Multiple scarring alopecia patches on scalp

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A 39-year-old male patient complained of multiple areas of hair loss on scalp for about 4 years. The lesions were gradual in onset and had slowly progressive course. Topical steroids were used as an initial treatment for about 5 months, but without any significant improvement. Five sessions of interalesional steriods failed to show any improvement, and the patient stopped treatment for about two years. The patient was then treated with combination of potent topical steroids and mesotherapy with PRP for 3 months with significant improvement. There was no past history of similar condition, and family history for similar disease was also negative. Skin examination showed multiple area of alopecia scattered randomly on scalp. The periphery of some lesions were slightly indurated, scaly and pinkish (Fig. 1).

General examination was irrelevant, while routine laboratoratory investigations including CBC, liver and kidney profile were within normal level. In addition, serological investigations for lupus erythematosus and other connective tissue diseases were also negative.

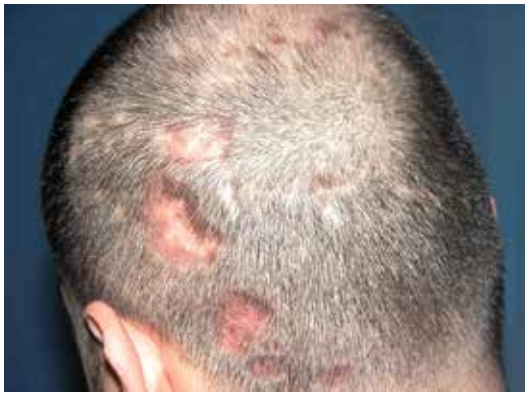

Fig. 1 Multiple non-hairy areas of scalp.

What is the clinical diagnosis?

1. Discoid lupus erythematosus
2. Lichen planus
3. Syphilis

A skin biopsy from the lesion showed mild interface dermatitis, vacuolar alteration at basal cell layer. An epidermal atrophy and hyperkeratosis. Superficial perivascular and periadenxal inflammatory infiltrate formed of lymphohistiocytic cells admixed with few plasma cells. In addition,

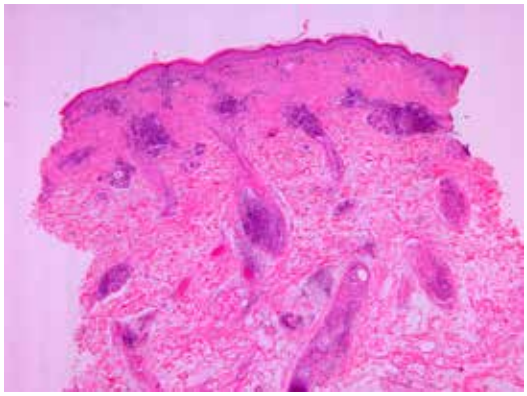

Fig. 2 Mild interface dermatitis with vacuolar alteration at basal cell layer. An epidermal atrophy with hyperkeratosis. Superficial and deep perivascular and periadenxal inflammatory infilterate formed of lymphohistiocytic admixed with few plasma cells. Middle and lower dermis showed mucinous deposiotions inbetween collagen bundles.

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mucin depositions were observed in-between collagen bundles (Fig. 2). PAS stain was positive and revealed eosinophilic thickening of basement membrane (Fig. 3).

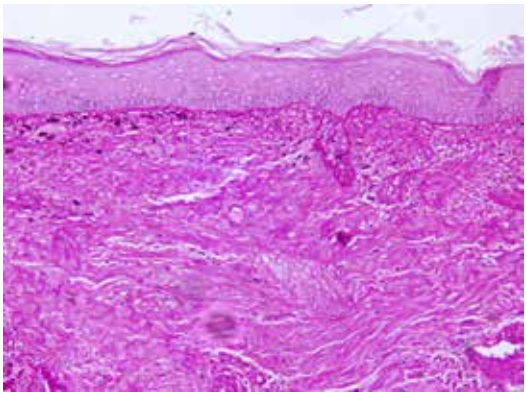

Fig. 3 PAS stain was positive showing eosiniphilic theckining of dermoepidermal junction.

