Discoid Lupus Erythematosus of The Ears
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Abstract
Discoid Lupus Erythematosus is characterized by scaly, discoid plaques occurring over sun exposed areas such as scalp, face and ears. Here, we present a case of Discoid Lupus Erythematosus of the ears. We would like to highlight the occurrence of lupus erythematosus of the ear with the presentation similar to a non-infective aetiology.

Skin biopsy was performed over the lesion and the diagnosis was confirmed with histopathological examination. We seek to characterize the presentation and treatment in hope for better understanding and treatment.

Keywords: Lupus erythematosus; External ear disease

Introduction
Lupus Erythematosus (LE) is a chronic, autoimmune, multisystem disease with immunological abnormalities. It is classified as a connective tissue disease into two broad spectrums that presents clinically as systemic manifestations, which most of us are familiar with and another form which is cutaneous manifestations. The disease often has a chronic and relapsing course that can be induced or aggravated by ultraviolet light.

The diagnosis of systemic manifestation is based on Systemic Lupus International Collaborating Clinics 2012 (SLICC) [1]. Classification as having Systemic Lupus Erythematosus by the SLICC criteria requires either that a patient satisfy at least 4 of 17 criteria, including at least 1 of the 11 clinical criteria and one of the six immunologic criteria, or that the patient has biopsy-proven nephritis compatible with Systemic Lupus Erythematosus in the presence of antinuclear antibodies (ANA) or anti-double-stranded DNA (dsDNA) antibodies.

According to American dermatologists Gilliam and Sontheimer, cutaneous manifestations of Lupus Erythematosus can be divided into Lupus Erythematosus-specific and Lupus Erythematosus-non-specific skin manifestations [2, 3].

Lupus Erythematosus-specific skin manifestations can be further subdivided into Acute Cutaneous Lupus Erythematosus (ACLE), Subacute Cutaneous Lupus Erythematosus (SCLE) and Chronic Cutaneous Lupus Erythematosus (CCLE).

Acute Cutaneous Lupus Erythematosus may be localized most often as a malar or a generalized rash. Subacute Cutaneous Lupus Erythematosus is highly photosensitive, with predominant distribution on the upper back, shoulders, neck, and anterior chest. This is frequently associated with positive anti-Ro antibodies and may be induced by a variety of medications.

In Chronic Cutaneous Lupus Erythematosus, the most common form is Classic Discoid Lupus Erythematosus (DLE) [4]. Chronic Cutaneous Lupus Erythematosus is characterized by sharply circumscribed, scaly, infiltrated and later atrophic (‘discoid’) plaques. These plaques are usually occurring on sun-exposed areas such as scalp, face, and ears with the characteristic scarring and

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pigmentary changes. Less common forms of Chronic Cutaneous Lupus Erythematos include Hyperkeratotic Lupus Erythematosus, Lupus Tumidus, Lupus Profundus and Chilblain Lupus.

Common cutaneous disease associated with, but not specific for Lupus Erythematosus includes vasculitis, livedo reticularis, alopecia. There can also be digital manifestations such as periungual telangiectasia, Raynaud phenomenon, photosensitivity, and bullous lesions.

In most cases this skin disorder is purely cutaneous without any systemic involvement. It is vital to confirm the diagnosis of cutaneous Lupus Erythematosus by a skin biopsy as there can be several differential diagnosis in dermatological diseases of the external ear. They include infectious, tumor and noninfectious inflammatory.

A recent study has shown that about 25% of the Discoid Lupus Erythematosus patients are already diagnosed with Systemic Lupus Erythematosus when they get their Discoid Lupus Erythematosus diagnosis [5]. The probability of receiving an additional diagnosis of Systemic Lupus Erythematosus was 18.1% during the first three years after being diagnosed with Chronic Lupus Erythematosus.

Case Report

An otherwise healthy 27 years old male smoker presented with a three years history of bilateral ear discomfort with intermittent pruritus. He denied having hearing loss, ear discharge, tinnitus or vertigo. There was no past medical history to suggest atopy. This patient had consulted several general practitioners over the last three years and was treated as otitis externa with a trial of oral antibiotics, anti-inflammatories and anti-fungal, all to no effect.

Ear examination showed bilateral hyperpigmented, irregularly shaped scarring plaques on the left concha (Figure 1). The otoscopy findings were unremarkable.

Incidentally, examination of the facial skin revealed two well-demarcated, erythematous atrophic papules with central keratotic plugging on the right cheek, adjacent to the right nasolabial fold as well as on the vermilion border of the left side of the upper lip (Figure 2). These lesions appeared much later than the ones in the ears, approximately one year ago. There were no other lesions identified elsewhere on the body.

A skin biopsy from the left concha revealed focal basal cells vacuolation with perivascular lymphocytic infiltration suggesting a diagnosis of discoid lupus erythematosus (DLE) (Figure 3).

Another punch biopsy performed subsequently on the atrophic papule on the right cheek demonstrated marked hyperkeratosis, parakeratosis with mild acanthosis of the epidermis layer. There is basal cells lichenoid cells infiltration. There is prominent perifollicular and periappendageal lymphocytes infiltrations with interstitial mucin deposition. There were no malignant cells seen.

