

Lymphomatoid Papulosis –3 Case Reports

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Abstract: We present two cases of childhood and one case of adult Lymphomatoid papulosis (LyP), an entity which is commonly misdiagnosed and poorly described in the paediatric dermatology literature. Clinically and histologically, the features of LyP in children can mimic insect bite reaction, with prominent dermal neutrophils and eosinophils. However, CD30 immunohistochemical staining of atypical lymphocytes within a mixed inflammatory infiltrate should point to the diagnosis of LyP. There is no consensus to guide management of childhood LyP due to its rarity and largely unknown natural course. We discuss our experience with LyP in two children who are blood related and one adult case. Although none of our cases have experienced malignant transformation to date and life long monitoring is advocated.

Keywords: CD30+, Cutaneous lymphomas, Childhood LyP, Lymphomatoid papulosis, Lymphoproliferative disorder.

I. Introduction

Childhood lymphomatoid papulosis (LyP) is an entity within the spectrum of CD 30+ cutaneous lymphoproliferative disorders that behaves in a clinically benign fashion but histologically mimics a malignant cutaneous lymphoma. The features of LyP in children are similar to those of adult form, which is familiar to dermatologists and for which exists well established management guidelines. Childhood LyP is rare, with approximately 100 cases reported in the literature and a general lack of consensus regarding its clinical course, risk of malignant transformation and recommendations for management and follow up.

Similar to adult onset LyP, the papulonodular lesions of childhood LyP may ulcerate and become necrotic, they typically heal slowly over 3 to 12 weeks. The rhythmic eruption of these lesions may recur for months or years before spontaneous and persistent resolution. Frequently atrophic or varioliform scars are left in testimony to the preceding papulonodular lesions. For this reason treatment for childhood LyP has been advocated predominantly to minimize scarring¹⁻³.

In this article we are sharing our experiences with three cases of LyP.



Fig: 1. Two siblings showing papular lesions

II. Case Reports

2. 1.Case.1: A 4 yr old male child was brought by his mother with complaints of rash over face and trunk since 6months of his age (3 ½ yrs). Recurrent episodes of asymptomatic erythematous raised lesions over the face and trunk at monthly intervals which are healing with hyperpigmentation. Not associated with fever or any other systemic symptoms. H/o consanguinity (1st degree) present in the parents. No h/o of drug intake prior to onset of rash. Similar type of history is present in mother's brother. On dermatological examination multiple erythematous papules & nodules are present over face & trunk which are healing with hyperpigmentation. Few lesions are crusted. Few lesions are not blanchable. The general examination was normal except for dermatological findings.



Fig. 2&3: Lesions on the face.



Fig. 4&5: Shows popular lesions on the body

FNAC of lymph nodes shows- Non specific lymphadenitis.

HPE: Sections shows skin with mild orthokeratosis. Epidermis is unremarkable with preserved basal layer. The dermis shows moderate interstitial perivascular and periappendegeal atypical cells, lymphocyte, plasma cell infiltrate extending transdermally. Admixed atypical cells shows irregular nuclear margins and cleaved nuclei.

