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CASE STUDY

PRIMARY CUTANEOUS CD30-POSITIVE LARGE T-CELL LYMPHOMA- A SPECTRUM OF DISEASES

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ABSTRACT

Primary cutaneous CD30- positive large cell lymphoma (CD30+ PCLCL) represents a spectrum of disease, with lymphomatoid papulosis (LyP) at the benign end, and primary cutaneous anaplastic large cell lymphoma (PCALCL) at the other. We present two cases of CD30+PCLCL. First case presented with papules and nodules with severe itching. Biopsy of the skin showed atypical multinucleate RS like cells. Second patient had a history of ulcerated plaques and papules over scalp, back and thigh which showed many atypical cells. Atypical cells in both cases stained positive for CD30, CD3 and CD4 and negative for ALK, EMA. After a thorough clinicopathological correlation a diagnosis of CD30+PCLCL – lymphomatoid papulosis was made in first case and CD30+ PCLTCL – Anaplastic type in second. CD30+ PCLCL are characterized by an excellent prognosis. Therefore, it is essential to consider other differentials before making a final diagnosis.

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INTRODUCTION

Primary cutaneous CD30-positive large cell lymphoma (CD30+ PCLCL) is the second most common type of cutaneous T-cell lymphoma, accounting for approximately 30% of all cases (Querfeld, 2009). In cutaneous lymphomas CD30 expression is a more important prognostic parameter than is the extent of skin disease at presentation. Beljaards RC *et al.,* 1993 in his study stated that only 7% of patients with CD30+PCLCL died of progressive disease compared to 80% of patients with CD30 negative PCLCL. It comprises of Primary cutaneous anaplastic large cell lymphoma (PCALCL) and lymphomatoid papulosis (LyP). These conditions represent a spectrum of disease with LyP at the benign end and PCALCL at the malignant (Paulli *et al*., 1995). When PCLCL is suspected, it is important to consider other differential diagnosis of CD 30+ PCLCL in order to reduce the use of unnecessarily aggressive chemotherapy regimens. We present two cases of CD 30+ lymphoproliferations, one at each end of the spectrum with brief discussion on important differentials to be considered.

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Case History

First case was a 35 year female who presented with generalized itching since one year. Numerous nodules and papules were seen on forearm, back and thigh ranging in size from 0.2 to 1cm. Second case was a 68 year male with 8 month history of itchy small nodules and ulcers on scalp, back and leg. There were polymorphic lesions seen in form of umblicated papules and ulcerated plaques which ranged in diameter from 1.5 cm to 2.2 cm (Figure 1). In both the cases no lymphadenopathy was noted. Hematological, biochemical, radiological investigations and bone marrow biopsy were normal. Cutaneous biopsy from the thigh lesions in first case showed epidermis showing parakeratosis and irregular acanthosis. Superficial and mid dermis revealed a predominantly perivascular mixed inflammatory infiltrate comprised of lymphocytes, histiocytes, eosinophils, neutrophils and many atypical multinucleate R.S. like cells (Figure 2). Biopsy taken from the plaques in the second case revealed focal ulceration of epidermis with neutrophilic infiltration and acanthosis. Superficial and deep dermis revealed perivascular and interstitial infiltrate composed of lymphocytes, plasma cells and neutrophils with many atypical cells showing condensed nuclei with focal binucleation and multinucleation (Figure 3). Immunohistochemistry in both

cases showed atypical cells positive for CD3, CD4, CD30 and negative for CD20, ALK, EMA. CD8 was focally positive in first case while it was negative in the second case (Figure 4 8). Based on these clinical, histopathological and immunohistochemical findings a diagnosis of CD30+PCLCL-lymphomatoid papulosis was made in the first case and a diagnosis of CD30+PCLCL – anaplastic type in second case.

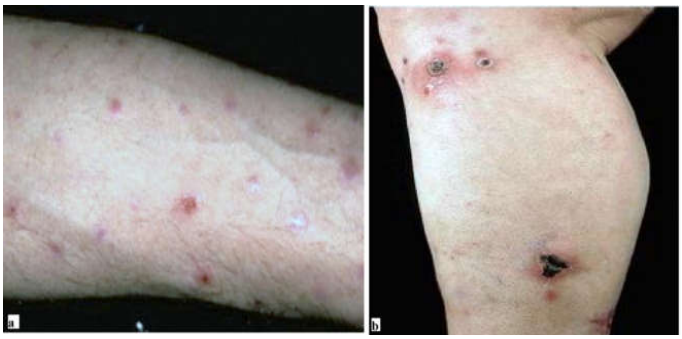

Figure 1(a). First case- Reddish papules and nodules on thigh (b) Second case - Ulcerated lessions on leg

