



Autoimmune Resource and Research Centre

Information Sheet

Cutaneous Lupus Erythematosus

Lupus is an autoimmune disease, which affects multiple organs and systems in the body. An individual's own immune system attacks various cells causing a wide variety of signs and symptoms. With regards to the skin, there are lupus-specific skin lesions and non-specific skin lesions. Cutaneous lupus may be categorized into three main entities: chronic cutaneous lupus (CCLE), subacute cutaneous lupus (SCLE) and acute cutaneous lupus (ACLE). It is important to note that lupus may be seen as a spectrum with CCLE on one end and systemic lupus (affecting other organs and systems) on the other end. Therefore, it is critical to see a dermatologist to properly evaluate the extent of skin involvement versus systemic involvement, as they do not always directly correlate. Cutaneous lupus most often affects women ages 20-50 and is often induced by sunlight. A wide variety of rashes may be seen and are described below.

The most common form of CCLE is discoid lupus (DLE), named for its coin-like shape. These lesions are categorized by thickened, red scaly patches that often appear on the cheeks, nose and ears. These lesions may also generalize to the v of the neck, upper back and dorsum of hands. They are often not itchy or painful. Once the lesions resolve, they may leave dark or light pigmentation as well as atrophy (thinning of the skin). If lesions are in the scalp or involve the hair follicles, areas of hair loss may develop which could be permanent if the hair follicle is completely destroyed. It is important to note that should these lesions affect the inside of the mouth or lips they may cause ulcers and carry a risk of future squamous cell carcinoma. Long standing lesions may also predispose to future skin cancer. Individuals with DLE on the head and neck do not usually develop systemic disease. Variants of DLE include hypertrophic LE, which may look wart-like and very thick as well as palmoplantar LE (occurring on palms and soles). DLE may also affect fatty tissue below the skin resulting in firm deep nodules called lupus profundus or lupus panniculitis. Once, these lesions resolve they may leave indented scars called lipodystrophy due to destruction of the fat cells.

SCLE is often characterized by two forms including papulosquamous lesions and annular lesions. Papulosquamous lesions often appear as red scaly patches that look psoriasiform. Annular lesions are ring-shaped with a small amount of scale on the edge of the lesions. These lesions do not itch and often appear on the chest as well as the upper back and neck, however, may also be seen on the face and arms. SCLE is not often associated with significant systemic disease however it is always important to be evaluated by your physician. Furthermore, it is not uncommon for patients with SCLE to have associated joint disease.

ACLE is often seen in patients with active systemic disease. Flat red patches on the cheeks and nose called a butterfly rash that looks quite like sunburn characterize the most common form of ACLE. Individuals may also present with generalized flat red patches on arms, legs and trunk. These lesions are sensitive to the sun (photosensitive to both sun rays and tanning rays) and therefore commonly appear on sun-exposed areas. While these lesions do not often result in scarring, they may leave dark or light pigment changes. Other lesions associated with ACLE include oral ulcers, hives, and temporary hair loss, which are replaced by new hair once the disease flare is treated. Systemic lupus erythematosus (SLE) can also affect the

kidneys heart, lungs, liver, brain, joints, blood cells and blood vessels. Damage to the blood vessels results in small red-purple lesions on the lower legs called vasculitis.

Neonatal lupus syndrome may occur when an infant is born to a mother who has autoantibodies in her blood during the pregnancy. The baby may develop skin lesions, which usually resolve by six months. There is a small risk of having a baby with neonatal lupus or congenital heart damage in mothers with SCLL lesions. Therefore, it is critical to discuss this possibility with your physician.

It is essential to be seen by a physician to properly evaluate and individually assess each patient with a complete history, physical examination and laboratory evaluation. Patients with cutaneous LE may have no lab abnormalities, especially if lesions are limited to the discoid type. However, anaemia or a decrease in an individual's white blood cells may exist in addition to elevated antibodies such as ANA (anti-nuclear antibodies). It is also essential to evaluate other organs, which may be affected such as kidneys, heart, liver, lungs and brain. Regarding individual skin lesions, a skin biopsy is helpful in making the diagnosis in addition to direct immunofluorescence tests, which reveal antibody deposition within the skin.

Many different treatments exist for the different forms of cutaneous lupus. Sunscreens are vital in protecting patients from UVA and UVB rays which provoke skin lesions. Apply broad-spectrum sunscreen with at least SPF 30 every 2 hours. Avoid excess sun exposure by wearing sunscreen, wide-brim hats, avoid sunlight during peak hours of UV exposure (10am - 2pm) and wear tightly woven clothing. CLE (discoid lesions) may be treated with corticosteroid creams or ointments in addition to corticosteroid injections. ACLE may be treated with systemic treatment such as prednisone or other immunosuppressive drugs including but not limited to methotrexate, cyclosporine, azathioprine and mycophenolate mofetil. All of these drugs have important side effects and should always be discussed with your physician prior to starting treatment. Lastly, patients with widespread cutaneous lupus lesions may be treated with oral antimalarial pills, which work due to their anti-inflammatory properties (hydroxychloroquine, chloroquine). These medications also involve regular eye exams and routine blood tests. Your dermatologist will properly evaluate which treatment is right for you.

American Skin Association, 2016 www.americanskin.org

© ARRC 2016

The Autoimmune Resource and Research Centre (ARRC) is a Not for Profit registered health promotion charity. ARRC provides education, support and research services for people living with a range of systemic and organ-specific autoimmune diseases. For more information, education and support contact ARRC

www.autoimmune.org.au

HNELHD-arcc@hnehealth.nsw.gov.au

Pathology North Bldg, John Hunter Hospital, New Lambton Heights, NSW Australia 2305

ARRC information for patients, carers & Health Professionals

Disclaimer

This document has been developed and peer reviewed by ARRC and is based on expert opinion and the available published literature at the time of review. Information contained in this document is not intended to replace medical advice and any questions regarding a medical diagnosis or treatment should be directed to a medical practitioner. The development of this document is not funded by any commercial sources and is not influenced by commercial organisations. For more information about ARRC and its *policies & procedures* please refer to our website.

Content last updated July 2016

Reviewed by Judy Knapp, Clinical Trial Nurse