Case Report

A Case of Infraorbital Lichen Sclerosus

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Abstract

We present a 57-year-old man with erosive lichen sclerosus isolated to the infraorbital area.

Keywords: Lichen sclerosus, infraorbital, extragenital, hypopigmented patch

Introduction

Lichen sclerosus (LS) is a chronic, inflammatory disorder of the anogenital and extragenital skin. Women are more commonly affected than men with the highest prevalence occurring in the peri- and post-menopausal period. The lesions of LS typically begin as polygonal papules that coalesce into porcelain white plaques. Edema, telangiectases and comedo-like plugs may commonly accompany the primary eruption. Although, LS most commonly occurs around the anus, vulva, and foreskin of the penis, approximately 15% of patients present with extragenital lesions of the neck, shoulders, and upper torso (1,2). Most cases of extragenital LS are asymptomatic, but pruritis occasionally occurs.

Case Report

A 57 year-old man presented to an ophthalmologist for a hypopigmented lesion of the right lower eyelid. The clinician reported the lesion as subtle and no further evaluation was recommended. Ten months thereafter the patient re-presented to the ophthalmologist for a 3.0 mm erosion with associated crust that had developed in the middle of the lower eyelid lesion. A biopsy was performed and the patient was referred to a dermatologist. Physical exam demonstrated an infraorbital hypopigmented patch within which was a surgical biopsy site with suture (Fig 1). The lesion was not sclerotic to palpation and there was no associated erythema or telangiectasia. Examination of the patient’s remaining skin surface, with special attention to the anogenital region, revealed no evidence of pathologic lesions. He was treated with topical tacrolimus with minimal benefit.
Histologic examination of the biopsy demonstrated characteristic features of LS including thinning of the epidermis with loss of rete ridges, mild focal vacuolar degeneration, edema and hyalinization of the papillary dermis, ectatic thin walled vessels, and a mid-dermal lymphohistiocytic infiltrate (Fig 2 and 3).

Figure 1. Infraorbital hypopigmented patch with central biopsy site and suture.

Figure 2 and 3. 2, Epidermal atrophy with associated loss of rete ridges and a pale homogenized papillary dermis with a lymphohistiocytic infiltrate. (Hematoxylin-eosin stain.) 3, Mild focal vacuolar degeneration is seen at the dermal-epidermal junction with focal dermal pigment incontinence (Hematoxylin-eosin stain.).

Discussion

This case represents the third reported patient with exclusive infraorbital LS and the first reported case occurring in a man [1,3,4]. In all three reported cases the lesion presented on the right infraorbit as a well-demarcated hypopigmented patch in patients ranging in age from 12 to 57 years (Table 1). Histologically, all three cases displayed classic features including epidermal atrophy, loss of rete ridges, and homogenization of the papillary dermis. Treatment with methylprednisolone resulted in complete resolution of the lesion in the case described in Kim Y. et al. However, treatment with tacrolimus and corticosteroids resulted in minimal to no benefit in the current case and the one described by Klenk-Pfeifer E., respectively (Table 1).

Exclusive extragenital LS has been reported to occur in up to 15% of patients presenting with LS [1]. Women are more commonly affected than men and the most common extragenital sites include the neck, shoulders, arms, breasts, and upper portion of the trunk. The strong association between autoimmune disease and LS highlights the importance of the dermatologic exam in patients with autoimmune disease [1,2].

Thus far, there have been no reported cases of malignant transformation in extragenital LS. In contrast, genital LS is associated with an increased risk of squamous cell carcinoma and verrucous carcinoma [2]. It is interesting to note that despite the apparent higher prevalence of LS in women the risk of malignancy seems to affect both sexes equally with vulvar and penile malignancy occurring in up to 5% in both women and men, respectively [2].

Clinical and histopathologic studies have suggested that there may be subtle differences in the pathobiology between genital and extragenital forms of LS. Patients with extragenital LS are commonly asymptomatic but in rare cases present with pruritis. In contrast, genital LS commonly presents with pruritus, dyspareunia and genital bleeding in women and phimosis, painful erections and meatal stenosis in men [1,2]. On dermoscopy, extragenital LS more commonly shows scales, keratotic plugs, chrysalis structures, and erosions, but it less commonly shows linear and dotted vessels than its genital counterpart [5]. It has also been demonstrated by histology that there is more epidermal atrophy and decreased rete ridges, but there are fewer associated dermatoses observed in extragenital LS [6]. Furthermore, immunohistochemical studies have demonstrated decreased immunoreactivity to Ki-67 and p53 in extragenital LS suggesting that there are differences in cell proliferation rates between genital and extragenital sites [6]. This finding may partly explain the observed difference in risk of malignancy by location.
Table 1. Summary of reported cases of exclusive infraorbital lichen sclerosus

<table>
<thead>
<tr>
<th>First author</th>
<th>Age/Sex</th>
<th>Race</th>
<th>Laterality</th>
<th>Size</th>
<th>Morphology</th>
<th>Histology</th>
<th>Treatment</th>
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</thead>
<tbody>
<tr>
<td>Klenk-Pfeifer E.</td>
<td>12/F</td>
<td>NS</td>
<td>Right</td>
<td>2.5 x 1.5 cm</td>
<td>Well-demarcated, porcelain-white, coarse patch with comedone-like follicles</td>
<td>Typical results of lichen sclerosus</td>
<td>Topical and injected corticosteroids without improvement</td>
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<tr>
<td>1974</td>
<td></td>
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</tr>
<tr>
<td>Kim Y.</td>
<td>28/F</td>
<td>Korean</td>
<td>Right</td>
<td>NS</td>
<td>Well-demarcated, linear, porcelain white, atrophic patch</td>
<td>Compact hyperkeratosis, loss of rete ridges, atrophy of stratum spinosum and homogenization of collagen in papillary dermis</td>
<td>Methylprednisolone 0.1% cream for one month with complete resolution of lesion</td>
</tr>
<tr>
<td>2007</td>
<td></td>
<td></td>
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<td></td>
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<td></td>
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<tr>
<td>Current case</td>
<td>57/M</td>
<td>White</td>
<td>Right</td>
<td>NS</td>
<td>Well-demarcated, hypopigmented, non-sclerotic patch</td>
<td>Epidermal atrophy, loss of rete ridges and edema and homogenization of the papillary dermis</td>
<td>Topical Tacrolimus with minimal benefit</td>
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NS: Not stated

Conclusion

We present this case to demonstrate a unique site of presentation and raise awareness of exclusive extragenital LS. It should be emphasized that 15% of patients with LS present with only extragenital lesions most commonly on the neck, shoulders, and rarely on the face.

References