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Case Report

Angiocentric lymphomatoid papulosis in a child: uncommon benign clinical entity with malignant histology

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ABSTRACT

Lymphomatoid papulosis (LyP) is a rare form of chronic inflammatory skin disease with histologic features of a malignant lymphoma.It presents clinically with history of recurrent crops of pruritic papules that occur on the trunk and limbs that resolve spontaneously. We report an unusual case of angiocentric LyP in a 4 year old child and review the literature.

Key words: Lymphomatoid papulosis; child; cutaneous lymphoma

INTRODUCTION

The term lymphomatoid papulosis originally was used by Macaulay in 1968 to describe "a self-healing rhythmical paradoxical eruption, histologically malignant but clinically benign" [1]. Awareness of this entity is important as the histological features may result in a mistaken diagnosis of malignant lymphoma, if clinical picture is not taken into account. Follow-up is essential as it can progress to malignant lymphoma in a subset of patients.

CASE REPORT

A 4 year old male child presented with itchy erythematous and skin colored papules over trunk, upper and lower limbs of 3 months duration (Fig. 1). There were few excoriated papules. The child did not have genital or web space lesions. There was no history of prior viral infection, drug intake etc. A clinical diagnosis of prurigo simplex was considered and a skin biopsy was taken. Histopathological examination revealed wedge shaped superficial and deep aggregates of atypical lymphoid cells with hyperchromatic nuclei and scanty cytoplasm, along with few mononuclear Reed Sternberg like cells with prominent nucleoli, admixed with plasma cells and histiocytes. Folliculotropism of the lymphoid cells was noted (Figs 2 and 3). Angiocentric infiltration around dermal blood vessels with necrosis of blood vessel wall was seen (Fig. 4). The lymphoid cells were predominantly CD 3 positive with admixed CD 20 positive cells, while the large atypical cells stained positive for CD 3 and CD30 (Fig. 5). A diagnosis of Lymphomatoid papulosis was made based on the clinical and histopathological features.

The patient's informed consent was obtained. Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

DISCUSSION

Lymphomatoid papulosis (LyP) is a chronic inflammatory skin disease which can histologically be mistaken for malignant lymphoma. It presents clinically with recurrent crops of itchy papules that occur on the trunk and legs that heal spontaneously over 1-2 months leaving slightly depressed oval scars.

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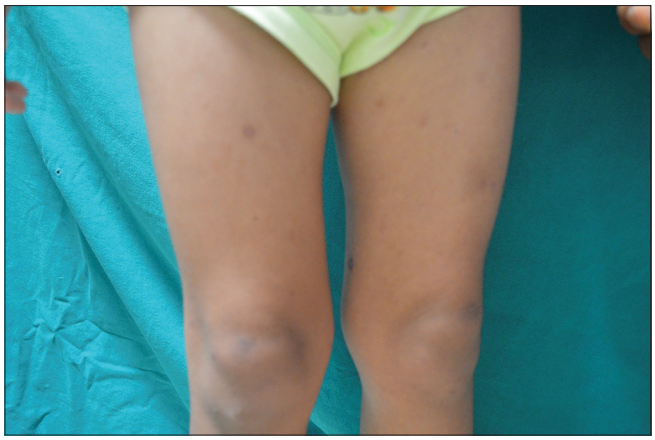

Figure 1: Clinical picture showing multiple resolving erythematous papules on the legs

