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A perspective on Systemic lupus erythematosus (SLE)

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Review Article

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ABSTRACT

Systemic lupus erythematosus (SLE) is a clinically heterogeneous disease, which is autoimmune disorder. It affect multiple organs like skin, joints, kidneys, brain, and other organs. Although liver dysfunction is not considered the main organ pathology or prognostic factor in patients with Systemic Lupus Erythematosus (SLE), it is not uncommon during the course of SLE. Liver complications in patients with SLE may be caused by lupus hepatitis (SLE-related hepatitis); autoimmune liver diseases, such as Autoimmune Hepatitis (AIH) and Primary Biliary Cirrhosis (PBC); viral hepatitis; and drug-induced liver injury. Here, liver complications in patients with SLE are reviewed with a predilection for females, with a female-to male ratio of between 4.3 and 13.6, and a mean age at diagnosis of 34.3 years. Most are young women of childbearing age who suffer from such symptoms as intense fatigue and exhaustion, joint pains, thinking and memory problems, and skin rashes. In young adult women the peak incidence of about 5 in 100 000 per year.

Introduction

Systemic lupus erythematosus (SLE) or lupus is asystemic autoimmune disease. Erythematosus is from the Greek word for red and refers to the color of the rash. Patients with lupus have unusual antibodies in their blood that are targeted against their own body tissues. Systemic lupus erythematosus (SLE) is a complex autoimmune disorder with diverse clinical manifestations. Women (especially African-American and Asian women) have a higher risk than men for developing SLE. Atherosclerosis is a pathological inflammatory process of the artery Myocardial Infarction (MI), stroke and critical limb ischemia are all end-stage consequences of progressive atherosclerosis, termed Cardiovascular Disease (CVD). The 'traditional risk factors' for CVD have been identified from longitudinal studies on a population from Framingham in the USA, and include increasing age, male sex, hypertension, smoking, dyslipidemia and diabetes mellitus. Involving a complex interplay of endothelial dysfunction, monocyte and T-lymphocyte intimal invasion and inflammatory cytokine production, producing lesions known as atheromata that are responsible for infarction of target organs, through occlusion of the affected vessel. Since the 1970s, improved survival rates in SLE patients have resulted in an increase in disease duration with the concomitant accrual of organ damage. Cardiovascular disease accounts for nearly 30% of the organ damage suffered by patients with lupus for 15 years.

Types of lupus:

1) *Cutaneous lupus erythematosus: There are 3 different types of cutaneous lupus:*

a) Chronic cutaneous lupus (CCLE): The most common form is discoid lupus. These lesions may also generalize to the v of the neck, upper back and dorsum of hands. These lesions are found in only about 20% of SLE patients. Discoid lesions that occur on the scalp may cause the hair to fall out. If the

lesions form scars when they heal, the hair loss may be permanent. 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.

b) Subacute cutaneous lupus (SCLE): Subacute cutaneous lupus erythematosus (SCLE) is a non-scarring, non–atrophy-producing, SCLE is often characterized by two forms including papulosquamous lesions and annular lesions, The lesions occur most commonly on the sun-exposed areas of the arms, shoulders, neck, and body.

c) Acute cutaneous lupus (ACLE): The lupus butterfly rash or malar rash is the most common form of ACLE. The most typical form of acute cutaneous lupus is a malar rash–flattened areas of red skin on the face that resemble a sunburn. Photosensitive dermatosis that is most often present in white women aged 15 to 40, consisting of skin lesions that are scaly and evolve as polycyclic annular lesions or psoriasiform plaques.

2) Drug-induced lupus: Drug-induced lupus erythematosus (DILE) is a variant of lupus erythematosus that resolves within days to months after withdrawal of the culprit drug in a patient with no underlying immune system dysfunction. The list includes medicines to treat heart disease, thyroid disease, hypertension, and neuropsychiatric disorders.

3) Neonatal Lupus Erythematosus: Neonatal lupus is a uncommon diseases form of lupus. At birth, the infant may have a skin rash, liver problems, or low blood cell counts but these symptoms disappear completely after several months with no lasting effects. Some infants with neonatal lupus can also have a serious heart defect. With proper testing, physicians can now identify most at-risk mothers, and the infant can be treated at or before birth. It is a rare condition that affects infants of women who have lupus and is caused by antibodies from the mother acting upon the infant in the womb.

4) Childhood Lupus: The most common types of lupus which are easily diagnosed. This kind of lupus can easily affect any part of body. And systemic lupus is more sever or serious than the other forms of lupus. It is marked by chronic inflammation, especially of the kidneys, joints, and skin. In the United States, the highest incidence is found in African-Americans.

5) Systemic lupus: Mainly childhood lupus occurs in the child although many of the symptoms are same as the adult lupus. Childhood lupus mainly affects the boy and it more likely to affects the kidney. Childhood lupus may require more aggressive treatment than adult lupus.

