Current Practice

DISEASES OF THE SKIN

Management of Cutaneous Lupus Erythematosus

STEPHEN GOLD,* M.D., F.R.C.P.

Discoid lupus erythematosus is a relatively uncommon chronic skin condition which affects females twice as frequently as males, and the majority of patients are in their third or fourth decades at the time of onset.

The exact relationship of the discoid or purely cutaneous form with the systemic disease (S.L.E.) is still undecided. There is much evidence that they represent different ends of one spectrum. Support for this is the occasional (5%) change from the cutaneous form to the systemic, and more particularly the existence of a great variety of intermediate patterns which bridge the gap between the two extremes. Furthermore, the more critically forms which are seemingly cutaneous are screened with modern sophisticated techniques the more frequently unsuspected haematological and immunological disturbances are revealed. It should be mentioned, however, that Burch and Rowell^{1,2} have produced ingenious, if complex, data which they feel are evidence of a genetic difference between the cutaneous and systemic forms. It is known that close relatives of affected persons are more likely to reveal humoral and immunological evidence of L.E. than is the general population.

Clinical Features

Patches of fixed erythema appear, especially on the exposed skin. They are well defined and most commonly seen on the face; they may be single or multiple. Symmetrical lesions on the cheeks, spreading across the bridge of the nose, often produce the characteristic butterfly or batwing pattern. Patches may also appear on the forehead, chin, and ears, and on the backs of the hands, and—especially in women—on the upper chest where it is exposed to sunlight. Such patches may enlarge centrifugally and fuse with others to result in polycyclic and bizarre shapes.

In the early stages the areas are somewhat raised, more as a result of oedema than of cellular infiltration, and they may be circular or quite irregular in shape. Certain diagnosis is not possible until other confirmatory signs develop. Later the erythematous patches may become scaly, and scaling is an important sign of epidermal involvement; it is often most marked at the edge of the lesion, especially in the more chronic forms. When the scales are removed they are found to be adherent, because spinelike keratinous processes anchor them to the underlying dilated pilosebaceous follicles. Rarely, keratinous follicular plugs may be seen actually extruding from the skin surface as threadlike projections.

If such patches persist, then scarring will follow gradually. The smooth, shiny, dry, and depressed surface of the atrophic skin will often reveal telangiectasia. This is the fourth cardinal

sign. Whether it is the face, scalp, fingers, or toes which are affected, the same distinctive features of erythema, scaling, atrophy, and telangiectasia can result.

A pigmentary change may also occur. It is not uncommon to see suppression of pigment in the centre of patches, and such secondary leucoderma, if occurring in a negroid subject, can cause great cosmetic distress. In some cases peripheral hyperpigmentation may produce a darker halo zone to the patches.

The course of discoid L.E. varies. Some patients have persistent trouble over a great many years, while in others it is more labile and evanescent. There are several recognizably distinct clinical types:

Chronic Persistent Forms.—In this type the condition produces the grossest degree of scarring and disfigurement. It is peculiarly resistant to treatment, and perhaps a third of patients fall into this category. Hyperkeratosis and pigmentary disturbances are most likely to be seen.

Chilblain Lupus (Hutchinson's).—The areas predominantly affected are those parts subject to chilblains, particularly the tips of the fingers and toes. Changes may also be seen on the ears, nose, and possibly cheeks. Interference with the nail matrix may result, and nail growth may therefore be disturbed; a variety of changes from complete shedding to various distortions of the normal plate can be seen.

Labile Form.—The erythematous patches are more evanescent, and scaling is not so marked. The depth of dermal involvement is less than in the chronic persistent type, as is the degree of scarring. Such cases are more rewarding to treat, for complete recovery is the rule.

