

A Typical Discoid Lupus Erythromatosus: A Case Report and a Brief Review of the Literature

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Abstract: *We report a case of Atypical Discoid Lupus Erythromatosus. A 49 years old female presented with hyperpigmented and multiple scaly nodular lesions. She recovered very well with short steroid therapy. Discoid Lupus Erythromatosus is a chronic photosensitive skin eruption in exposed areas which can be localized or widespread. DLE can cause permanent scarring if treatment is delayed.*

Keywords; Discoid Lupus Erythromatosus, Lupus Erythromatosus

1. Introduction

Discoid Lupus Erythromatosus is an uncommon sub-acute or chronic auto-immune dermatosis and the most common clinical variant of Lupus erythromatosus. DLE is a benign disorder of skin, clinically characterized by red scaly patches which heal with atrophy, scarring, pigmentary, potentially disfiguring changes and histo-pathologically a vacuolar degeneration of the basal layer of epidermis with patchy dermal lymphocytic infiltrates. It is caused or triggered by exposure to ultraviolet radiation, cold or drugs. Early treatment may prevent and reduce scarring or atrophy.

2. Case Report

A 49 years old female presented with itchy, pigmented, multiple, itchy, nodular and minimally disfiguring lesions over the face, chest, upper back, and outer aspects of both the arms since 10 years. She also complaints of multiple joint pain (large and small joints) since 5 years. Within 2 years of the appearance of facial lesions, she developed numerous lesions on trunk and extremities. She had used high potency topical corticosteroids and various medications but with partial and temporary response.

On clinical examination, the patient was obese with weight of 86 kg and height of 5.2". Patient was conscious, oriented, afebrile with a pulse rate of 86 / min and blood pressure of 130/80 mm Hg. On examination, she had hyperpigmented scaly and erythematous lesions spread over the entire exposed skin surface with few (discoid) plaques on the face,

scalp, trunk and extremities. Few areas showed atrophy and hyperpigmentation at the margins. Palms, soles, nails and mucosa were normal. Systemic examination was unremarkable. Patient was a diagnosed case of hypertension since last 2 years and on regular treatment on temisartan 40 mg OD.

Laboratory evaluation revealed normal leucocyte count with ESR of 55mm at the end of 1 hour, absolute eosinophil count was 71 /cumm. Serum anti-nuclear antibody was positive, anti ds-DNA was also positive, serum rheumatoid factor was positive. Ultrasound of the abdomen showed fatty liver. Thyroid functions showed subclinical-hypothyroidism with a TSH of 10.5 uIU/ml. Colour doppler of both extremities showed normal arterial doppler study. MRI of brain was normal.

Clinical suspicion of Atypical Discoid Lupus Erythromatosus was confirmed. She was put on hydroxychloroquine phosphate 200 mg twice a day and a short steroid therapy. She improved with this treatment, the skin lesions regressed and vanished. She is under regular follow-up.

On discharge she was advised to continue with DMARD's, Anti-hypertensive and oral hypothyroid drugs. Patient recovered well after DMARD's and steroid therapy at the end of one month. Patient has been under followup and after 1 month of treatment, her lesions totally disappeared. We hereby describe a case report of atypical discoid lupus erythromatosus with extensive involvement and

disfigurement, which responded very well to short steroid therapy and hydroxychloroquine.



Figure 1: Pre treatment picture showing itchy, pigmented, multiple nodular lesions over face, both extremities.



Figure 2: Post-treatment- clearance of lesions

3. Discussion

We have discussed a case of Atypical Discoid Lupus Erythromatosus. Discoid lupus erythromatosus is a chronic dermatological disease that can lead to scarring, hairloss, and hyperpigmentation changes in the skin if it is not diagnosed and treated early and promptly. The skin is the second most frequently involved organ in lupus erythromatosus. Although cutaneous involvement is rarely life threatening, it is associated with major morbidity¹. Within the spectrum of disease included in LE, disease confined mainly to the skin and referred to as discoid lupus erythromatosus, whereas at the other end is a florid disease with systemic involvement of kidneys, joints, lungs, heart and brain in systemic lupus erythromatosus (SLE). Although at the benign end of the spectrum, 1 % to 5 % of patients with discoid lupus may develop SLE² and 25 % of the patients with SLE can develop typical chronic discoid lesions at same time during the course of their illness.³

Discoid lupus erythromatosus was first described by Behcet⁴ in 1942. Lupus occurs in all ages with a mean of 21 to 50 years⁵. Women are much more often affected than men.⁶ The prevalence is between 17 to 48 per 100000 people.⁷ Discoid lupus is by far the most common manifestation of SLE⁸.

