

Cutaneous Lupus Erythematosus (CLE)

What is systemic lupus erythematosus (SLE)?

SLE is known in Latin as the wolf. Similarly, the Chinese associate this condition with the aggressiveness of the wolf. It is a systemic disease where all the body parts and organs can be affected. This autoimmune disease is a chronic long standing condition with a relapsing and remitting course i.e. a period of active disease followed by quiescent period.

SLE is a disease seen mainly in women. The ratio of women to men having SLE is 9 to 1. It is a disease mainly seen in the reproductive age group. In Malaysia, SLE mainly affects the Chinese. The spectrum of this disease varies from mild to severe. Those with severe disease usually succumb to their illness if treatment is not instituted urgently. Most patients succumbed to infection and severe kidney disease.

Identifying CLE is important in the diagnosis of SLE as 20% of patients with SLE initially presents with skin manifestations. In fact 85% of patients have skin diseases during the course of their disease.

In active disease, SLE does not only affect the skin but also presents with other organs involvement. Kidney involvement leads to protein and blood in the urine that can progress to kidney failure. Involvement of the brain leads to seizures, personality changes and psychosis. Lungs and heart diseases leads to difficulty in breathing. Bone marrow disease of SLE leads low haemoglobin, white blood cells and platelets causing easy bruising and infections.

To diagnose SLE, doctors rely of good clinical examination and blood investigations. In addition investigations to look for abnormalities of the organs e.g. full blood count, liver and renal function tests, the two most important blood tests are the antinuclear antibody (ANA) and the double stranded DNA (dsDNA). If both these tests are positive together with a typical clinical and laboratory investigations, a diagnosis of SLE is made.

What is cutaneous lupus erythematosus (CLE)?

Cutaneous lupus erythematosus (CLE) is the skin manifestation of lupus erythematosus.

Lupus erythematosus is due to an overactive immune system attacking its own bodily protein or clinically called autoimmune disease. Cutaneous lupus erythematosus can be divided into three clinical types namely acute cutaneous lupus erythematosus (ACLE), subacute lupus erythematosus (SCLE) and the chronic cutaneous lupus erythematosus (CCLE). In Malaysia, the author have noted that among patients with cutaneous lupus erythematosus attending the Dermatology Clinic in Sarawak General Hospital, 47.5% had ACLE, 12.7% had SCLE and 39.8% had CCLE.

What are the skin manifestations of acute cutaneous lupus erythematosus (ACLE)?

Patients with ACLE commonly presents with a butterfly rash. Butterfly rash or malar rash is an erythematous or red rash on both cheeks, interconnected by the redness over the bridge of the nose.

Patients with ACLE can also present with a photosensitive rash. Photosensitive rash is redness seen on the skin exposed to the sun, namely the face sparing the hairline on the forehead (covered by the hair, hence no sun exposure), under the eyes (covered by the eyes), under the nose (covered by the nose), under the mouth (covered by the lower lips) and also under the chin (covered by the chin); on the exposed part of the neck and the chest (typically in a V fashion) and also on the exposed parts of the arms and legs.

Patients with ACLE also frequently presents with multiple painless mouth ulcers. These ulcers are usually seen on the upper part of the mouth or hard palate. All the clinical features of ACLE points towards a diagnosis of SLE. In the author's research, all patients with ACLE have SLE.



Figure 1: Malar rash – “Butterfly rash”

What are the skin features of subacute cutaneous lupus erythematosus (SCLE)?

Patients with SCLE present with two type of skin diseases, i.e. the psoriasiform type and annular type.

The psoriasiform type is more commonly seen than the annular type in Malaysia. The psoriasiform type have skin lesions resembling psoriasis. The skin lesions are raised, red, scaly and looks like map on the body. It is almost always confused with psoriasis.

Other psoriasiform disorders like pityriasis rubra pilaris can mimics this condition. The annular type have skin lesions that are round or circular in nature. These skin lesions are red and scaly, resembling skin lesions of ring-worm, a fungal infection of the skin clinically known as tinea. Other annular skin lesions like discoid eczema can also be confused with this condition.

Patients with SCLE is usually diagnosed by dermatologist via a proper clinical examination and skin biopsy. Skin biopsy entails cutting a small piece of the diseased skin, typically 4 to 6 mm, for histopathological examination under the microscope. Blood investigations usually show positive antibody towards SS-A (anti-Ro antibody) and positive ANA and dsDNA. In the author's research, 20% of patients with SCLE had positive anti-Ro antibody and 53% had positive ANA and dsDNA.



Figure 2: Psoriasiform type SCLE

What is the association between SCLE and SLE?

Approximately 50% of patients with SCLE progress to develop SLE. However, the disease activity in these patients might not be as severe as those with ACLE with SLE.

What are the skin signs of chronic cutaneous lupus erythematosus (CCLE)?

