being the type A classic form which shows extensive inflammatory infiltrate composed of CD30+ lymphocytes, histiocytes, neutrophils.

Treatment- Active treatment is not necessary in patients with relatively few non-scarring lesions. In case of cosmetically disturbing lesions systemic immunomodulatory like low dose methotrexate (5-20mg/week), interferon α, bexarotene can be given. Topical treatment like intralesional steroids, PUVA, regional radiotherapy.

An anti-CD 30 monoclonal antibody that is Brentuximab vedotin can selectively target the cells expressing CD30 antigen. Long-term follow up and surveillance is required in view of development of systemic lymphoma

2. Case Report

A 49year old male, presented with recurrent multiple raised dark colored lesions all over body since past 4 years. Lesions were associated with mild intermittent itching. Patient was a known case of Hodgkin's lymphoma for which he had taken 6 cycles of Adriamycin, Bleomycin sulfate, Vinblastin sulfate, Dacarbazine chemotherapy and radiotherapy and post-treatment Positron Emission Tomography scan was negative.

On Examination
Multiple papulonodular eruption with necrotic center, hemorrhagic crusting seen over few lesions. Lesions seen in different stages of evolution leaving hyperpigmented, atrophic oval scars predominantly on trunk and limbs.

On Dermoscopic examination- peripheral delicate dotted vascular pattern surrounding a central pinklight brown homogenous area with whitish scales at few sites is observed.

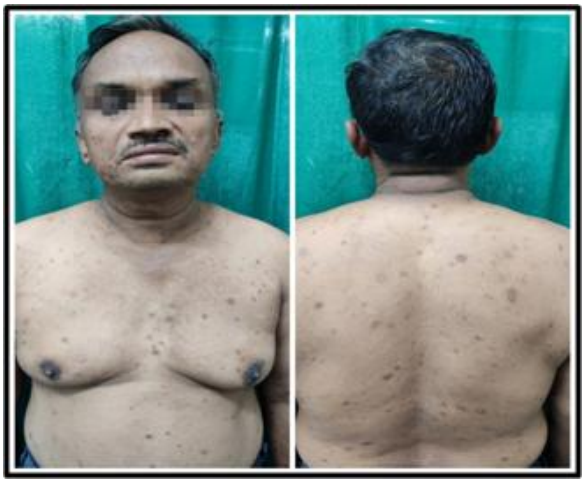

Figure 1: Papulonodular lesions over trunk

Figure 2: Papulonodular lesions over face

Skin Biopsy

Shows moderately dense superficial and deep perivascular and peri appendageal infiltrate of small and large lymphocytes (black arrow), few eosinophils (red arrow) with papillary dermal oedema. Overlying epidermis showed mild focal spongiosis and slight infiltration of small and large lymphocytes.

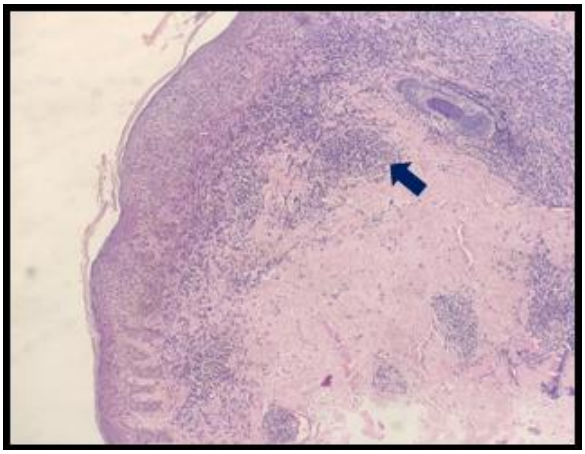

Figure 3: 10X view of skin biopsy

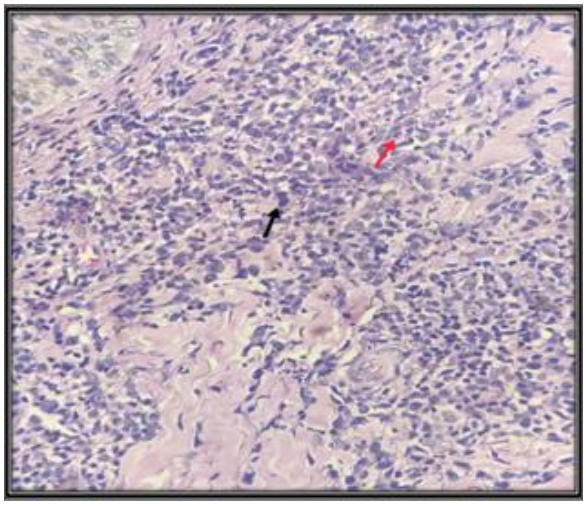

Figure 4: 40x view

Immunohistochemistry shows CD30+ lymphoid cells (arrow head). Which is in concordance with histopathological A subgroup of Lymphomatoid papulosis

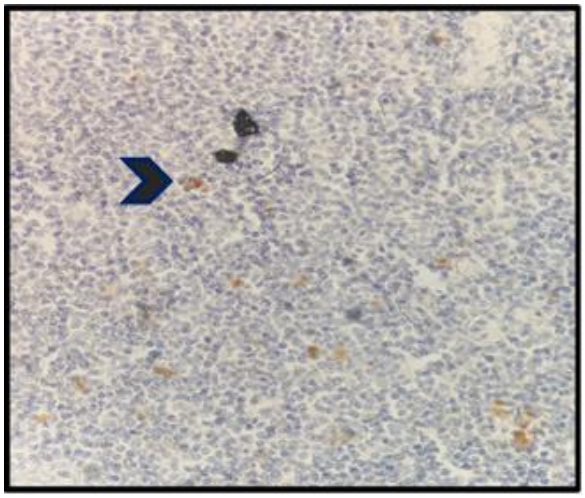

Figure 5: Immunohistochemistry – CD30+ cells seen

3. Conclusion

Despite malignant appearance on histology, LyP typically has an indolent relapsing and remitting course lasting for months to decades. The five-year survival is estimated to be 100%. (2)

Some cases may require long-term follow up and surveillance for systemic lymphoma development. TCR gene rearrangement is associated risk factor for it.

References

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