DLE mainly affects areas exposed to sunlight, like cheeks, nose, ears, arms, neck and back of hands. Any other deviation from this pattern of skin involvement in DLE is referred to as Atypical DLE. It may rarely occur on the palms and soles.⁹ DLE starts as erythromatous papules or plaques, usually on the hand and neck, with an adherent scale. The lesions tends to spread centrifugally and as it progresses there is follicular plugging and also pigmentary changes; generally hyperpigmentation at the periphery and hypopigmentation with atrophy, scarring and telengectasia at the center of the lesion.¹⁰ The lesions are usually asymmetric and they may present with mild pruritis or sometimes pain within the lesions. The scalp may be affected and cause permanent 'scarring alopecia'.

DLE may affect the lips and inside of the mouth, causing ulcers, scaling, and predisposing to squamous cell carcinoma. The incidence of SCC developing in DLE ranges from 3.3-3.4 % in various studies. The interval between developing of DLE and SCC have varied from 4-20 years¹¹.

Histopathological changes are characteristic but depend on the type and age of the lesion. Most of the patients with DLE show a direct positive immunofluorescence in biopsies of lesions but this is not very specific.⁶

Approximately 20 % of patients with DLE have a positive anti-nuclear antibody and raised ESR. Rheumatoid factor may be positive. Blood testes should be repeated periodically, perhaps annually to check for onset of systemic disease.

General measures include sun avoidance and the liberal application of sunscreens. Patients should be educated about the use of sunscreens and protective clothing and behavioral modifications to avoid the precipitating factors, particularly between 10 AM to 4 PM. They should be aware of water, snow and sand surfaces from which the UV rays are reflected.^{12,13} Topical steroids are the mainstay of treatment of DLE. Patients are usually started with a potent topical steroid applied twice a day, then switched to a lower potency steroid as soon as possible. A Cochrain review concluded that flucinonide cream may be more effective than hydrocortisone in treating people with SLE.¹⁴ When systemic treatment is required, Methylprednisolone (Pulse) is the first line of treatment.¹⁴ It is customary to start with hydroxychloroquine at the dose of 200 mg per day for adults and if there are no gastrointestinal intolerance, increase the dose to twice a day.¹⁵ It is important to emphasize to the patient that it may take between 4 to 8 weeks for any clinical improvement. Other possible treatment includes immunosuppressive agents. In 1995, Bottomley and Goodfield¹⁶ found that methotrexate may be helpful to the patients with DLE resistant to conventional treatment. In 1994 Yell and Burge tried cyclosporine in 2 patients with severe DLE and concluded that it was effective at the dose of 4-5 mg/kg/day. Blood pressure and kidney functions need to be monitored and hypertension is the most common side effect. Walker et al¹⁷ reported 2 patients with severe recalcitrant chronic discoid lupus that had not responded to topical steroids or antimalarials but dramatically responded to topical tacrolimus ointment. Goyal and Nousari¹⁸ described 2 cases of refractory discoid lupus involving the palms and soles that responded satisfactorily to mycophenolate mofetil. Azathioprine a potentially toxic drug has been used in refractory cases of discoid lupus, with particular success among those with the involvement of the palms of the hands and the soles of the feet¹⁹.

Surgical options include⁶ excision for burnt out scarring lesions and laser therapy can be considered for lesions with prominent telangiectasias. A minority of patients with DLE (less than 5 %) progress to systemic lupus erythromatosus. In our case short steroid therapy with hydroxychloroquine proved highly effective and the hyperkeratotic plaques lessened promptly within the first 3 months of treatment. ANA and histopathological features guided us to the diagnosis and then treatment with short steroid therapy and hydroxychloroquine helped patients in leading a normal life.

4. Conclusion

DLE is a chronic scarring and potentially disfiguring disease seen in all parts of the world and among all ethnic groups. It is an important cause of irreversible hair loss and is associated with considerable morbidity. It is extremely important to diagnose this relatively uncommon condition early because early effective treatment is important to prevent scarring. Most of the cases respond to short steroid

therapy and DMARDs.

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