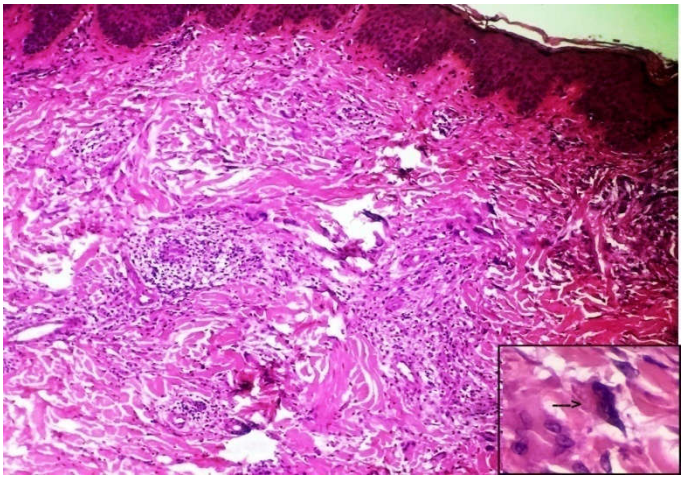

Figure 2. First case - upper and mid dermal perivascular inflammatory infiltrate (H and Ex10). Inset- Atypical multinucleate R.S. like cells (H and E, x40)

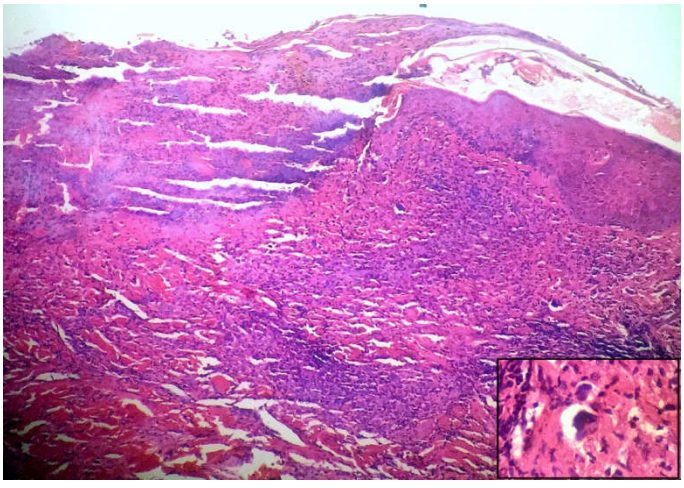

Figure 3. Second case- focal ulceration of epidermis with neutrophilic infiltration and acanthosis along with superficial and deep dermis showing perivascular and interstitial infiltrate composed of lymphocytes, plasma cells and neutrophils (40 X; H& E). Inset - atypical cells with focal binucleation (400 X; H&E)

