HISTOPATHOLOGICAL REVIEW ON DISCOID LUPUS ERYTHEMATOSUS MIMICKING GRANULOMA FACIALE: ONE CASE REPORT

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ABSTRACT

Discoid lupus erythematosus (DLE) is one of the most common forms of cutaneous lupus erythematosus. It is characterized by clinical manifestations of erythematous macules, papules, or plaques with a coin-like shape, and the face is the most common predilection site. Clinical features often resemble other lupus types and granuloma faciale. This case report aimed to distinguish discoid lesions on the face based on the histopathological examination. A 71-year-old male with a few reddish lumps appeared on his face three months ago. Physical examination showed multiple discrete erythematous plaques with overlying squamous. *Hematoxylin* and *eosin* staining on the epidermis demonstrated basket weave type orthokeratosis, basal vacuolar cell degeneration, epidermal atrophy with flat rete ridges, and follicular plugging while in the dermis obtained inflammatory cell infiltrates, especially in *periadnexal* areas. *Histopathological* features of DLE are *hyperkeratosis,* pilosebasea gland atrophy, follicular plugging, basal membrane thickening, and cellular infiltrate in *periadnexal* or perivascular areas more visible than in other types of CLE. In DLE, subepidermal green zone and eosinophil infiltrate were not found, like histological features of granuloma faciale. Histopathological examination can be used to establish a diagnosis for discoid lesions on the face, although serology examination remains the gold standard.

Keywords: Discoid lupus erythematosus; Granuloma Faciale; Histopathology

INTRODUCTION

Lupus erythematosus (LE) is an inflammatory disease of connective tissue involving autoantibody pathogens and immune complexes, which affect the entire system in the body, with the skin as the dominant organ affected.[1] Cutaneous lupus erythematosus is classified into three categories, namely: *acute cutaneous lupus erythematosus* (ACLE), *subacute cutaneous lupus erythematosus* (SCLE), and *chronic cutaneous lupus erythematosus* (CCLE). The incidence of LE in the United States varies from 2 to 7.6 cases per 100,000 people annually, and cutaneous manifestations range from 72% to 85% in SLE patients and about 23% to 28% of patients show skin symptoms as the first symptoms of SLE.[2] Establishing the diagnosis of CLE is based on clinical symptoms, histopathological and immunohistological examinations of skin lesions, and serological examination.[3] *Discoid lupus erythematosus* (DLE) is a form of *chronic cutaneous lupus erythematosus* (CCLE) that is a skin lesion in the form of the macule, papule, or erythematous plaque with a coin-like shape (discoid) and the predilection site is commonly on the face. The surface of the lesion is generally a thin squama. In its development, the lesions can become atrophy, depigmentation, and scar.[1] A discoid lesion with predilection on the face can resemble facial granuloma disorder, which is benign chronic vasculitis cutaneous disease and has a typical morphological pattern with unknown causes.[4] The results of histopathological examinations of each type of LE can show overlapping results. Therefore, it is necessary to combine clinical, histopathological, and serological examinations in making the definitive

diagnosis. LE histopathological examination can show a feature of epidermal atrophy with hyperkeratosis and follicular plugging, dermatitis interface with single keratinocyte necrosis, vacuolar degeneration in dermal-epidermal junction, thickening of basal membranes, infiltration of lymphocytes in the dermis region, especially periadneksa and perivascular, as well as degenerative changes in the connective tissue under the epidermis. This case report aimed to distinguish discoid lesions in the face by histopathological examination.

CASE PRESENTATION

A 71-year-old farmer presented to the Dermatovenereology outpatient clinic of Dr. Moewardi Hospital Surakarta with the complaint of a reddish lump on his cheeks since three months ago that sometimes itchy. One month later, the reddish lump widened, multiplied, some scaly, and appeared on some other face areas. The reddish lump is felt increasingly red and itchy, especially when the Patient does activity under the sun. This Patient experienced the same complaints about a year ago, especially when he was often exposed to sunlight. At the time he was diagnosed with allergic photodermatitis and given ointments and medication, and then the complaints resolved. There were no histories of allergy, atopy, eczema, numb spots, and sores in his mouth. This Patient is a farmer who works from 07.00 a.m until 1.00 p.m, and he never uses sunscreen. None of his family members has experienced a similar skin disease before. Dermatologic examination obtained multiple discrete erythematous plaques as well as thin squama. (Figure 1)