DIAGNOSIS

Discoid lupus erythematosus

DISCUSSION

Discoid lupus erythromatosus (DLE) is a chronic cutaneous disorder that can be complicated by scarring, hair loss, and hyperpigmentation, these changes can occur in the skin if it is not diagnosed and treated early. The skin is the second most frequently involved organ in lupus erythromatosus. Although cutaneous involvement is rarely life threatening, it is associated with major morbidity.[1] 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 SLE[2] and 25% of the patients with SLE can develop typical chronic discoid lesions at same time during the course of their illness.[3]

In in 1942, discoid lupus erythromatosus was described by Behcet.[4]

Lupus occurs in all ages with a mean age of 21 to 50 years.[5] Females are much more often affected than males.[6] The prevelance is between 17 to 48 per 100,000 people.[7] Discoid lupus is by far the most common manifestation of SLE.[8]

Sites exposed to sunlight, like cheeks, nose, ears, arms, neck and back of hands are commonly liable for occurrence of discoid lupus lesions. 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 erythematous 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 development of 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.[11] DLE in our case was located on scalp.

DLE may occur on 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 development of DLE and SCC has varied from 4-20 years.[11]

Microscopic features are characteristic but depend on the type and age of the lesion. Examination of the skin for deposits of immunoreactants is called direct immunofluorescence (DIF). DIF of lesional skin does not replace routine histologic staining as the method of choice for establishing a diagnosis of cutaneous LE. However, in those

cases where the routine histopathology is equivocal, DIF can be a valuable asset in establishing a diagnosis. The most characteristic DIF finding in cutaneous LE is antibody deposition at the dermal-epidermal junction and around hair follicles. These deposits are typically granular, and they are composed primarily of IgG and/or IgM, although IgA can also occasionally be seen. In addition, deposits of complement proteins are to be expected. Some investigators have reported that in SCLE, granular deposits of IgG and IgM are observed primarily within the epidermis rather than at the dermal-epidermal junction. There is evidence that the epidermal deposits are due to anti-SSA/Ro autoantibodies depositing directly within the skin. In active lesions of ACLE, SCLE and DLE, DIF of lesional skin is positive in the majority of cases. In general, a positive DIF supports the diagnosis of cutaneous LE, but a negative DIF does not exclude the diagnosis. It has been noted that DIF is most likely to be positive in well-established, active lesions. DIF is often negative or nonspecific in LE tumidus. In lupus panniculitis, DIF may show immunoreactants around dermal vessels, but granular deposits at the dermal-epidermal junction are not uniformly present.[6]

Around 20 % of patients with DLE have a positive anti-nuclear antibody and raised ESR. Rheumatoid factor may be positive. Laboratory investigations should be repeated periodically, perhaps annually to check for the 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 mainstray 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 Cochrane review concluded that flucinonide cream may be more effective than hydrocortisone in treating patients 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 treatments include immunosuppressive agents. In 1995, Bottomley and Goodfield[16] 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 observed. Walker et al[17] reported 2 patients with severe recalcitrant chronic discoid lupus that had not responded to topical steroids or antimalarials but dramatically improved with topical tacrolimus ointment.

Antimalarial therapy has been used for more than a half-century for cutaneous LE, and it remains the gold standard for systemic therapy. Hydroxychloroquine sulfate is the most commonly chosen antimalarial, as it is usually well tolerated. Chloroquine and quinacrine (mepacrine) are alternatives. In patients, who are not responsive to hydroxychloroquine sulfate, quinacrine may be added to the regimen. Quinacrine can turn the skin yellow,

although it does not invariably do so. The dose of hydroxychloroquine sulfate chosen is usually 200 mg once or twice per day. It has been reported that if the dose does not exceed 6.5 mg/kg ideal body weight/day, eye toxicity is quite unlikely. For chloroquine, the usual dose is 125-250 mg/day, with the eye toxicity-minimizing dose being no more than 3.5-4 mg/kg ideal body weight/day.[17]

Goyal and Nousari[18] described 2 cases of refractory discoid lupus involving the palms and soles that responded satisfactorly 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 feet.[19] Surgical options include[6] excision for burnt out scarring lesions and laser therapy can be considered for lesions with prominent telengiectasias. A minority of patients with DLE (less than 5%) progess to systemic lupus erythromatosus. our patient was treated with hydroxychloroquine and topical tacrolimus and showed marked improvement and scarring areas lessened markedly within the first 2 months of therapy.

DLE can be considered as one of main causes for chronic scarring alopecia all over the world. It is the reason of irreversible hair loss and it is associated with considerable morbidity. Early diagnosis and treatment are recommended to prevent scarring and disfigurement.

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