Papular eruption characteristic of persistent agmination of lymphomatoid papulosis

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Received: 12 November 2019 / Accepted: 13 January 2020 / Published online: 20 January 2020
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Abstract

A 48-year-old man presented with a 1-year history of a nontender, pruritic, well-circumscribed crop of waxing and waning papules. The histological findings were compatible with lymphomatoid papulosis (LyP) in spite of the fact that cutaneous distribution was not typical. Attending to the clinicopathological correlation, we established the diagnosis of persistent agmination of lymphomatoid papulosis (PALP). PALP is a rare variant of CD30+ primary cutaneous T cell lymphoproliferative disorder, with only a few reported cases in the literature. It is still controversial whether PALP is a subtype of localized LyP or an own entity. In any case, there is potential of developing malignancy, so at least a cautious long-term follow-up is recommended in these patients.

Keywords Lymphomatoid papulosis · Persistent agmination · CD30+ cutaneous T cell lymphoma · Localized lymphomatoid papulosis

Introduction

Lymphomatoid papulosis (LyP) is classically defined as a chronic, recurrent, and self-healing skin disease featuring papular eruption in a generalized distribution. However, in a small subset of patients, the eruption is limited to a single anatomic region making the diagnosis more difficult for clinicians.

Clinical history

A 48-year-old man with no past medical history presented with a 1-year history of a nontender, pruritic, well-circumscribed crop of waxing and waning papules on the right side of the abdomen that never resolved completely.

Physical examination showed multiple erythematous papules within an orangey plaque on the right side of the abdomen

(Fig. 1). No lymphadenopathy in any lymph node chains was found.

Material and methods

A punch biopsy from one of the papules showed a polymorphic superficial and deep dermal infiltrate with perivascular and interstitial disposition. The infiltrate was composed of lymphocytes, histiocytes, and eosinophils (Fig. 2). Some of the lymphocytes were large-sized

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Fig. 1 Multiple erythematous papules within an orangey plaque on the right side of the abdomen

Fig. 2 Haematoxylin and eosin stain showing a mixed-cell dermal infiltrate with some large-sized lymphocytes with cerebriform nuclei

with cerebriform nuclei. Epidermotropism was not present. Immunohistochemical stains for CD3, CD4, CD8, CD43, CD30, CD20, and ALK-1 showed a CD3+, CD4+, CD8–, CD43+, CD20–, ALK-1– pattern, and CD30+ in more than 20% of the neoplastic cells (Figs. 3 and 4). Monoclonal rearrangement of the γ T cell receptor was demonstrated on polymerase chain reaction. Routine blood extension analyses were normal. Based on the clinical presentation and histological findings, we made the diagnosis of persistent agmination of lymphomatoid papulosis (PALP). Treatment with mometasone cream was initiated with partial response after 3-month follow-

Fig. 3 CD3 immunostaining

Fig. 4 CD30 immunostaining

up, so we initiated clobetasol cream with complete response after 1 month. No recurrences at the 6-month follow-up were seen. The patient is still on regular follow-up every 3 months.

Discussion

PALP is a rare variant of CD30+ primary cutaneous T cell lymphoproliferative disorder, with only a few reported cases in the literature. LyP is classically defined as a chronic, recurrent, and self-healing skin disease characterized by papular, papulonecrotic, or nodular lesions that typically occur in a generalized distribution, with multiple lesions over the trunk and extremities. However, in a small subset of patients, the eruption is limited to a single anatomic region defining the localized or regional lymphomatoid papulosis. The concept of PALP has been frequently challenged. Some authors suggest that it is a form of localized LyP and others that it represents an own entity.

Classically, PALP is a recurrent disease with papulonecrotic lesions circumscribed within a mycosis fungoides-like patch area, usually on the trunk and thighs [1].

Scarlsbrick et al. were the first to group 4 cases of LyP limited to specific regions [2]. Interestingly, one of the patients had mycosis fungoides (MF), and the LyP lesions were confined to the same region where MF lesions were observed. Steinhoff et al. highlighted the different clinical manifestations of localized LyP, ranging from scattered solitary circumscribable papules in between normal skin to densely packed and confluent papules forming an “MF-like” plaque lesion that in contrast to MF is not sharply demarcated [3]. Localized LyP has been described in

Table 1 Differential diagnosis of PALP

	PALP	Localized LyP	Papular MF	Transformed MF	Lymphomatoid PL
Clinical history	Persistent eruption with waxing and waning papules	Self-healing papular eruption	Persistent papular eruption. Not waxing and waning pattern	Previous history of patches and plaques of MF	Papules or nodules in a generalized distribution
Physical examination	Crops of papules that erupt and crust but within a well-circumscribed area typical for MF	Crops of papules that erupt, crust, and resolve. Not always on areas typical for MF. Usually with a pleomorphic pattern	Small erythematous, monomorphous, partly scaly papules in the absence of patches or plaques. Not well-circumscribed	Frequently nodules arising in plaques or de novo	Papules that develop a micaceous scale and then regress. Polymorphous appearance leaving macular pigmentary alterations
Histology	Consistent with LyP, most commonly with type A. Usually without fibrosis	Polymorphous infiltrate with atypical lymphocytes. Six histologic types*	Lymphoid infiltrate within a fibrotic papillary dermis with some intraepithelial lymphocytes	Dense infiltrate with large atypical lymphocytes often admixed with epidermotropic small cells of MF	Vary according to morphology of the lesion. PLC is more likely to have histopathologic features that overlap with LyP-B or papular MF
IHC	CD30+	CD30+	Classical MF phenotype (CD3+ CD4+ CD8–) with CD30–	Classical MF phenotype with CD30+ in 50% cases	CD30–

PALP, persistent agmination of lymphomatoid papulosis; *LyP*, lymphomatoid papulosis; *MF*, mycosis fungoides; *PL*, pityriasis lichenoides; *IHC*, immunohistochemical stains; *PLC*, pityriasis lichenoides chronica

*Histologic distinction between papular MF and type B of LyP may be impossible without accurate correlation with the clinical features. Although CD30 is negative in cases of papular MF, rare, questionable cases of LyP type B with lack of CD30 expression have been described

anatomic regions affected by other conditions like radiotherapy, red tattoo, and Becker melanosis [4–6]. Nevertheless, the pathogenesis for regional distribution of LyP remains uncertain. In 2007, Heald et al. first described PALP showing that the behavior of this entity is more like MF than the self-healing eruption of LyP. More recently, Karai et al. have found a type of LyP with chromosomal rearrangements on 6p25.3 that usually affects a single body area [7]. Unlike PALP, localized LyP may remit spontaneously [8]; however, some treatments have been used with variable results. These include topical and intralesional steroids, injections of γ -interferon, radiation therapy, minocycline, oral methotrexate, and phototherapy [9]. It is well known that patients with classic LyP have an increased risk of developing lymphoma; however, this relation is not so clear with localized LyP. To date, only one patient with localized LyP has developed lymphoma, and one has developed mycosis fungoides [2, 10].

All of this make the diagnosis of PALP challenging, and it could be difficult to distinguish from localized LyP, lymphomatoid pityriasis lichenoides, and papular or transformed MF (Table 1), sometimes presenting more similarities than differences. [1, 11, 12].

In conclusion, PALP may be more common than previously believed because a precise definition of the condition has not been established. PALP should be considered in the differential diagnosis of a regional and persistent papular eruption. It is still controversial whether PALP is a subtype of

localized LyP or an own entity. In any case, there is a risk of developing malignancy, so at least a cautious long-term follow-up is recommended.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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