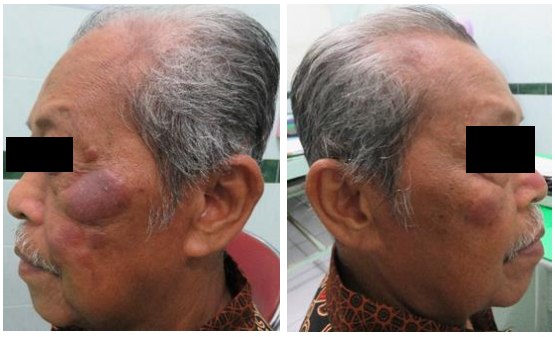

Figure 1. Multiple discrete erythematous plaques, partially covered in fine squama on both cheeks.

Laboratory examination results were unremarkable results within normal limits. Dermoscopic examination of the lesion demonstrated follicular opening accompanied by slightly dilated blood vessels. (Figure 2)

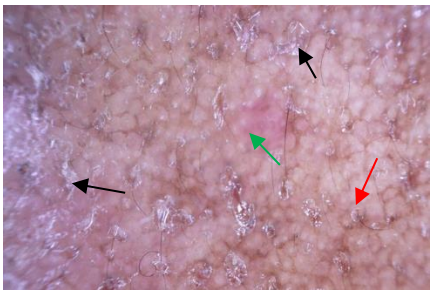

Figure 2.: Dermoscopic findings: widening of the estuary follicle (red arrow), halo in the perifollicular area (green arrow), and squama in several places (black arrow).

Based on all these findings, we temporarily diagnosed this Patient with cutaneous lupus erythematosus, and the differential diagnosis was granuloma faciale. Microscopic preparations of the skin lesions taken from the left cheek using *hematoxylin* and *eosin* staining revealed orthokeratotic basketball-type weave with thinning / epidermis atrophy, rete ridge lander, plugging keratosis. Vacuolar degeneration of basal cells in the epidermis. At the same time, there was visible edema in the dermo-epidermal area in the dermis, melanin pigment in the upper dermis, and periadnexal inflammatory cell infiltrates (Figure 3). Histopathological biopsy results supported the diagnosis of discoid lupus erythematosus.

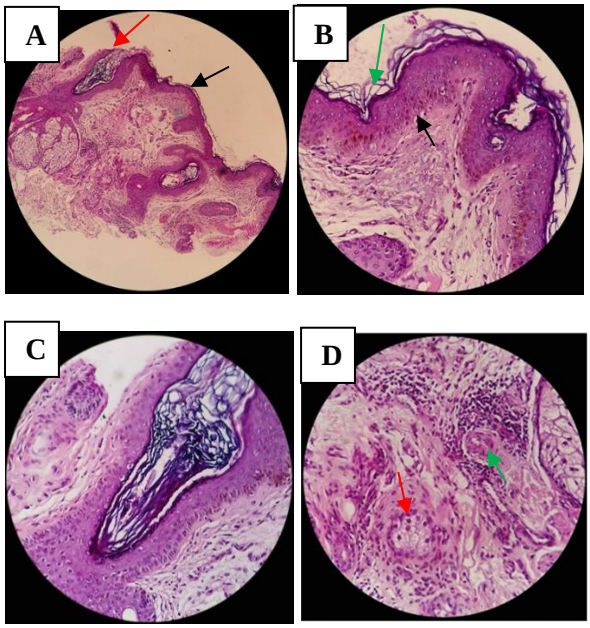

Figure 3. A. Atrophy/thinning of the epidermis (black arrow) and follicular plugging (red arrow) in the epidermis layer (H&E, 10x). **B.** Ortokeratotic basketball type weave (green arrow) and visible degeneration of basal cells (black arrow) in the epidermis (H&E, 40x). **C.** visible follicular plugging (H&E, 40x). **D.** In the dermis: inflammatory cell infiltrate especially in perivascular (green arrow) and periadnexal (red arrow) (H&E, 40x).