The histological features of the above types will show various degrees of hyperkeratosis with keratinous plugging of the follicles. The older the lesion the more gross the changes, and it should be stressed that biopsy of early lesions is not profitable. The epidermis is frequently thinned, and the dermo-epidermal junction blurred with intracellular oedema and cellular vacuolation, the so-called liquefaction-degeneration. The most uniform dermal change is a patchy lymphocytic infiltrate focused on the neurovascular bundles and around the skin appendages. There is capillary dilatation, but otherwise gross vascular changes are unusual.

Lupus Erythematosus Profundus (Kaposi-Irgang).—This is a rare variety in which epidermal changes are usually absent. Histological diagnosis is difficult, but often there is a necrobiotic zone deep in the dermis surrounded by a non-specific inflammatory infiltrate. The patches are red, firm, persistent, and infiltrated, whereas surface scaling, atrophy, and telangiectasia are absent; deep scarring may well result, but ulceration rarely occurs. The coexistence of more typical lesions is reassuring supporting evidence for this difficult diagnosis.

Lupus Tumidus.—A further variant of L.E. most likely to be seen on the ear lobes are livid, plum-coloured, thickly

* Physician, Skin Department, St. George's Hospital, London S.W.1.

infiltrated plaques clinically suggesting a skin lymphoma. Compact foci of lymphocytes, reminiscent of lymphoid follicles, are a distinctive histological feature.

Buccal L.E.—In some cases the lips may be affected. The vermilion may show cracking, dryness, and fissuring, sometimes becoming completely covered with thick adherent scales. The lower lip is more commonly affected than the upper (compare actinic cheilitis). Occasionally warty excrescences may develop on top of long-standing cheilitis.

Lesions inside the mouth may appear as erythema, submucous haemorrhages, and erosions. Blistering is not unusual, though white plaques may occasionally occur, especially in long-standing lesions, making clinical differentiation from lichen planus and leukoplakia important. Long-standing L.E. of the buccal mucosa will, as with lesions of the ordinary skin, be followed by scarring.

L.E. of the Scalp.—On the scalp patches usually appear as well-defined atrophic bald areas. For this reason it may easily be confused with other atrophic alopecias, notably lichen planus. Generally there will be some patchy erythema with scaling; the white areas will be shiny and entirely hairless. It is easy to observe follicular plugging and perifollicular hyperkeratosis. Secondary infection of surface erosions may be a feature during the active phases. Lesions on the scalp are notoriously resistant to treatment, but can usually be camouflaged without much difficulty.

Complications

The development of squamous carcinoma (epithelioma) in patches of long-standing scarring L.E. is occasionally seen. This is no more common than in scarring lupus vulgaris, and is probably more a sequel to the atrophy and chronic inflammation than to anything specific about the process itself.

Management

One of the most striking clinical facts is that the cutaneous areas affected can be explained on a basis of solar damage, and many patients are aware of the worsening of their condition after sunburn. Occasionally the initial patch appears after a patient has been sunburnt. The distribution of the lesions on the nose, cheeks, dorsal surface of the hands, and the V of the female neck are all evidence of the effect that sun may have on this condition. However, it is surprising to find there is little evidence of any increase or alteration of response when such patients are carefully tested to assess photosensitivity. When one considers that neither buccal lesions nor scalp lesions can be explained in this way, it seems that it is trauma to the skin which really triggers off the changes in a subject who is susceptible and ready to react. There is in fact almost uniform incidence of the conditions throughout the world, in spite of the great difference of available sunlight—it is, for instance, seen in negroid subjects, in whom a built-in sunscreen is available. L.E. can occur in abraded skin, after burns, vaccination, and after various types of trauma. It is therefore important to minimize skin damage, and careful advice should be given to patients concerning sun protection. It is customary to warn patients to take their holidays in less-sunny climates, and it is wise to suggest protective clothing, such as wide-brimmed hats, long sleeves, and the use of a parasol—advice that is rarely popular. There are a variety of sun-screens available which can be useful, the most popular being either 15% para-aminobenzoic acid in ung. emulsificans aquosum or a proprietary preparation Uvistat 2211 (Ward, Blenkinsop), which contains a variety of benzophenones. These preparations should be applied to the skin before exposure. They form a protection for four to five hours.