CCLE has most disfiguring skin lesions as the skin lesions heal with scarring. There are multiple types of skin lesions seen in CCLE. The most common type is a skin lesion called discoid lupus erythematosus (DLE).

DLE is a scarring skin lesion seen mainly on the scalp, face, ears, neck and arms. The skin lesions start as small well demarcated red, purplish scaly lesions that expand to form a round coin shaped lesions. The round lesion has a scarred pale centre with multiple stuck on scales and brownish outer ring. The scales are due to blocked hair follicles.

If the skin lesions are found on the scalp, these areas will be devoid of hair causing bald patches. This is because of the scarring caused by the skin lesions. The bald patches are irreversible as the scarring damages the hair follicles. The most characteristic area to find DLE is the ear. This ear sign is the hallmark to allow diagnosis of DLE.

Diagnosis of CCLE is by a careful clinical examination and also by taking a skin biopsy for analysis. Blood investigations in patients with CCLE are almost always normal.



Figure 3: DLE – scarring patches over sun exposed area



Figure 4: DLE over scalp – scarring alopecia

What is the association of DLE with SLE?

Patients who first present with skin lesions of CLE rarely progress to SLE. However, among patients with SLE, it is estimated that 20% have skin lesions of CLE.

What is the treatment for cutaneous lupus erythematosus (CLE)?

Treatment depends on whether the patients have skin disease alone or with combination with systemic organs involvement (SLE).

For patients with only skin involvement, the aim of the treatment is to relieve the skin disease and prevent further cutaneous complications. Further workup to look for underlying systemic organs involvement is essential.

The principle of treatment involve sun protection, topical or oral corticosteroids and oral antimalarials with cessation of smoking.

Sun protection is important for patients presenting with CLE. Sun exposure with worsen CLE. It is vital for these patients to use good sunblock and to wear sun protective clothing. They should also avoid mid-afternoon sun.

Topical corticosteroids are helpful to relieve CLE lesions. The strength and potency of the steroids should be used correctly as directed by the dermatologists. Those with more severe skin disease might warrant use of oral steroids. The risks and benefits of each treatment must be discussed in length before commencement of such treatment.

Oral antimalarials especially hydroxychloroquine is one of the mainstay of treatment for CLE. However, long term use of this medication can affect the eyes. Thus consultation with ophthalmologist is vital before the start of treatment and yearly thereafter. For greatest efficacy of this medication, smoking cessation is essential.

For patients with systemic involvement of SLE, treatment will be more focused on the internal organs involved. A multidisciplinary approach involving rheumatologist, neurologist, dermatologist and other specialists are important in treating the patients. Antimalarials and immunosuppressive treatment are the mainstay of treatment.

Immunosuppressive treatment involve use of medications like corticosteroids, azathioprine, mycophenolate mofetil, methotrexate and cyclosporine. These medications suppress the hyperactive immune system.

Patients and families should also be given psychosocial support. These support can be given by the healthcare professionals and also the patient support group. Patients should also be advised on pregnancy and contraception. Pregnancy can worsen SLE and the medications used to treat SLE can cause problems to the unborn baby. Counselling and consultation with obstetrician and gynaecologist is important.

What skin diseases can mimic cutaneous lupus erythematosus (CLE)?

Skin lesions of cutaneous lupus erythematosus (CLE) is often confused with other skin diseases. A high index of suspicion among treating doctors are essential for early pick up of the condition.

Butterfly rash of ACLE can be confused with other skin conditions like seborrheic dermatitis, rosacea and allergic contact dermatitis.

Skin rash of SCLE, annular type can be mistaken for skin diseases like tinea corporis and nummular eczema. Whereas the psoriasiform type can be mistaken for dermatoses like psoriasis, seborrheic dermatitis, pityriasis rubra pilaris and mycoses fungoides.

Discoid lupus erythematosus can be confused with skin conditions like sarcoidosis, squamous cell cancer and actinic keratosis.

Thus, it is important for patients with these skin lesions to seek proper dermatological assessment to get a proper diagnosis and early treatment. Early treatment in lupus erythematosus allows for better outcome in terms of skin and systemic involvement.

Who should I see for cutaneous lupus erythematosus (CLE)?

Patients suspected of having CLE must consult a qualified dermatologist for proper diagnosis and treatment. The names and place of practice of qualified dermatologists in Malaysia can be found on the Malaysian Association of Dermatologists (Persatuan Dermatology Malaysia) website – www.dermatology.org.my

Prepared by,

Associate Professor Dr Felix Yap Boon Bin
Faculty of Medicine and Health Sciences
Department of Medicine
University Tunku Abdul Rahman

Member of Dermatological Society of Malaysia

Consultant Dermatologist
Subang Jaya Medical Centre (SJMC)



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