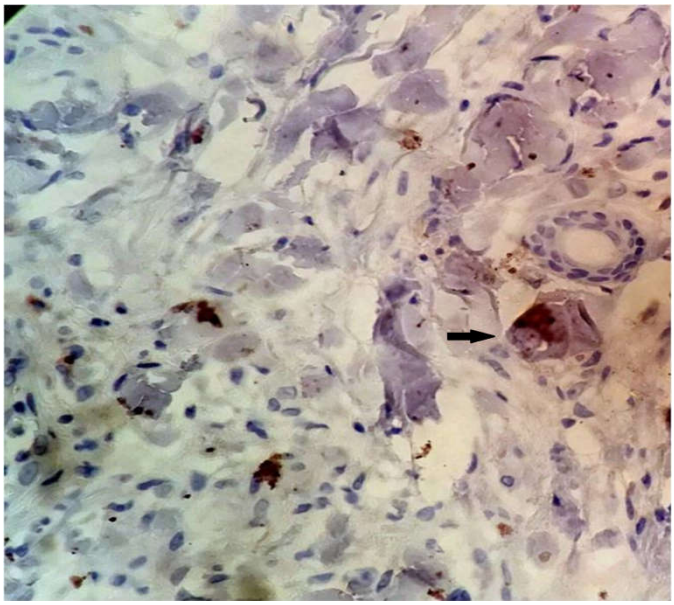

Figure 4. CD 30 – positive in case 1

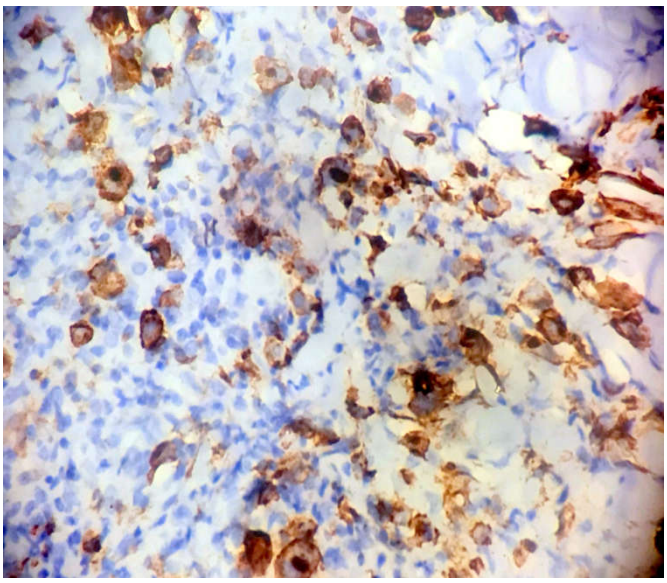

Figure 5. CD 30 – positive in case 2

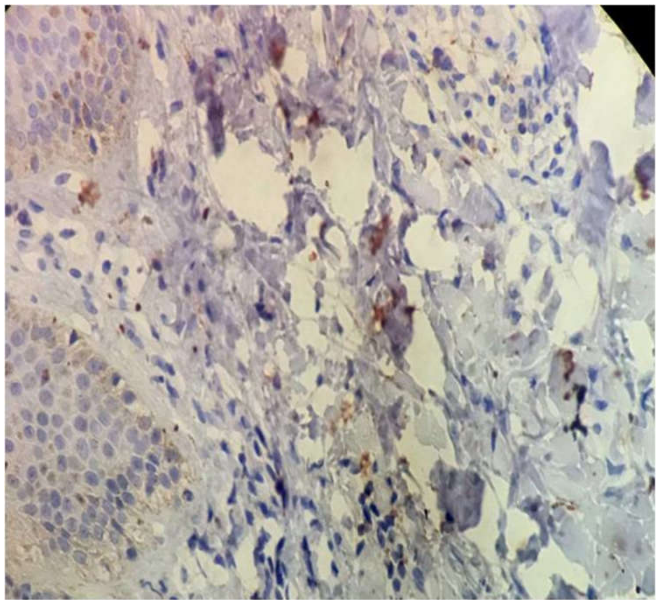

Figure 6. ALK – negative in case 1

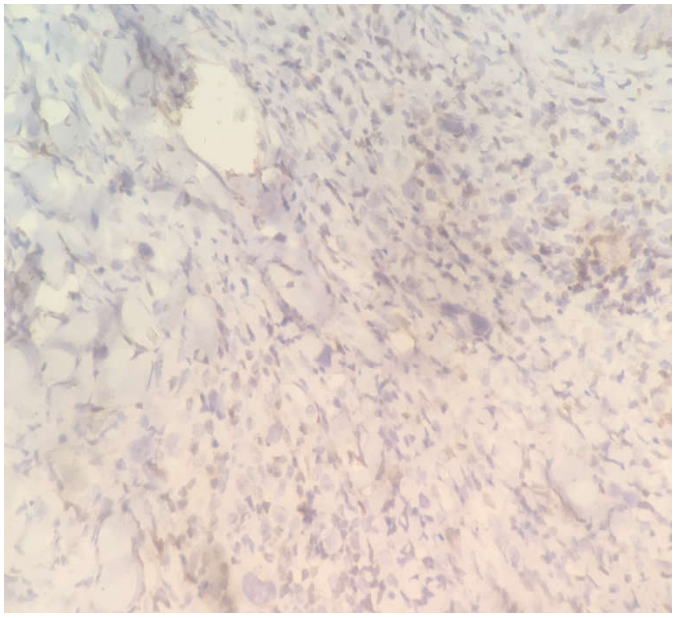

Figure 7. EMA - negative in case 2

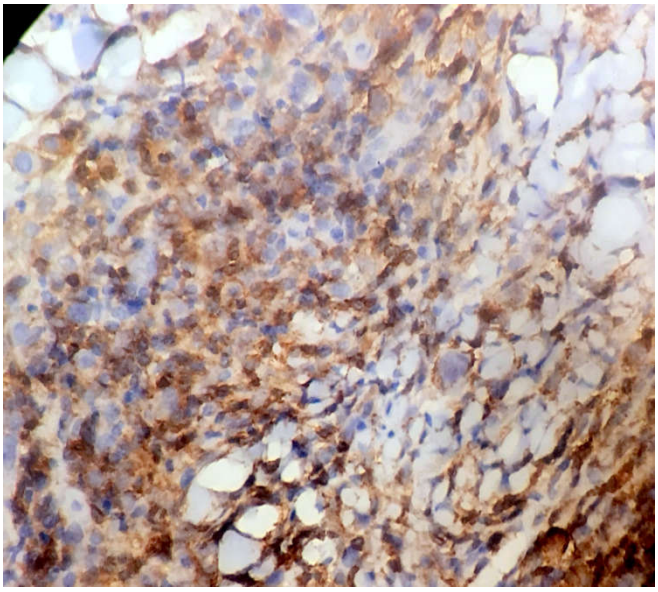

Figure 8. CD 4 Positive in case 2

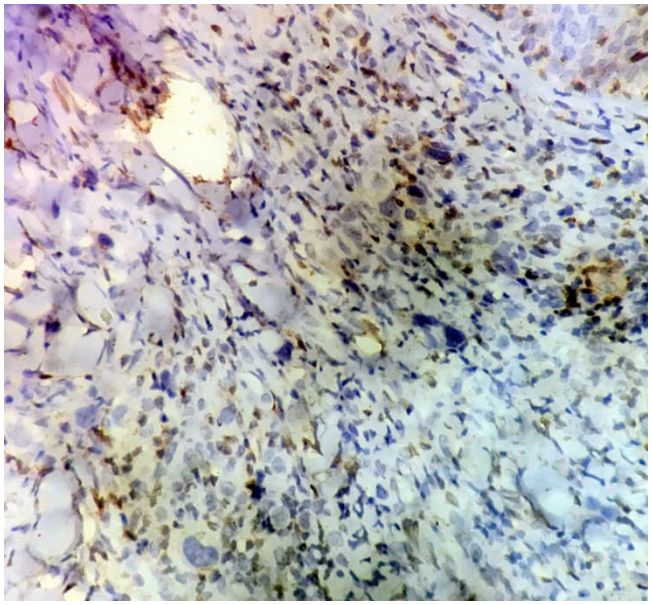

Figure 9. CD 20 negative in case 2

DISCUSSION

CD30+PCLCL is a rare subtype of cutaneous T-cell lymphoma. It represent a spectrum of disease, with LyP at the benign end, and PCALCL at the other. Although clinically typical cases of LyP can be easily distinguished from classical cases of PCALCL, a huge area of overlap exists and these entities are better understood in continuum. Both LyP and PCALCL lesions show histology of high grade lymphoma. However, while LyP patients have clinically benign self-healing lesions (Macaulay WL. 1968), in PCALCL only 25% of lesions regress spontaneously (Kempf, 2006). Also, patients with LyP have a 10–20% increased risk for developing lymphoid malignancy in comparison to PCALCL (Nijsten T *et al*., 2004). Thus, it is important to distinguish among them as the therapeutic strategy also differs. Individual lesions of LyP are <2 cm in diameter, whereas those of PC-ALCL are generally >2 cm. The distinction between the two entities is related to the depth of invasion of the infiltrate as well as to the clinical features and evolution of the disease in the patient. Several newer markers like Bcl-2, fascin, CLA, and CD56 and clonality by T-cell receptor (TCR) gene rearrangement are under study to distinguish between the two (Droc *et al*., 2007).