(a) Systemic Treatment

1. **Corticosteroids.**—The prescription of corticosteroids should virtually be confined to the frankly systemic forms, though for acute exacerbations of the discoid type they occasionally may be justified. Small doses of prednisone daily (10/15 mg.), or twice this dose given on alternate days, are unlikely to provoke serious side-effects even over long periods, provided the usual precautions are observed. It is probably wise to leave this type of treatment to experts.

2. **Antimalarials.**—For no explicable reason it has been observed that various antimalarial drugs have a suppressive effect on the activity of this disease. Quinine, the first to be introduced, was frequently prescribed between the wars. Around 1951 to 1952 mepacrine was observed to have a beneficial effect on patients with the discoid form. This drug has to be given over prolonged periods, and as well as causing a yellow discoloration of the skin it has the tendency to provoke other undesirable side-effects, notably blood dyscrasias, gastrointestinal upsets, and various skin reactions. It was replaced by chloroquine, a related drug which does not have the staining qualities of mepacrine and seems altogether better tolerated.

Chloroquine (Nivaquine; Avoclor) in doses of 200 to 600 mg. daily has an appreciable effect on the course of chronic discoid L.E. in the majority of patients. However, once the drug is stopped it is apparent that the effect has been suppressive rather than curative; relapse generally occurs within a few weeks of withdrawal. For this reason chloroquine has been prescribed over long periods. It is interesting to note that chloroquine has also been used for the treatment of rheumatoid arthritis, and patients with S.L.E. will often report an improvement in a variety of symptoms after its use—not only in the arthralgias but in less well defined symptoms such as malaise and loss of energy.

Side-effects.—Various side-effects have been reported after prolonged use of chloroquine. These include the occasional haematological disturbance, especially thrombocytopenia, rashes of the skin, often closely resembling lichen planus, as well as generalized pruritus. In addition, depigmentation or discoloration of the hair resulting in a white or yellow forelock, or patchy discoloration of the scalp, brows, or lashes, may produce a striking appearance.

It is, however, the ocular complications which should take pride of place in considering whether or not it is justifiable to prescribe a drug of necessity to be given over prolonged periods.

The development of corneal opacities is fairly common. It is insidious in its progress and usually bilateral, resulting in various visual disturbances, notably blurring of vision, the development of haloes round lights, and occasionally double vision. Its detection necessitates skilled examination with a slit-lamp microscope. Such toxic changes in the cornea are reversible, and improvement occurs gradually after the drug is stopped.

A more disturbing side-effect is the possible development of a retinopathy in which oedematous changes occur in the fundus, with vascular narrowing and deposition of pigment. Macular involvement is distinctive. This process may lead to progressive blindness, and its progression is not immediately stopped by withholding the drug. Furthermore, damage may start before any symptoms of scotoma or field defects are noticed. Chloroquine is in fact retained in the body for many months, and this should partly account for the sinister progressive deterioration of the retinal disease. The earliest signs of developing retinopathy can be detected by plotting visual fields, for they will tend to be reduced. Another useful test in expert hands is provided by the electro-oculogram. This is based on the measurement of potential difference between the cornea and the fundus. If the figures obtained with those in bright light are compared with those in darkness a marked

variation is apparent between the two readings once retinopathy is starting.

It would therefore be permissible to prescribe chloroquine for only short courses at the height of summer, or possibly in the spring, when sun damage is likely to be maximal. Ocular disturbances seem to be related to the total dose, as well as to the duration of administration. A three months' course with a daily dose of not more than 400 mg. is unlikely to cause eye complications, especially if it is followed by a prolonged time off the drug to allow total elimination. If such a course is adopted supervision by an ophthalmologist who can assess the visual fields and make the necessary slit-lamp examination from time to time is advisable.

3. *Bed-rest*.—For the few patients with widespread lesions, which seem to be resistant to the usual forms of treatment, a period of bed-rest for four to six weeks is often rewarding.

