Linear Lichen Planopilaris of the Face: Case Report and Review

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Introduction

Linear lichen planopilaris of the face is a rare variant of lichen planopilaris (LPP) and only few cases have been reported in the literature. The etiology is unknown and is characterized by pigmented follicular papules that progress to atrophy in a linear distribution. Histological findings are similar to those found in LPP, but this rare variant predominantly affects middle-aged men.
Case Report

A 45-year-old man presented to the Dermatology Clinic with a 5-month history of a pruritic unilateral linear erythematous lesion on the chin and mandibular area. Physical examination showed a 0.5 × 12 cm linear well-defined papular pigmented lesion with atrophy and absence of beard hair (fig. 1a). The rest of the skin, oral mucosa and nails were normal. The patient was otherwise healthy.

Dermoscopy showed areas of cicatricial alopecia with absence of follicular openings, perifollicular erythema and pigment (fig. 2a). A skin biopsy specimen showed an intense lymphocytic infiltrate around the upper portion of the hair follicles, vacuolization of the basal layer and destruction of the outer epithelial sheets of the follicles (fig. 3, 4). Atrophic follicles and fibrosis were also present (fig. 5).

The diagnosis of linear LPP of the face was made. The patient initially received treatment with high-potency topical steroids, and then changed to topical tacrolimus (0.1%) to minimize corticosteroid side effects, and oral hydroxychloroquine (200 mg once daily) with partial improvement. For cosmetic purposes we treated the red coloration with pulse dye laser (Syneron Candela 585 nm) and the atrophy with hyaluronic acid filler (Restylane Perlane) with good results (fig. 1b).

Discussion

LPP or lichen planus follicularis, a subtype of lichen planus, is an inflammatory condition characterized by patchy or diffuse hair loss with erythematous papules and keratotic follicular lesions that evolve to cicatricial alopecia [1, 2]. Although rare, it represents 25% of the cases of cicatricial alopecia [3] and is more frequent in women (70–80%), with a peak age of onset between 30 and 60 years [1, 4]. The etiology of this condition is poorly understood. It has been proposed that it is caused by an autoimmune reaction against follicular antigens mediated by T-lymphocytes [1, 4, 5].

Traditionally, it can be divided into three different clinical types: classic type, frontal fibrosing alopecia, and Graham-Little-Piccard-Lassueur syndrome [2, 3]. Other variants reported in the literature include scarring alopecia of the vulva [6], lichen planus follicularis tumidus [7], and linear LPP of the face.
**Fig. 3.** **a** Presence of folliculocentric lichenoid infiltrate at the infundibulum sparing the lower portion (hematoxylin and eosin stain, original magnification ×20). **b** Close-up showing perifollicular lymphocytic infiltrate, vacuolization of basal cells and pigment incontinence (hematoxylin and eosin stain, original magnification ×40).

**Fig. 4.** Lymphocytic infiltrate that predominates in the infundibulum and does not involve the sebaceous glands (hematoxylin and eosin stain, original magnification ×20).

**Fig. 5.** Fibrosis and atrophic hair follicles are seen (hematoxylin and eosin stain, original magnification ×10).

**Table 1.** Reported cases of LPP of the face

<table>
<thead>
<tr>
<th>First author</th>
<th>Year</th>
<th>Sex</th>
<th>Age, years</th>
<th>Histopathology</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pullmann [8]</td>
<td>1977</td>
<td>M</td>
<td>47 and 46</td>
<td>LPP</td>
<td>Peeling and dermabrasion</td>
</tr>
<tr>
<td>Küster [19]</td>
<td>1989</td>
<td>M</td>
<td>52</td>
<td>LPP</td>
<td>Not treated</td>
</tr>
<tr>
<td>Gerritsen [18]</td>
<td>1998</td>
<td>M</td>
<td>44</td>
<td>LPP</td>
<td>Topical steroids, topical tretinoin</td>
</tr>
<tr>
<td>Yanaru [16]</td>
<td>1999</td>
<td>M</td>
<td>53</td>
<td>LPP</td>
<td>Cyclosporine A</td>
</tr>
<tr>
<td>Infante [20]</td>
<td>2009</td>
<td>M</td>
<td>56</td>
<td>LPP</td>
<td>Topical pimecrolimus</td>
</tr>
<tr>
<td>Cañadas [9]</td>
<td>2010</td>
<td>F</td>
<td>14</td>
<td>LPP</td>
<td>Tacrolimus</td>
</tr>
<tr>
<td>Junco [10]</td>
<td>2010</td>
<td>F</td>
<td>43</td>
<td>Pigmented LPP</td>
<td>Topical steroids, topical tretinoin</td>
</tr>
<tr>
<td>Zhao [14]</td>
<td>2012</td>
<td>M</td>
<td>41</td>
<td>LPP</td>
<td>Hydroxychloroquine, topical tacrolimus</td>
</tr>
<tr>
<td>Andrews [21]</td>
<td>2013</td>
<td>M</td>
<td>46</td>
<td>LPP</td>
<td>Topical tretinoin</td>
</tr>
</tbody>
</table>
Linear LPP of the face is a rare variant of LPP first described by Pullmann and Gartmann in 1977 [8], and to our knowledge, this is the 14th case reported in the literature (table 1). In contrast to LPP, linear LPP of the face is more common in men between 33 and 56 years, with only three female patients described in the literature [9–11].

Physical examination reveals pigmented red-brown papules, perifollicular erythema, scaling and secondary atrophy in a linear distribution [8, 9]. Although usually asymptomatic, it can be associated with itching or pain. LPP of the face appears most commonly on the cheeks, mandibular area and chin. Only two cases on the trunk have been documented [2, 12]. It has been proposed that it may appear following the lines of Blashko and in areas of koebnerization [13, 14].

The clinical differential diagnosis includes red linear lichen planus, linear atrophoderma of Moulin and linear morphea. Red linear lichen planus never resolves leaving skin atrophy, linear atrophoderma of Moulin is more common in children and predominates on the trunk and extremities, and linear morphea presents with skin sclerosis [13, 14].

Histological findings are the same as the ones found in LPP with folliculocentric lichenoid infiltrate at the level of the infundibulum sparing the lower portion, destruction of the basement membrane with vacuolization of the basal layer and damage of the outer epithelial sheets of the follicles [14, 15]. Hypergranulosis, acanthosis, hyperkeratosis, Civatte bodies, pigment incontinence, and follicular plugging can be seen. There are only a few reports of the immunofluorescence in this entity and the results are not consistent. Immunofluorescence is important to differentiate LPP from lupus erythematosus.

There is no generally accepted treatment regimen for linear LPP of the face. Successful regimens reported include oral cyclosporine [16], topical tacrolimus and pimecrolimus [9], hydroxychloroquine or topical steroids [14, 17]. Little success was reported with tretinoin cream [18]. The lesions of the patients reported by Küster et al. [19] and Giménez-Garcia et al. [13] improved spontaneously without therapy.

Prognosis of disease is uncertain and permanent atrophy has an esthetic impact on the patients. We propose the use of laser and fillers to improve the cosmetic results.

Statement of Ethics

The patient’s consent was obtained. The authors have no ethical conflicts to declare.

Disclosure Statement

The authors have no conflicts of interest to disclose.

References