

Discoid Lupus Erythematosus of the Eyelids

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Introduction:

Discoid lupus erythematosus (DLE) is a benign, autoimmune disorder of the skin. While the face, trunk and extremities are frequently affected, eyelid involvement is uncommon and often presents as a diagnostic problem. Several interesting variants of DLE have been described outside periocular tissues. We present here a patient who developed DLE in the eyelids and conjunctiva and was unsuccessfully diagnosed for a decade and a half.

Case:

A 58-year-old, white female presented with a 15-year history of bilateral ocular and lid redness, pain and progressive thickening of both eyelids (**Figure 1**). Despite numerous consultations and therapy for rosacea, meibomian gland dysfunction, allergic and seborrheic blepharitis, her symptoms continued to worsen. She underwent the first of three lid biopsies in 1990; all biopsies were read as benign, chronic, granulomatous inflammation of the dermis.

In July, 1993, she was referred for diagnosis and management. External examination revealed: 1) bilateral patchy erythema and thickening of the infraorbital skin; 2) erythema, thickening, and lash loss of the lateral lower lid margins; 3) a three to four millimeter papillomatous mass under the right upper lid with anterior keratinization; 4) irregularities of the tarsal conjunctivae; and, 5) a medial lid defect of right lower lid corresponding to biopsy sites. (Figures 1,2) Other ocular findings were normal.

Fig. 1 Bilateral eyelid thickening and redness

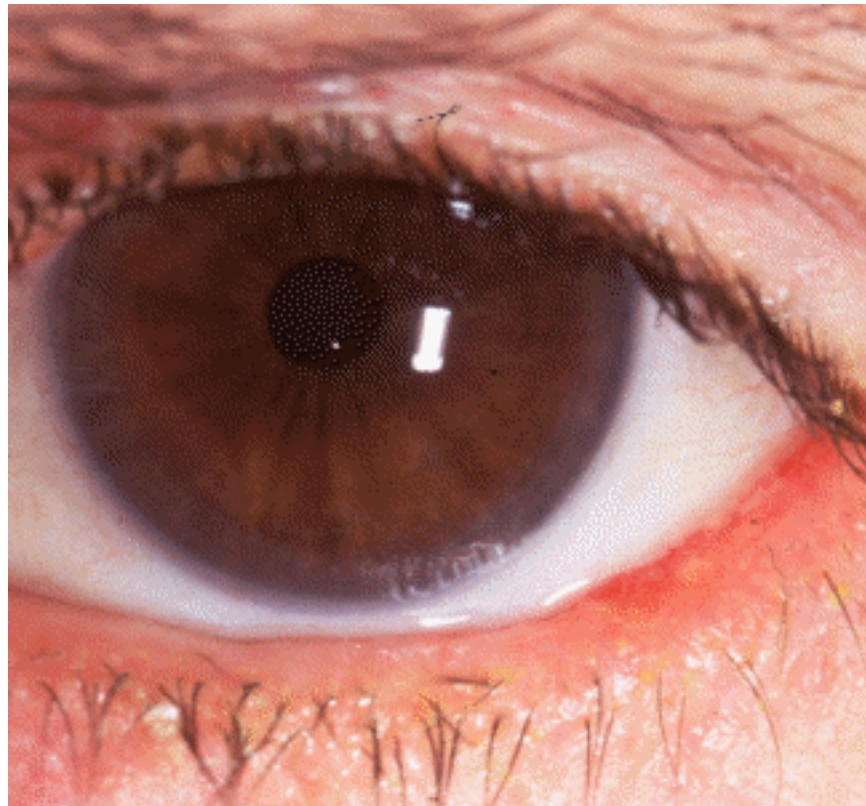
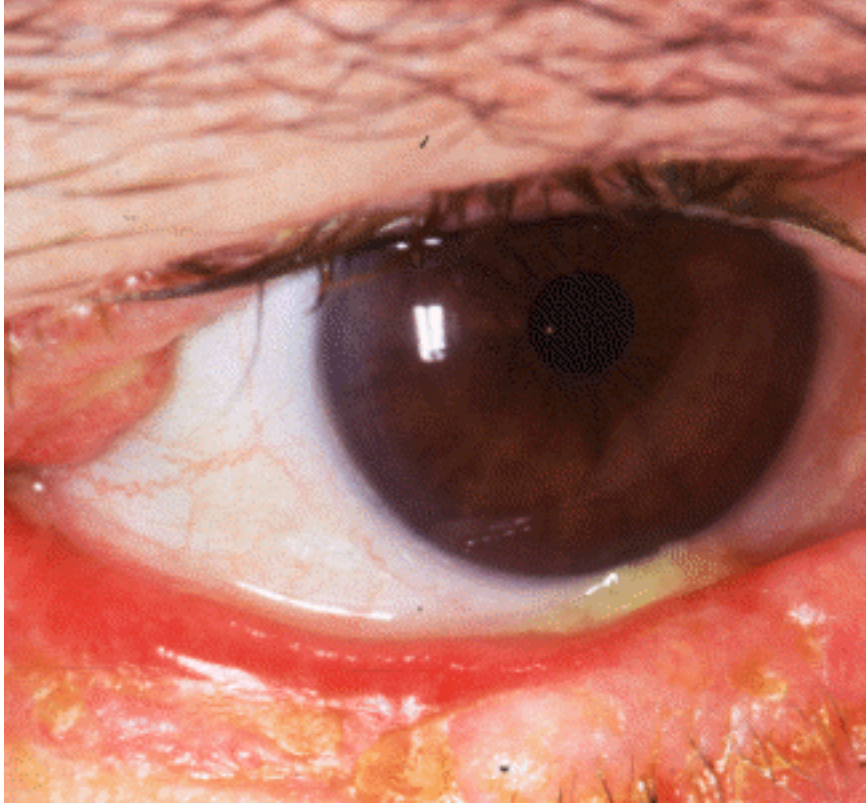