Immunofluorescence study shows Immunoglobulin M, weak Immunoglobulin G and Immunoglobulin A deposition at dermoepidermal junction, all of which are features of a Discoid Lupus Erythematosus lesion.

Further questioning did not reveal any symptoms of systemic lupus and his blood investigations were also within normal

limits (Hemoglobin 17.1g/dL,: Total White Blood Cells 7500, Platelet 200000/mm³ with Erythrocyte Sedimentation Rate :10mm/hr, Creatinine 81µmol/L and Urea 5.0mmol/L. This includes a non-reactive antinuclear antibody.

He was commenced on topical corticosteroids and was advised to stop smoking. He is on follow-up 3 monthly with regression of plaques over the mentioned areas.

Discussion

Discoid Lupus Erythematosus is the most common subtype of Chronic Lupus Erythematosus and 60-80% of lesions is localized above the neck [2, 3]. It is estimated 70-90% of the patients suffer from photosensitivity and areas such as the scalp, ears and cheeks are the most commonly involved areas as these are sun exposed areas.

Discoid Lupus Erythematosus is a chronic inflammatory disease consisting of fixed, indurated, erythematous papules and plaques that are often distributed on the head and neck, although any cutaneous region can be affected. Without intervention, Discoid Lupus Erythematosus lesions may last for many years and are associated with extensive scarring, a feature that helps distinguish Discoid Lupus Erythematosus from Subacute Cutaneous Lupus Erythematosus (SCLE) [4].

When Discoid Lupus Erythematosus occurs on the scalp, permanent scarring alopecia occurs. Pigmentary changes of both hyperpigmentation and hypopigmentation are also frequently associated with lesions of Discoid Lupus Erythematosus. The external ears are often involved in Discoid Lupus Erythematosus, like in our patient [5]. Thus, this area should be carefully examined in patients with suspected Discoid Lupus Erythematosus. In most patients, the initial discoid lesions appear on sun-exposed areas of the head and neck as well as upper limbs and only tend to involve the concha later during the course of disease. This patient had a rather unusual presentation with the initial lesions involving the ears first.

There are many cutaneous manifestation of external ear disease as enumerated in Table 1. [3,5].

With the above mentioned differential diagnosis, a biopsy will confirm the histopathological diagnosis. This is important in our patient as there are reported cases in literatures of squamous cell carcinoma developing in Chronic Discoid Lupus Erythematosus [6-8]. Hence, the need for close follow-up with repeat biopsies should be performed if there is failure to respond to conventional therapy.

As with other types of cutaneous lupus erythematosus, sunscreens and sun-protective measures are the foundation of therapy [9]. Potent topical steroids and intralesional corticosteroids are often helpful. Anti-malarial drugs are also used. Less often, therapy with immunosuppressive medications or systemic corticosteroids is offered [6, 9]

Conclusion

A clinical diagnosis of discoid lupus is difficult to make as most of us are only familiar with Systemic Lupus Erythematosus. Here, the criteria and its application is well established. The diagnosis of Systemic Lupus Erythematosus is based upon the judgment of an experienced clinician who recognizes characteristic constellations of symptoms and signs in the setting of supportive serologic studies, after excluding alternative diagnoses. This is often challenging due to the great variability in the expression and severity of Systemic Lupus Erythematosus.

As skin is the largest organ, it is vital to appreciate various skin disorders that has a predilection to occur in the head and neck region.

Repeated treatment of otitis externa which has no improvement

<table>
<thead>
<tr>
<th>Infectious Dermatologic Diseases of The External Ear</th>
<th>Tumoral Dermatologic Diseases of The External Ear</th>
<th>Non-Infectious Inflammatory Dermatologic Diseases of The External Ear</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Acute diffuse otitis externa</td>
<td>(A) Benign</td>
<td>• Cold injury of the external ear</td>
</tr>
<tr>
<td>• Chronic otitis externa</td>
<td>• Seborrheic keratosis</td>
<td>• Burn injury of the external ear</td>
</tr>
<tr>
<td>• Furunculosis</td>
<td>• Granuloma fissuratum</td>
<td>• Seborrheic dermatitis</td>
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<tr>
<td>• Otomyocis</td>
<td>• Keloid</td>
<td>• Contact dermatitis</td>
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<tr>
<td>• Lymphocytoma</td>
<td>• Chondrodermatitis nodularis helicis chronicus</td>
<td>• Atopic dermatitis</td>
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<tr>
<td>• Auricular chondritis and perichondritis</td>
<td>• Melanocytic nevus</td>
<td>• Acne</td>
</tr>
<tr>
<td>• Lupus vulgaris</td>
<td>• Blue nevus</td>
<td>• Lupus erythematosus</td>
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<tr>
<td>• Herpes zoster oticus</td>
<td>• Cylindroma</td>
<td>• Psoriasis</td>
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Table 1: Cutaneous manifestation of external ear disease.
would require further investigations and assessment with a consultation from otorhinolaryngologists and dermatologists. A biopsy will confirm the histopathological diagnosis and close follow up is warranted.

References


