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Case Report

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Lichen Sclerosus Presenting as Vitiligo: A Case Series

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Abstract

Depigmenting skin lesions have a limited differential in dermatology. Although the depigmenting process of vitiligo can be clinically striking, it is at best a cosmetic issue, and may or may not be indicative of other pertinent autoimmune process. However, the early diagnosis of lichen sclerosus (LS) is of the utmost importance, since it is associated with potentially severe pain, disfigurement and relatively increase risk of squamous cell carcinoma. We present a series of biopsy proven five cases of LS that clinically presented as vitiligo.

Introduction

LS is a chronic inflammatory dermatosis that classically presents with chalk-white plaques, epidermal atrophy and follicular plugging. Recalcitrant genital cases may result in severe pain and dysfunction [1]. Also, due to its association with squamous cell carcinoma, it is of utmost importance to obtain early recognition and diagnosis of LS [2]. Early intervention and therapy is life-altering [2]. On the other hand, vitiligo is the loss of Melanocytes resulting from cytotoxic T cells in genetic predisposed individuals [3]. Although it presents clinically with depigmented patches, the delay in its diagnosis does not portend a poor prognosis [4]. Unlike LS, it is not associated with scarring, dyspareunia, dysuria, intractable pruritus or squamous cell carcinoma [4]. Also, the first line therapy for both vitiligo and LS are similar, which is classically topical steroids [1,5]. Without a biopsy to confirm the diagnosis, the cutaneous depigmentation may be treated, but the underlying risks may be left unaddressed indefinitely. Therefore, it is our recommendation that any depigmented patch in mucosal skin warrants a biopsy to differentiate conclusively between LS and vitiligo.

Case Series

Case 1

Miss A, a 5 -year- old girl presented with depigmented patches on the genetalia. Initially, she was given clobetasol propionate 0.05% ointment (Dermovate) and showed some improvement. On a subsequent visit, the girl started to complain of pruritus. Accordingly, Dermovate ointment was hold and skin biopsy was taken. The result of the biopsy was consistent with LS.

Case 2

Mrs B, a 28-year-old married lady presented to our clinic with three years history of white patches over the face, upper and lower limbs, and trunk. At that time she was diagnosed to have vitiligo and underwent narrowband ultraviolet B phototherapy for several months; however, there was no improvement. Consequently, skin biopsy from the right arm was obtained. Microscopic examination showed upper dermal fibrosis and pigment incontinence. Melanocytes are normal in number. The features are suggestive of LS.

Case 3

Miss C, an 11- year- old girl came with depigmented spots on the thigh and peri-anal area. The differential diagnosis includes vitiligo and LS. Skin biopsy was taken. Microscopic examination revealed intact hyperpigmented epidermis. The dermis exhibits heavy sclerosis and band like chronic inflammatory infiltrate with some scattered melanophages. LS diagnosis was made.

Case 4

Mr. D, a 23-year-old single male presented to our clinic with two years history of hypopigmented patches on the left side of the neck with thick texture and violaceous colored border. The differential diagnosis at that time was LS, morphea or vitiligo. Consequently, skin biopsy was taken and showed fibrous and hyalinization mainly within the upper dermis and patchy lymphoid infiltrate. LS was diagnosed based on these findings.

Case 5

Miss E, a 4-year-old girl came with hypopigmentation over the labia majora. Vitiligo and LS were considered in the differential diagnosis. Skin biopsy confirmed LS.

Discussion

LS of the mucosal skin is a debilitating disorder [6]. Although it may present initially with hypo-or depigmented patch of skin, one is obligated to biopsy such a lesion to provide a definitive diagnosis [6]. In such a case, vitiligo is a diagnosis of exclusion [6]. There have also been other previous reports that LS can masquerade as other diseases, such as early mycosis fungoides [6]. Only a biopsy is sufficient to

differentiate between LS and cutaneous T cell lymphoma [6]. Despite a pre-existing family history of vitiligo, one of our patients had a biopsy of lesion that classically presents with the phenotype of vitiligo, which histopathologically showed LS. This further emphasizes that it is imperative to biopsy a clinically suspicious lesion prior to the introduction of therapy. Only with proper diagnosis can an appropriate therapeutic course be implemented.

LS has also been shown to clinically simulate lichen planus [7]. It is also pertinent that LS can overlap with morphea (localized scleroderma) [8]. Classically, LS presents with a band-like lichenoid infiltrate at the dermo-epidermal junction, compact hyperkeratosis and prominent papillary dermal edema, which eventually results in homogenous fibrosis1. This histopathological picture is characteristically different from the scenario associated with vitiligo. Since they are very different, one is obligated to do a biopsy, since the treatment approaches and resulting consultations may differ.

It is important to know that although topical calcineurin inhibitors have been shown to be efficacious in both vitiligo and LS, their use in a case of biopsy-proven LS may be discouraged [1,9]. The FDA added a black-box warning to topical calcineurininhibitors and their connection to neoplasia, which presents a clinical conundrum [10]. Since LS is a chronically debilitating disease, the long term use of the topical calcineurin inhibitors may pose a theoretical, albeit important, risk of lowering the threshold for the development of squamous cell carcinoma [2,10].

Furthermore, the frequency of autoimmune diseases in those suffering from vitiligo is increased [11]. These autoimmune diseases include autoimmune thyroiditis, diabetes mellitus, pernicious anemia, systemic lupus erythematosus, and Addison disease [11]. Therefore, it is advisable to investigate for these medical illnesses in patients with vitiligo for early detection and appropriate consultations [11]. Additionally, there is increased incidence of auto-immune antibodies in LS and an association with auto-immune disease such as vitiligo [12,13].

In conclusion, this case series identifies that LS can clinically mimic vitiligo. Therefore, all depigmented mucosal lesion should be biopsied to allow for the proper diagnosis. Furthermore, treating all depigmented patches as vitiligo, without a conclusive biopsy, may delay the diagnosis of LS, which potentially could have devastating consequences.

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