Fig. 2 Marked erythema and scaling especially at the outer lower lid margins. Note hypertrophic lesion, right upper lid

The service reviewed her biopsy slides together with Dr. Frederick Jakobiec . The histopathologic findings was clear to Dr. Jakobiec: 1) hyperkeratosis of the epithelium; 2) thickened basement membrane; 3) basal cell vacuolation; 4) telangiectasia of the substantia propia; 5) lichenoid infiltrates; and, 6) dermal inflammation. (Figures 3,4) The histopathologic diagnosis was discoid lupus erythematosus, hypertrophic or verrucous variant.

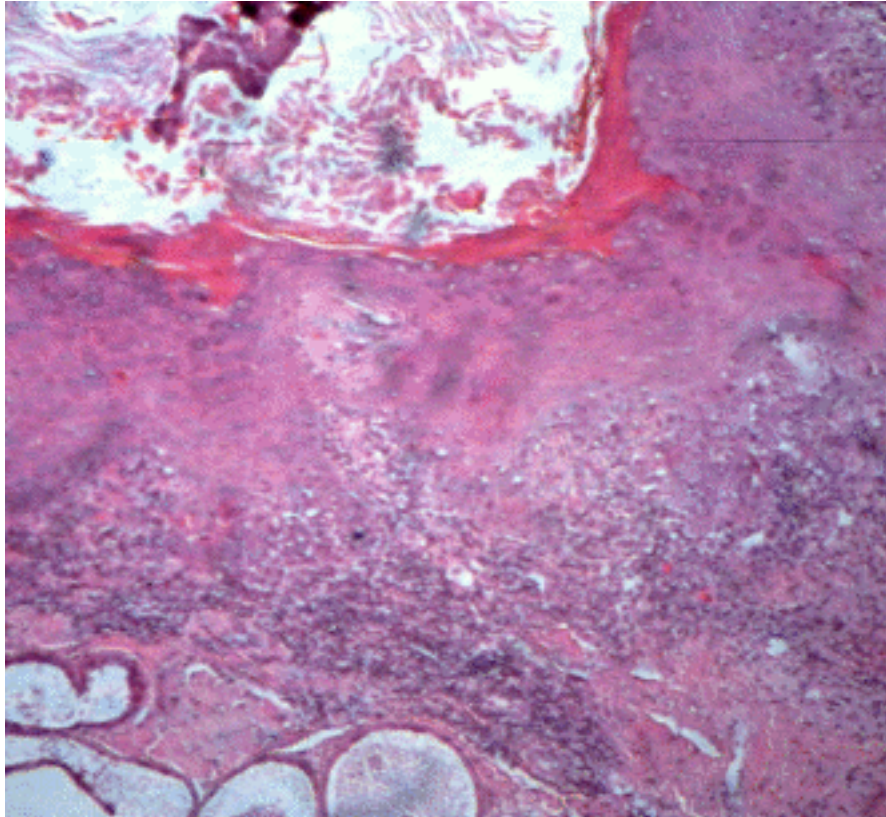


Fig. 3 Hyperkeratosis and dermal inflammation

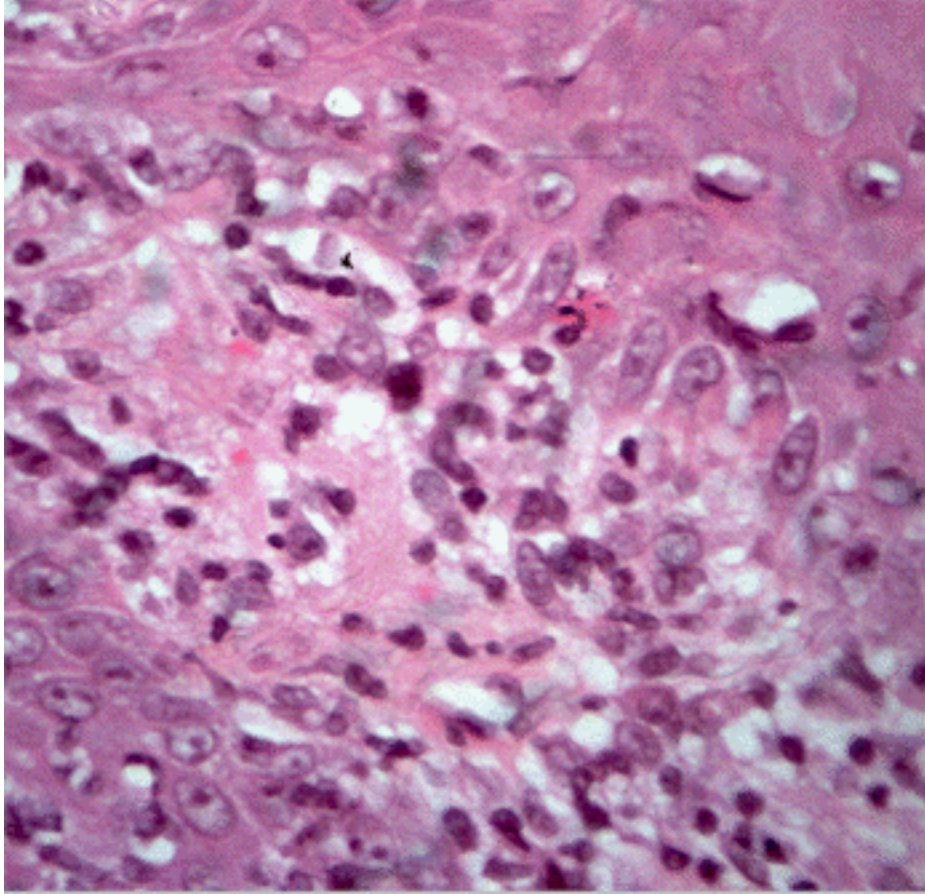


Fig. 4 Basal cell vacuolation and basement membrane thickening

The patient was started on hydroxychloroquine (Plaquenil 200 mg) twice a day per orem. After two weeks, she noted decrease in lid margin redness and irregularity; there was also decrease in pain. The lid lesions gradually regressed. She continued to improve over the next two years (**Figure 5**) No signs of Plaquenil toxicity were revealed by visual field or by dilated funduscopic examination.