DISCUSSION

Lupus erythematosus may have different patterns of clinical and serological manifestations that determine the division of clinical types. It occurs predominantly in women.[1] In this case, CLE occurs in a 71-year-old man. An epidemiological study of the incidence and prevalence of SLE and CLE conducted in the United States reported that CLE incidence in men was higher than SLE and CLE incidence increased with age.[6]

LE pathogenesis is an interaction between genetic, hormones, and the environment with unknown etiology. Genetic susceptibility factors (e.g., haplotypes of human leukocyte antigens) interact with environmental trigger factors (infections, drugs, ultraviolet rays), resulting in decreased immune system tolerance, producing autoantibodies and lymphocyte-specific antigens manifesting in tissue damage, one of which is skin.[7] In this case, our Patient is a farmer who is often exposed to sunlight, and he never uses any protection. Environmental factors, especially sun exposure, play an essential role in the pathogenesis of CLE occurrence. UV rays can trigger keratinocyte apoptosis as well as reduce the clearance of apoptotic cells themselves. UVB radiation induces the release of several pro-inflammatory cytokines and chemokine, including CCL27 (Cutaneous T cell-attracting chemokine) which activates autoreactive T cells.[8]

Our Patient complained about a reddish lump on his face, which had been experienced for three months. In physical examination, we found discrete multiple erythema plaques with squamous on some lesions. Based on these visible skin disorders, we differential diagnosed with *discoid lupus erythematosus* (DLE) and granuloma faciale. A study conducted by Fabri et al.[5] and Walling Sontheimer[12] described several criteria for clinical diagnosis of DLE. A clinical overview of spherical active DLE regarding areas exposed to sunlight and follicular plugging.[9] Granuloma faciale is a chronic benign vasculitis cutaneous disease with a typical morphological pattern and unknown etiologies. The clinical feature of granuloma faciale is brownish-red plaques on the face, especially on areas exposed to sunlight. Sometimes it is accompanied by the dilation of the estuary follicle glands and superficial teleangiectasis. Histopathological examination of granuloma faciale, staining with HE shows a normal epidermal layer in the presence of subepidermal green zones and infiltrates on the superficial and perivascular dermis. Infiltrates can be neutrophils, lymphocytes, plasma cells, and eosinophils as dominant infiltrates.[4]

The histopathological feature of CLE depends on its clinical variation. But in practice, this feature often overlaps, sometimes even difficult to determine the type based on the histopathological finding. A prominent feature of CLE includes:

Liquefaction degeneration of basal epidermal cells, degenerative changes in the connective tissue under the epidermis include (hyalinization, swelling, and fibrinoid changes), lymphocyte infiltration (plasma cells and histiosit) in the dermis area (dominant around the appendix). Other possible features are epidermal atrophy with hyperkeratosis and follicular plugging, which are sometimes accompanied by pilosebaceous atrophy, thickening of the basal membrane, and premature degeneration of collagen connective tissue.[1,10] The comparison of DLE histopathological features also follows the stage of the disease. Hyperkeratosis, pilosebaceous gland atrophy, follicular plugging, thickening of the basal membrane (more visible in PAS staining), and cellular infiltration in periadnexal or perivascular areas are more visible than in SCLE types. In SCLE type, vacuolar cell degeneration, epidermal atrophy, infiltration of the superficial dermis, subepidermal edema, keratinocyte undergoing apoptosis (Civatte bodies) can be more prominent.[5,10]

Histopathological findings of our Patient showed, orthokeratotic basketball type weave with thinning / atrophy epidermis in the epidermis so that the epidermis looked flat. The important histopathological change seen to indicate LE is vacuolar/hydropic degeneration of basal cells. This change is marked by the presence of a vacuolar-shaped space below and between basal cell keratinocytes.[12] The rete ridge pattern looks flatter. Visible follicular plugging as part of epidermal invaginated is filled keratin and associated with estuary hair follicles.[13] Melanin pigment is observed on the upper dermal layer. Inflammatory cell infiltration spread in the dermis area is partially visible in periadneksa, a typical sign of DLE lesions.

In our case, we treated the Patient with high-potency topical corticosteroid therapy, a mometasone furoate cream used twice a day on lesions, and advised him to use sunscreen protectors to prevent a recurrence.

A previous study about CLE in Asian people stated a correlation between clinical, Histopathology, and serologic profile of the various types of CLE. Histopathological examination can be an alternative diagnostic tool to differentiate between various types of CLE, while serology profile has been identified as a marker of systemic involvement of CLE.[14]

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