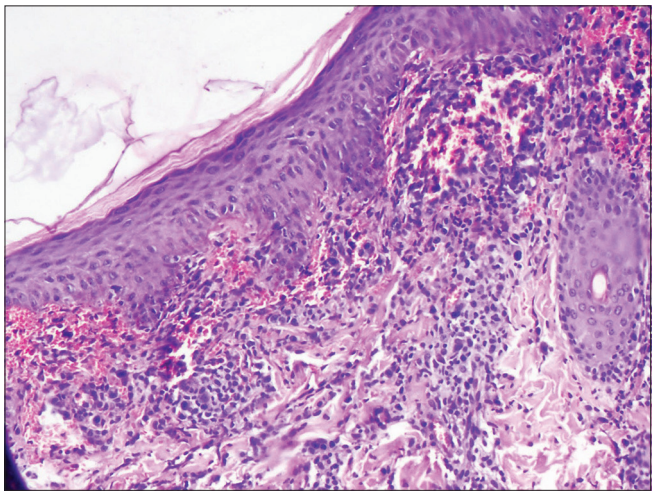

Figure 2: Wedge shaped superficial and deep diffuse and perivascular inflammatory infiltrate. H&E 200X

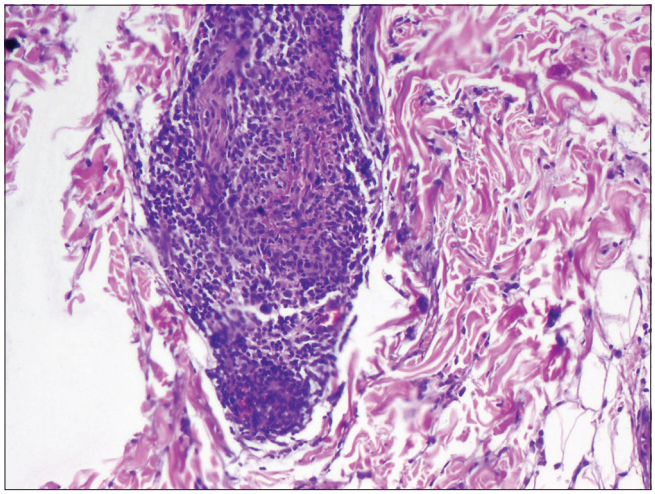

Figure 3: Angiocentric infiltration by lymphoid cells with fibrinoid necrosis of vessel wall. H&E, 200X

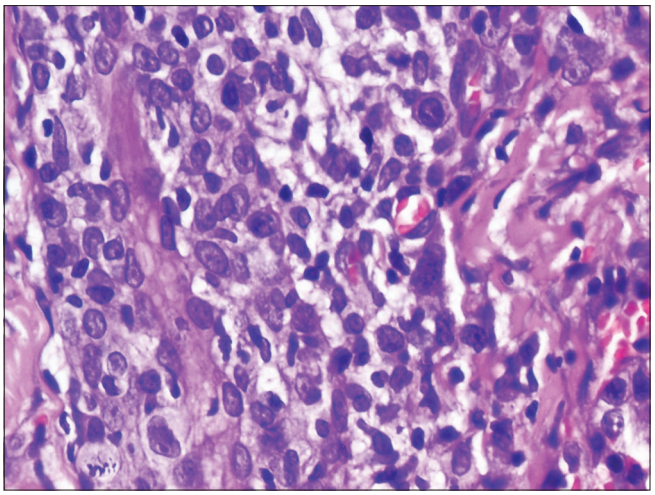

Figure 4: High power pictute showing large atypical lymphoid cells, and occasional mitosis. H&E, 400X