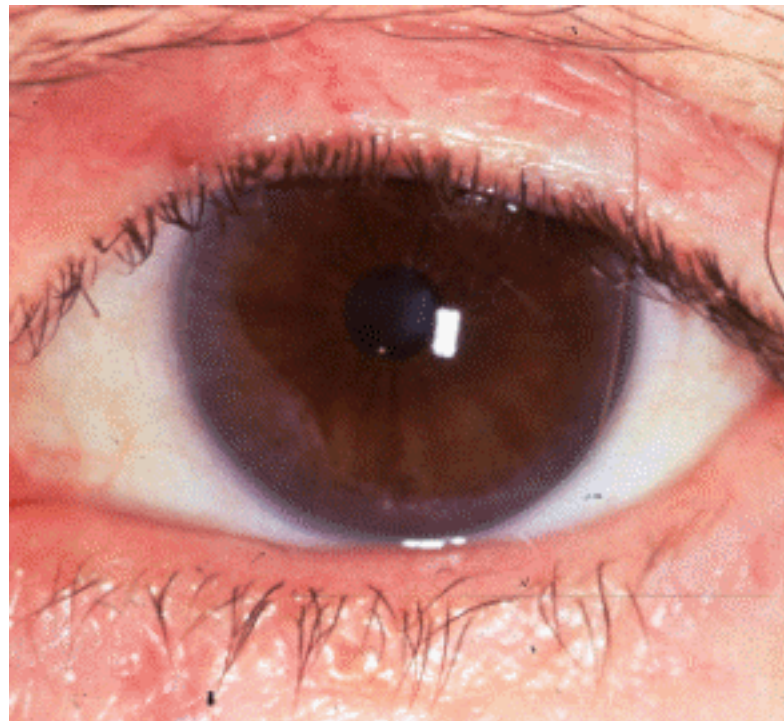
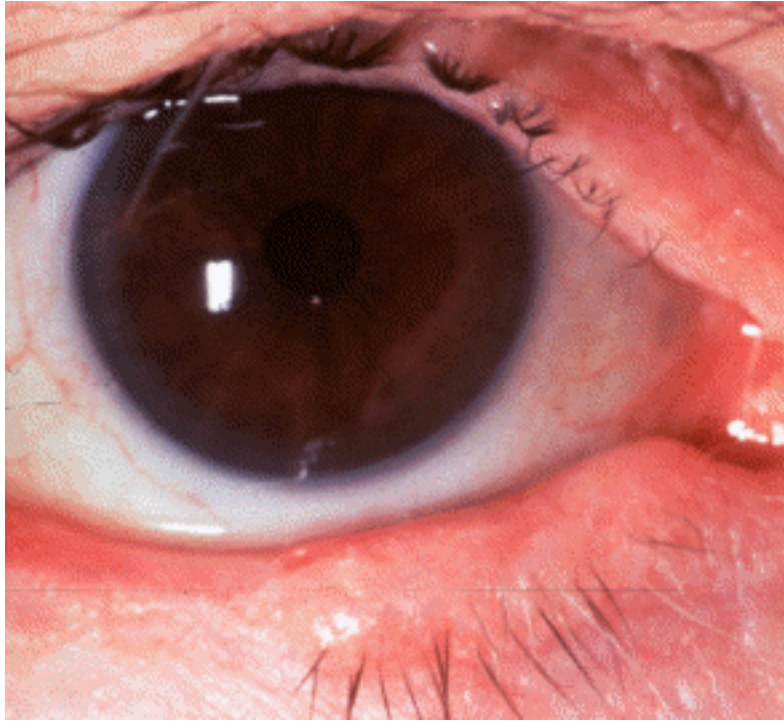


Fig 5. One month after Plaquenil therapy, marked diminution of eyelid redness and thickening.

Discussion:

Discoid lupus erythematosus (DLE) or chronic cutaneous lupus erythematosus (CCLE) is a benign skin disorder characterized by well-defined, raised, erythematous lesions. These margins

of these lesions are irregularly outlined and spread slowly while the central portions undergo scaling, atrophy and scarring. Areas frequently involved include the face, ears, and scalp. The trunk, extremities and mucosal surfaces are occasionally affected [1,2].