(b) Local Procedures

Treatment therefore depends largely on local measures, and with the introduction of the modern fluorinated cortisone-type compounds there has been a distinct advance in this respect. Fluocinolone (Synalar), betamethasone valerate (Betnovate), triamcinolone acetonide (Adcortyl, Ledercort) are such compounds (and there are a number of others in this highly competitive commercial field), which if applied frequently to the inflamed areas will generally cause improvement. If simple local inunction of such a preparation is advised it is important that frequency of application is stressed; it is more effective to apply a little four or six times in 24 hours than merely to apply it twice daily. The choice of vehicle, whether it be in a greasy base, in a vanishing-cream, or in a lotion, depends partly on which area of the skin it is to be applied to and partly on which is most acceptable to the patient. Trial and error will decide which choice is the more agreeable and effective.

It may be possible to enhance penetration by occluding the treated area with a film of polyethylene sheeting. Having applied the steroid, a sheet of impermeable polyethylene fabric is kept in place with adhesive strapping, so that the underlying skin is sealed from the air and becomes warm and moistened with sweat. It is important to seal the polyethylene down tightly, and if adhesive strapping is used some ingenuity is required so as to ring the changes on the areas that are subject to repeated adhesive. For scalp lesions a polyethylene sheet can be kept close to the surface overnight by means of a well-fitting hair-net or even a bathing-cap. Ingenious plastic devices have been devised for use on difficult contours, such as the nose and ears, by using acrylic moulds which exactly conform to the site; they may be held in place (on top of the polyethylene sheet) by means of an elastic band round the back of the head. By pretreating the skin with hot wet flannels, applied for 10 minutes before rubbing in the steroid, similar effects may also be obtained.

As a result of these new techniques efficacy of surface applications has been improved, and it is not often necessary to resort to intracutaneous injections of the steroid preparations. Such injections, which were first undertaken with an ordinary intradermal needle and later by dermojet, can have the disadvantage of producing depressions of the skin, even scarring.

They may be useful, however, in the profundus and tumidus varieties.

Buccal erosions may be helped by simple bland mouth-washes and the sucking of hydrocortisone hemisuccinate lozenges (Corlan). Occasionally a special ointment (Orabase) will adhere to the buccal surface, in which case triamcinolone (Adcortyl in Orabase) may be useful. The lips can be treated by ordinary ointments and creams.

(c) Physical Treatments

If warty excrescences develop and persist for long they are best destroyed, bearing in mind that on rare occasions squamous carcinoma is a sequel to the scarring variety of L.E. Such lesions are most likely to occur on the lower lip or on the helix of the ear.

Carbon dioxide slush (dry ice moistened with a few drops of acetone) was a favourite local procedure in the "presteroid" era, and it certainly seemed useful in speeding on the natural development of scarring; it is rarely used nowadays.

Treatment by x-irradiation or ultraviolet rays is not used in the treatment of L.E.

Plastic surgery is best deferred until all activity has ceased. This should be assessed by the presence of erythema and scaling, extension of existing areas, and development of new ones. A gap of several years between such "dying out" and contemplation of any plastic surgical procedure is essential. Bearing in mind that various forms of trauma are likely to provoke the process into renewed activity, it is easy to understand how ill-judged surgery can be followed by disastrous recurrence.

(d) Camouflage

Covering the ugly patches is not the least important aspect of improving a patient's morale. A thick cosmetic cream can be used on top of an area already treated with steroid, and the cream may also serve as a mechanical screen against the sun's rays.

A variety of cover creams can be bought quite cheaply (Boots), and a more expert service is available (Covermark—Denver Laboratories; Keromask—Innox), where patients are actually taught how best to apply the appropriate masking agents. Preparations are also available for use on dark negroid skin, and should certainly be offered, especially when the secondary leucoderma presents such an unusual appearance in these patients.

REFERENCES

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