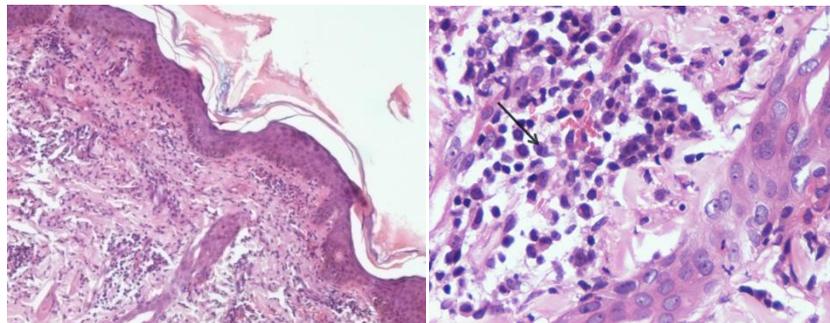


Fig. 6&7: Shows reactive lymphoid infiltrate in dermis

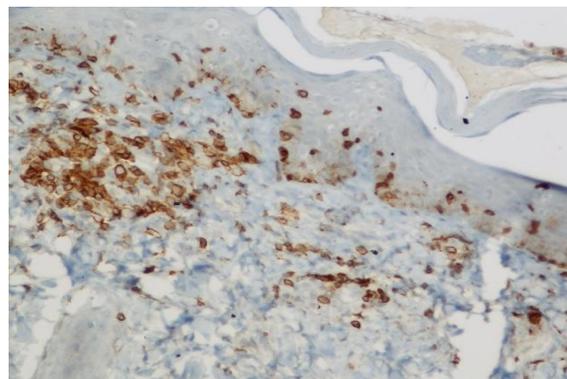


Fig. 7: IHC, CD30: Scattered cells are positive in dermis

2.2. Case.2: A 12 yr male child brought with similar complaints as case 1 since 6 months of his age and he is the brother of case 1. On dermatological examination, multiple erythematous papules and nodules present predominantly over trunk and few lesions over the face. All are healing with hyperpigmentation. The general and physical examination was normal, except for dermatological findings.



Fig: 8,9,10. Macules and Papular lesions on the trunk

FNAC of lymph nodes : Shows non specific lymphadenitis.

HPE: Section from skin shows mild orthokeratosis, mild lymphocyte exocytosis. There is upper dermal moderate lymphocyte infiltrate with few scattered large cells. Section from wedge biopsy shows mild orthokeratosis, lymphocyte exocytosis and preserved basal layer. There is prominent upper dermal, perivascular and periadnexal infiltrate of lymphocytes, admixed with few atypical large cells. These atypical cells have irregular nucleus with conspicuous nucleoli. There is haemorrhage in deeper layers.

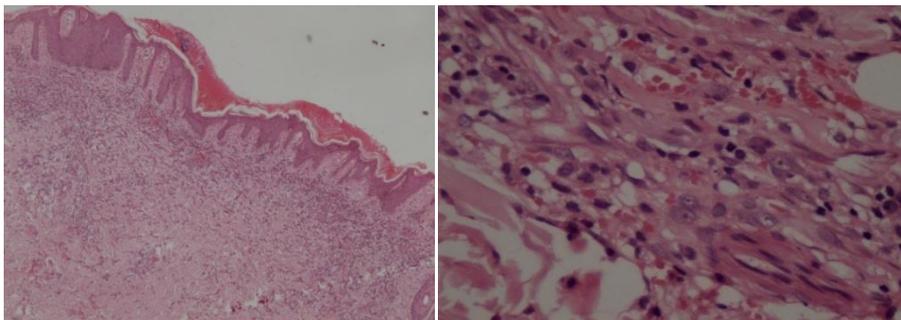


Fig: 11. Shows atypical lymphoid infiltrate

Fig:12. Shows IHC: Scattered CD 30 cells + in dermis

2.3.Case.3: A 33 year old female presented with asymptomatic rash on the body of 15 yrs duration. The lesions started 15 years back with recurrences on and off. Erythematous grouped papular eruption started on the trunk, face, neck and extending to limbs, abdomen and back. Lesions progressed rapidly over few days to pustules followed by ulceration and crust formation and few healing with hyperpigmented scars. No history of fever, insect bite allergies or drug intake prior to the eruption. No history suggestive of exposure to chemicals & metals. No abdominal pain. She has taken treatment from a private hospital for a period of 5 years with no improvement. The left central group of axillary lymph nodes, and left inguinal lymph nodes are enlarged which are firm and non tender.

2.4.Cutaneous examination: Multiple papulo nodular & ulcerated necrotic lesions over the back, face, neck, abdomen, lower limbs and genitalia. Few had erythematous base and few resolved with varioliform scars. Routine investigations are normal except elevated ESR. Ultra sonography of abdomen showed normal study.



Fig.13. Shows Papulo pustular lesions with hyperpigmentation on the the body

FNAC of lymph nodes: shows non specific lymphadenitis.