Epidemiology

DLE is not an uncommon disease although its actual prevalence is unknown. Females are affected twice as often as males. Most patients are between 25 to 45 years old; however, neonatal and childhood cases have been reported [3]. There is no known racial predilection. While a significant number (15-20%) of systemic lupus erythematosus (SLE) patients manifest DLE lesions; only about 5-10% of patients with DLE go on to develop SLE [4].

Pathogenesis:

The exact etiology of DLE is unknown. A genetic role is suspected on the basis of increased prevalence among first degree relatives, the association with HLA-B7 and -B8 and the development of DLE-like lesions in genetic syndromes. The presence of virus-like structures on electron microscopy and the elevation of certain anti-viral antibody titers suggest an infectious etiology. Skin trauma and ultraviolet light exposure have also been reported to induce or exacerbate lesions. Sex hormones may also play a role. Exacerbation may occur during pregnancy, menstrual or pre-menstrual periods and with intake of oral contraceptives. Finally, drugs such as procainamide, hydralazine, isoniazide, diphenylhydantoin, methyldopa, penicillamine, guanides and lithium may precipitate DLE lesions [2,5,6].

Like SLE, DLE is believed to be an autoimmune process. Unlike SLE, however, DLE patients do not manifest the same serologic abnormalities [7]. A comparison of immunologic characteristics of SLE and DLE is shown in Table 1 [8].

Table 1. Comparison of immunologic features of DLE and SLE

	Discoid Lupus	Systemic Lupus
Systemic manifestations	5-10 % of patients	All patients
Serologic Abnormalities	Few	All patients
ESR	Normal or increased	Increased
Serum immunoglobulins	Normal	Increased IgG, M, A
Serum complement	Normal	Low
ANA	Mostly negative	Mostly positive
Biopsy	Only lesions are positive	Extralesional skin positive
Association with episcleritis	No	Yes

Though both humoral and cellular-mediated pathways have been implicated in the pathogenesis of DLE, cellular-mediated immunity may play a greater role. There are several observations in favor of this. The vast majority of infiltrating cells are T-lymphocytes instead of antibody-producing B lymphocytes. Synkowski and associates have demonstrated that T cells and Ia cells are in abundance in DLE lesions. The number of helper and suppressor T cells has been found to be in equal proportion [7,9,10,11].

More than 90% of these are HLA-DR positive. HLA-DR positive cells were present among epidermal keratinocytes and dermal cells but not among the endothelial cells. These HLA-DR positive cells may function as antigen presenting cells involved in major histocompatibility complex (MHC) restricted cellular cytotoxicity [9,10,11].

The absence of autoantibodies in most patients with DLE suggests that helper T-cells stimulate macrophages and cytotoxic T cells via cytokines. [12] Elimination of helper T-cells in mice models retards or prevents disease [13,14] Immunohistochemical studies done by Tebbe and associates revealed expression of adhesion molecules (ICAM-1) in epidermal keratinocytes, dermal inflammatory cells and endothelial cells. Leucocyte function associated antigen (LFA-1) was present only in the dermis. ICAM-1 and LFA-1 most likely play a role in target/effector recognition between target cells, T cells and endothelial cells. HLA-DR was also present in epidermal keratinocytes and dermal cells indicating a role for major histocompatibility complex restricted cellular cytotoxicity. Interleukin 2 receptor (IL-2R) expression in DLE was found to be weak with less than 1% of cells positive for IL-2R. This suggests non-specific activation of T lymphocytes [9,11]. B7-CD28 costimulatory pathways have also been detected in discoid lupus erythematosus lesions and may be important to propagation of T cell hyperactivity [15]. Among the population of dermal inflammatory cells, most were T helper inducer cells, few were B cells. There was also an increased number of Langerhans cells in peripheral lesions.

De Jong and coworkers demonstrated an increase of keratinocyte proliferation factors, Ki-67 and keratin 16 as well as of differentiation factors, involucrin and filagrin. These may partly account for the hyperkeratosis and follicular plugging of the skin [16].

Clinical Features

The typical cutaneous discoid lesions begin as erythematous patches or papules located at the malar area, nose, face and scalp (80-90% of patients). Later, slowly growing, sharply demarcated but irregularly outlined, bright, erythematous papules and plaques develop with edema and elevation of the skin. This is followed by formation of adherent thick plaques with follicular plugging and scaling. Older lesions demonstrate atrophy with pigmentary changes. These lesions are usually asymptomatic but may sometimes be pruritic. The typical discoid lesions follow a progression of erythema, hyperkeratosis and atrophy with pigmentary changes. The limbs and trunk are less frequently affected [1].