CD30+PCLCL are characterized by an excellent prognosis with a disease-specific survival at 5 years for LyP patients being 100% and for PCALCL patient around 90% (Bekkenk MW, *et al*., 2000). Therefore, the pathologist must be aware of the other differential diagnosis of CD 30+ PCLCL. An important differential to be considered is nodal/systemic ALCL manifesting as cutaneous lesion. This distinction is difficult by clinical examination or routine histopathology. Immunohistochemistry plays an important role in this. ALK protein and Epithelial membrane antigen (EMA) is expressed in tumor cells of most patients with nodal/systemic ALCL but not in the patients with CD30+PCLCL while CLA being more frequently expressed in CD30+PCLCL (de Bruin PC, *et al*., 1993). Other cutaneous infiltrates characterized by CD30 expression should also be considered in the differentials. These conditions include reactive and neoplastic diseases, such as arthropod bites,scabies, pityriasis lichenoides, Langerhans cell histiocytosis, cutaneous B-cell lymphomas with immunoblastic or large-cell features and CD30+ large-cell transformation of mycosis fungoides (Werner B, *et al*., 2008). Exclusion of these diseases is essential before establishing a diagnosis of PC-ALCL.

Conclusion

CD30+PCLCL may present in a variety of ways. For LyP no treatment is advised. However, methotrexate and low dose PUVA/ UVB therapy may be used. In single or localized PCALCL lesion either local excision or radiation is used. Chemotherapy is reserved for patients with systemic involvement.

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