HPE: Epidermis shows mild hyperkeratosis, Irregular acanthosis with exocytosis of lymphocytes. Dermis shows nodular infiltrate of lymphocytes with irregular nuclear margins with few large lymphocytes with nucleolus.

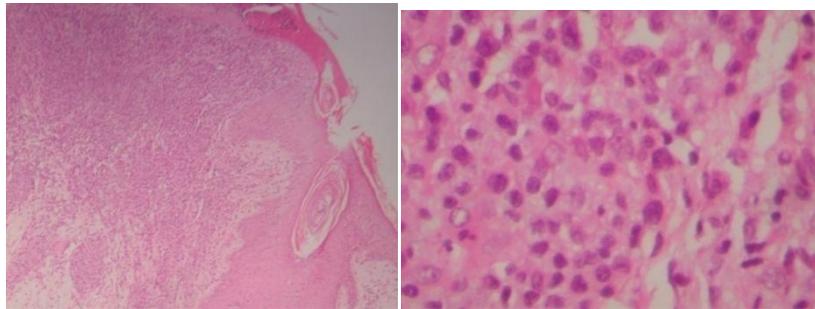


Fig.14: IHC : CD 3 and CD 30 Positive in dermis

Table.1: Shows Clinical and Histopathological findings of LyP

case	Age of patient	Age of onset Of disease	Clinical Features	Initial diagnosis	Histopatological Examination	Treatment
1.	4years	6 months	Papules and Nodules on face and trunk	LyP	Moderate dermal infiltrate with atypical cells. Atypical cells staining positive for CD 30.	Fluticasone Propionate 0.05% ,natural sunlight
2.	12 years	6 months	Papules and nodules predominant over trunk	LyP	Dense dermal infiltrate with atypical cells. Atypical cells staining positive for CD 30.	Fluticasone propionate 0.05% ,natural sunlight
3.	33 years	15 years	Papules and nodules, ulcerated and crusted lesions over trunk, neck & abdomen	LyP	Nodular infiltrate with atypical cells staining positive for CD 30	Oral Methotrexate Acitretin

All of the three cases are under follow up.

III. Discussion

The decision to actively treat childhood LyP is controversial since this disease is self resolving. However, the rhythmic eruption of lesions may lead to unsightly scarring. Treatment for childhood LyP has been advocated only if symptomatic or if cosmetically significant scarring is a concern.¹⁻³ Little evidence and few recommendation exist for the treatment of childhood LyP. Multiple treatment modalities including topical, systemic and intra-lesional corticosteroids,³⁻⁶ systemic antibiotics³⁻⁶ and natural sunlight,⁴ psoralen and UVA therapy,^{7,8} and narrow band UVB therapy^{4,7} have been described but with varying success. None of these modify the course of disease or lead to permanent remission, but merely produce partial and transient responses. Nijsten et al. surveyed a registry of childhood LyP consisting of 35 children reported that phototherapy was superior to topical corticosteroids and systemic antibiotics.¹ The same author also reported that low dose Methotrexate was effective in two children and may be associated with less active disease.

The risk of malignant transformation of adult onset LyP is 10-20%, which may precede, coexist with or follow malignant cutaneous lymphomas.⁸ The risk of childhood LyP transforming in to cutaneous lymphoma is less known but has been estimated to be approximately 10%.¹ Although none of our cases have developed malignant transformation till date. Long term follow-up is required to closely monitor for this risk. No guidelines exist to recommend long term follow up at 6 to 12 monthly intervals with additional investigations performed only when abnormalities were detected at routine evaluations.

IV. Conclusion

Childhood LyP is commonly misdiagnosed due to its rarity and clinico- histological similarities to insect bite reactions.¹ Pathologists should be alerted to the clinical suspicion of LyP in children with multiple nodules and perform CD30 immunostaining if appropriate. Treatments for childhood LyP have had varying success, but Methotrexate is a safe and effective alternative. Life-long monitoring for potential malignant transformation of childhood LyP is recommended.

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