Mucosal involvement is observed in 15-24% of patients. The oral and nasal mucosa may develop asymptomatic plaques with irregular, well-defined, scalloped white margins with radiating peripheral short white striae and telangiectasia giving a honeycomb appearance. The palate, lips, vagina, anus and conjunctiva may also be involved. [17,18]

Several morphologic variants have been reported. Chilblain lupus lesions appear as infiltrated papules and plaques of the hands and feet which are worsened by cold weather [2]. Bullous lesions may also occur rarely.

Hypertrophic DLE is found in only 2% of patients and manifests as raised, indurated, hyperkeratotic, wartlike lesions found mostly in the face, upper extremities, palms and soles. These chronic lesions should be differentiated from hypertrophic psoriasis, lichen planus,

verrucae, keratoacanthomas and squamous cell carcinomas. To the best of our knowledge, there have been no previous reports on hypertrophic discoid lesions of the conjunctiva. [1,19-21]

The profundus lesion manifests as well-defined subcutaneous nodules. In these cases, a panniculitis affects the subdermal fat pads and appears as movable nodules. These lesions are usually found in the arms, buttocks, thighs and breasts. Histopathologically, hyaline necrosis of fat, lymphocytic infiltration and calcification may be seen. These heal leaving depressed scars. [19,21-23]

Ocular involvement is uncommon. Fewer than 50 cases have been reported with only 17 having solely eyelid involvement. Because of lack of familiarity by ophthalmologists, eyelid DLE are usually misdiagnosed with a 2 year average time to diagnosis [24]. While DLE lesions elsewhere have a distinctive appearance, eyelid lesions are more subtle in appearance resembling non-specific blepharoconjunctivitis. The outer third of the inferior lids is a frequent site of involvement [25].

The different ocular manifestations and complications of DLE are listed in Table 2 [1, 21, 24-32].

Table 2: Ocular manifestations of discoid lupus erythematosus

Orbit

Proptosis

Periorbital edema

Eyelids

Chronic blepharitis

Lash loss

Scarring, destruction and disfigurement

Entropion, ectropion

Trichiasis

Pannaliculitis

Pigmentary changes

Conjunctiva

Conjunctivitis

Hypertrophic/verrucous lesion

Scarring

Symblepharon

Cornea

Stromal keratitis

The differentials for DLE are listed in Table 3. Blepharitis is probably the most common differential. DLE lesions are dry, have fine adherent scales, appear purplish-red and are associated with lash loss. Blepharitis lesions are moist, have discharge, are brick red and have matted lashes [1].

Table 3: Differentials for DLE of the eyelids

Infectious

Mycosis

Syphilis

Hordeolum

Trachoma

Immune/Allergic

Psoriasis

Seborrheic dermatitis

Contact/Allergic dermatitis

Polymorphic light eruption

Tumor

Lymphocytic infiltration

Sebaceous cell carcinoma

Diagnosis

Serologic testing is not as helpful in the diagnosis of DLE but screening for SLE should be done to determine presence of potentially life-threatening systemic disease.

Definitive diagnosis is established by biopsy. Findings in DLE are listed in Table 2. Routine histopathologic and immunopathologic studies should be performed. Electron microscopy and may be necessary. The highest yield rate is from conjunctiva where 48% positivity on immunopathology. Skin and lips are only positive 3% of the time [33].

Table 2. Histologic characteristics of discoid lupus erythematosus [21,34,35]

Layer	Finding
Stratum corneum	Hyperkeratosis, parakeratosis Follicular plugging
Epithelium	Stratum malphigii thinning and flattening Decreased rete ridge pattern Hydropic/liquefactive degeneration of basal cells Basal cell necrosis
Basement Membrane	Thickening and reduplication
Stroma	Lymphocytic predominant infiltrates (dermal-epidermal junction, hair follicles) Appendage atrophy Stromal edema, vasodilation, mucin deposition, lymphocyte extravasation Non-specific colloid bodies
Subcutaneous	Extension of inflammatory cells
Others	Pigment incontinence Atrophic Malphigian layer

By electron microscopy, widened intercellular spaces of basal cells; bifurcation and multiplication of the epithelial basement membrane; deposits of ground substance in the subepithelial connective tissue spaces, and tubuloreticular lesions are observed [35].