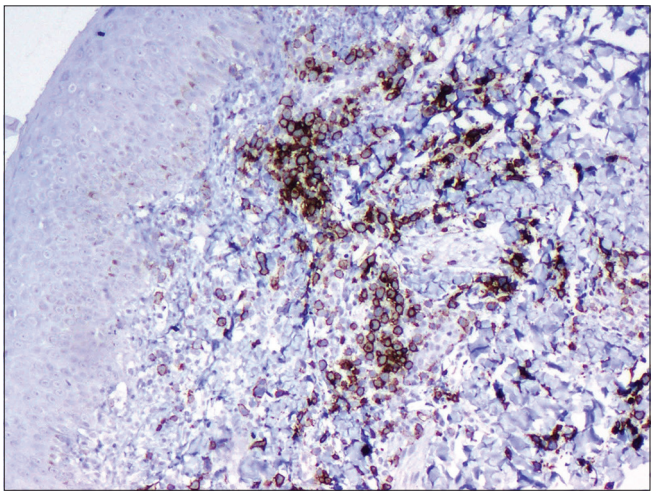

Figure 5: CD30 positivity among large atypical lymphoid cells. IHC, 200X

LyP was previously considered to be a pseudolymphomatous inflammatory process, because of the typical waxing and waning clinical course. However, World Health Organization—European Organization for Research and Treatment of Cancer (WHO-EORTC) classification, during the consensus meetings in 2003-2004 grouped lymphomatoid papulosis among the indolent cutaneous T-cell lymphomas [2]. The rationale behind classifying lymphomatoid papulosis as a cutaneous lymphoma is because of its association with other malignant lymphoproliferative disorders. Some experts consider this entity as chronic skin disease rather than a true malignancy because of its spontaneous resolution and benign clinical course.

Lymphomatoid papulosis belongs to the spectrum of CD30 (Ki-1)–positive cutaneous lymphoproliferative diseases ($CD30^+$ LPDs).This group includes lymphomatoid papulosis, primary cutaneous anaplastic

large cell lymphoma (pcALCL), and borderline CD30+ lesions [3].

The prevalence of lymphomatoid papulosis is estimated to be 1.2-1.9 cases per million population in the United States. It can develop at any age, but the peak incidence is in the fifth decade. LyP may rarely occur in children [4]. About 10-20% of LyP is thought to be associated with malignant lymphoma (ALCL, Hodgkin's disease, or Mycosis Fungoides) prior to, concurrent with, or subsequent to the diagnosis of lymphomatoid papulosis [5,6]. The etiology of lymphomatoid papulosis is not clear. There is debate about the nature of LyP,whether it is a benign or malignant condition. Histologically, it has features resembling malignant lymphoma; however, its clinical course manifested by recurrent, self-healing lesions suggests a benign nature.

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 morphologic features and immunohistochemistry. LyP Type A shows wedge-shaped, superficial and deep infiltrate of CD 30+atypical lymphoid cells, that resemble Reed Sternberg's cells along with small lymphocytes, often, plasma cells, neutrophils, and eosinophils. No epidermotropism is seen is seen in this subtype. LyP Type B is characterized by perivascular or band like dermal infiltrate of small to medium sized lymphocytes with cerebriform nuclei which are CD30+ or CD 30-, with epidermotropism, resembling Mycosis fungoides. LyP Type C shows monotonous population of CD 30+ large atypical cells with fewer inflammatory cells, and resembles ALCL. LyP Type D is a variant that histologically simulates an aggressive epidermotropic CD8 positive T-cell lymphoma. This cytotoxic variant of LyP may be histopathologically indistinguishable from primary cutaneous aggressive epidermotropic CD8+ cytotoxic T-cell lymphoma, and may be the source of pitfalls in the diagnosis and classification. A type E variant with clinical and histologic manifestations simulating highly aggressive angiocentric and angiodestructive T cell lymphoma has been described [7,8]. Patients present with a few papulonodular lesions that rapidly evolve to large ulcerations covered by a hemorrhagic and necrotic crust.However, our patient showed lymphomatoid papulosis, with angiocentric lymphoid infiltrate, but presented clinically with papular lesions, that regressed in 4 weeks.

Several inflammatory and reactive disorders like viral infection, drug eruption, arthropod bite reaction may contain a significant number of CD30+ cells and mimic lymphomatoid papulosis clinically or histologically [3]. Clinicopathologic correlation is hence necessary to establish an accurate diagnosis and to differentiate LyP from ALCL, Mycosis fungoides and Hodgkin's lymphoma and other inflammatory mimics.

Various therapeutic options are available to treat LyP, with methotrexate being the treatment of choice. Others include PUVA, potent topical steroids, interferon and topical mechlorethamine.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles.

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