With direct immunofluorescence, staining with IgG is seen in lesional skin. This may be seen as a well-defined broad immunofluorescent band at the dermo-epidermal junction (lupus band). This band may extend around the hair follicles. A narrow IgM band, prominent cytooid bodies and linear deposits of complement and fibrin bands may also be observed [36,37].

The presence of anti-nuclear antibodies is associated with a higher incidence of arthritis and photosensitivity and more acute lesions. A high level of single stranded DNA antibody is correlated with widespread and greater disease activity [31].

Treatment

1. **Reassurance** that this is a benign disease infrequently associated with systemic lupus.
2. **Sunscreen** to block UV radiation
3. Topical or intralesional **steroids**
4. Adjuvant therapy 20 ug/ml q 3 weeks;

5. Antimalarials:

- Antimalarials are the drug of choice and mainstay of treatment for lupus skin lesions. Hydroxychloroquine sulfate (Plaquenil), chloroquine phosphate (Aralen) or quinacrine hydrochloride have been shown to be useful in the treatment of cutaneous LE. Usually hydroxychloroquine is first tried as this drug has the least toxicity at a dosage of 200 to 400 mg per day. A period of 4 to 8 weeks is required to evaluate response to therapy. Chloroquine (250 mg daily) and quinacrine (100 mg daily) may be tried if unresponsive to hydroxychloroquine. Systemic side effects include: antimalarial retinopathy, pruritus, lichenoid drug eruptions, erythema multiforme, exfoliative dermatitis, cutaneous pigmentation, psychosis, seizures, cytopenias, neuromyopathy. Ocular side effects include retinopathy, extraocular muscle myopathy and corneal deposits. Regular examination including ophthalmoscopy and visual fields every 3 months may detect early toxicity and prevent permanent damage [1,19].

6. Dapsone

- Dapsone, a sulfone drug, has both anti-inflammatory and antimicrobial actions. It is believed to inhibit the alternative pathway of complement activation and polymorphonuclear (PMN) cell migration; it also suppresses production of hydrogen peroxide and hydroxyl radicals by PMN's ; leading to reduced cytotoxicity. Dosage is typically 50 mg/day for adults [38]. Severe idiosyncratic hematologic reactions such as aplastic anemia, agranulocytosis, hemolytic anemia as well as exfoliative dermatitis and Stevens-Johnson syndrome have been reported [39].

6. Methotrexate

- A competitive inhibitor of dihydrofolate reductase, methotrexate interferes with nucleic acid synthesis. Its antiinflammatory and antiproliferative effects are believed to stem from inhibition of DNA synthesis by immunocompetent cells. It has been reported to be effective against DLE refractive to steroid and antimalarial treatment [40].

7. Azathioprine

- Azathioprine is a purine antagonist that inhibits DNA and RNA formation and reduces cell-multiplication and modulates cell-mediated immunity. Callen and associates reported use of azathioprine in 6 patients with resistant cutaneous LE. Four of the six showed improvement. One patient developed pancreatitis and discontinued treatment. Because of potential systemic toxicity, administration of this agent should be monitored by physicians expert in its use [41].

8. Thalidomide

- Stevens, et al reported a case series of 16 patients with DLE refractive to conventional therapy who responded to thalidomide started at 50-100 mg/day with maintenance of 25-50 mg/day. About half of patients went into complete remission and a third more entered partial remission. Improvement was noted in 2-4 weeks with maximum benefit in 16 weeks. Effects of thalidomide on the immune system include decrease in tumor necrosis factor-alpha by monocytes, decreased peripheral CD4: CD8 ratios, enhanced cytokine production by T-helper lymphocytes, inhibition of angiogenesis and decreased levels of serum immunoglobulin. Side effects: somnolence, nausea, constipation, headache, rash, and polyneuropathy. Some patients relapsed on cessation of treatment. The potential for teratogenicity should be kept in mind in treatment of women of child-bearing age [42,43].

EMERGING THERAPIES

Interferon-alpha 2

Interferon-alpha 2 had been reported to be useful in the treatment of refractory DLE. Its mechanism of action is unknown but may be due to down-regulation of lymphokine production. It has been used parenterally and intralesionally. The intralesional dose was reported by Martinez, et al to be 5×10^6 IU twice a week given together with acetaminophen. Repeat biopsies confirmed clinical improvement after three weeks and maintenance treatment every 3 weeks was given. Side effects were mild flu-like symptoms [44].

AntiCD4 antibody, CM-T412

Anti-CD4 antibodies cause reduction of CD4+ counts and decrease in expression of activation molecules on the membranes of T cells or keratinocytes within the DLE lesions. There is a reduction in of inflammatory activity and clinical improvement. Prinz et al reported successful treatment of DLE with anti-CD4 antibodies in 4 patients. The total intravenous dose was 275-475 mg of cM-T412 given as 20-50 mg single doses over 5 to 8 weeks after premedication with metoclopramide 10 mg IV and per orem loperamide 4 mg. There was immediate response in all patients. There was relapse in all patients as early as 10 days after treatment; however, further control was achieved by use of previously ineffective conventional therapies such as topical and systemic steroids or hydroxychloroquine. Side effects included flu-like symptoms. CD4 acts as a coreceptor for the T-cell antigen receptor (TCR) by direct interaction with MHC class II molecules and participates in signal transduction of the TCR/CD3 complex by an intracellularly associated tyrosine kinase. TCR-independent engagement of CD4 molecules by anti-CD4 molecules leads to T cell inactivation resulting in immunotolerance. CD4 antibodies have also been shown to induce CD4+ T lymphocyte apoptosis in murine models, which may be Fas dependent [45].

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Discoid Lupus Erythematosus of the Eyelids Review Questions

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1. What are some clinical features of DLE?
 - a. erythema
 - b. hyperpigmentation
 - c. scaling
 - d. scarring
 - e. all of the above

2. Which population is at most risk for developing DLE?
 - a. children
 - b. Asians
 - c. retirees

d. Caucasians

e. females

3. Approximate risk of developing SLE in patients with DLE

a. 0 %

b. 15 %

c. 33 %

d. 50 %

e. 100 %

4. Risk factors for developing DLE

a. sunburn

b. seizure disorder

c. oral contraceptive use

d. menstrual period

e. all of the above

5. Differences between DLE and SLE, EXCEPT:

a. Most SLE patients produce anti-nuclear antibodies, normal in DLE

b. Most SLE patients have elevated serum immunoglobulins, normal in DLE

c. Serum complement is low in SLE, normal in DLE

d. ESR is elevated in SLE, normal in DLE

e. Clinical discoid lesions exhibit pathologic changes in SLE, are normal in DLE

6. Evidences for cell-mediated immunity in DLE, EXCEPT:

- a. 90% of lymphocytes are T helper/inducer cells in discoid lesions
- b. presence of B7-CD28 constimulatory molecules in discoid lesions
- c. absence of autoantibodies
- d. presence of adhesion molecules in discoid lesions
- e. none of the above

7. Manifestations of ocular DLE, EXCEPT:

- a. proptosis
- b. chronic blepharitis
- c. entropion
- d. keratitis
- e. none of the above

8. Histopathologic findings in DLE, EXCEPT:

- a. hyperkeratosis
- b. basal cell necrosis
- c. basement membrane thickening
- d. appendage hypertrophy
- e. none of the above

9. Characteristic immunofluorescent finding in DLE:

- a. reduplication of basement membrane
- b. diffuse staining of epithelium
- c. diffuse staining of stroma
- d. lumpy-bump fluorescence of basement membrane

e. none of the above

10. Mainstay of treatment for DLE:

a. systemic immunosuppressives

b. topical steroids

c. thalidomide

d. antimalarials

e. systemic steroids

Answers: 1E 2E 3B 4E 5E 6E 7E 8D 9D 10D