Signs and Symptoms

- **Severe fatigue:** two types of fatigue they separated physical fatigue from mental fatigue. Fatigue is one of the most common symptoms of lupus such as such as anemia, fibromyalgia, depression, or a kidney or thyroid problem.
- **Headaches:** Headache is a symptom commonly described by SLE patients Different studies have done on headache out of which migraine 31.7% and tension-type headache 23.5%. While headaches were linked to a lower health-related quality of life, Findings indicate that at the commencement of the study nearly 18% of participants had headache -- 61% with migraine, 37% tension, 7% intractable non-specific, 3% cluster and 1% intracranial hypertension
- **Hair loss:** Lupus hair loss tends to be minor and cause hair 'thinning' rather than bald patches. Most people don't even notice the 50 to 100 strands of hair that they usually lose every day. Lupus hair loss can be caused as the disease.
- **Anemia:** Anemia is usually defined as the less number of RBC counts and hemoglobin. Anemia is common in SLE patients. Mainly 50% of SLE patients the same problems.
- **Blood and lymph:** this is also called as mild anemia problems. They also include other problems of the blood like less number of blood cells or platelets counts. Sometimes the lymph also may get swell.

1. **Raynaud's syndrome:** Raynaud's disease is a rare disorder of the blood vessels, supplying the skin- such as your fingers and toes- to feel numb and cold in response to cold temperatures or stress. In most cases this discomfort will lasts only for short time. In raynaud's syndrome arteries that supply

blood to your skin narrow, limiting blood circulation to affected areas (vasospasm). Women are more likely than men to have Raynaud's disease, is known as Raynaud or Raynaud's phenomenon or syndrome. It appears to be more common in people who live in colder climates.

Common causes for Raynaud's syndrome:

- Diseases of the arteries (such as atherosclerosis and Buerger's disease)
- Drugs which narrow the supply to arteries (such as amphetamines, certain types of beta-blockers, some cancer drugs, certain drugs used for migraine headaches)
- Arthritis and autoimmune conditions
- Smoking
- Frostbite
- Thoracic outlet syndrome

Types of Raynaud's syndrome
There are two types of Raynaud's:

- Primary - when the condition develops by itself (this is the most common type)
- Secondary - when it's caused by another health condition

2. **Sjögren's Syndrome:** It is a common disorder of your immune system identified by its two most common symptoms-dry eyes and a dry mouth. Mostly sjogrens syndrome are occurs in the women's. Although you can develop Sjogren's syndrome at any age, most people are older than 40 at the time of diagnosis. The misdirected immune system in autoimmunity tends to lead to inflammation of tissues. This particular autoimmune illness features inflammation in glands of the body that are responsible for producing tears and saliva. About 50% of patients with Sjögren syndrome have cutaneous findings, such as dry skin (xeroderma), palpable and nonpalpable purpura, and urticaria.

Causes

Hormones: Hormones are the body's messengers. About 90% of patients are women, most diagnosed when they are in their childbearing years SLE affects more women than men. Women also experience worsening of symptoms during pregnancy and with their menstrual periods. The research is still need to prove that female hormone estrogen may play a role in causing SLE.
Genetic: The first mechanism may arise genetically. Research indicates SLE may have a genetic link. SLE does run in families, but no single causal gene has been identified. Instead, multiple genes appear to influence a person's chance of developing lupus when triggered by environmental factors. No gene or group of genes has been proven to cause lupus. Lupus does, however, appear in certain families, and certain genes have been identified as contributing to the development of lupus, but these associations alone are not enough to cause the disease.
Environment: There may be environmental triggers like ultraviolet rays, certain medications, a virus, physical or emotional stress, and trauma. Most researchers today think that an environmental agent, such as a virus or possibly a chemical, randomly encountered by a genetically susceptible individual, acts to trigger the disease. Researchers have not identified a specific environmental agent as yet but the hypothesis remains likely.

Treatment and Diagnosis

Nonsteroidal anti-inflammatory drugs (NSAIDs): Inflammation may be defined as the series of changes that occur in living tissues following injury. Injury may be caused due to various exogenous and endogenous stimuli, which in turn initiate protective response in the host cell. Examples of these are ibuprofen, naproxen and diclofenac. The main possible side-effects from NSAIDs are stomach and gut problems such as pain or bleeding in the stomach.

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Antimalarial drugs-Plaquenil (Hydroxychloroquine): Antimalarial drugs are used for skin and joint problems.

Corticosteroids: Prednisone and other types of corticosteroids can counter the inflammation of lupus. Steroid tablets are usually advised if you develop more severe symptoms. Steroids reduce inflammation and the dose is usually given at the lowest possible dose, in order to reduce any side-effects from the steroids. Steroid creams for rashes. Corticosteroids often produce several long-term side effects like weight gain, bruising easily, thinning of the bones, diabetes and increased risk of infection.

Immune suppressants: Such as: Belimumab (Benlysta), CellCept (mycophenolate mofetil), Cyclosporine, Cytoxan (cyclophosphamide), Imuran (azathioprine), Methotrexate (Trexall).

No single test can confirm or rule out SLE. A number of tests are required before SLE can be diagnosed definitively. The first symptoms of SLE can resemble one of many syndromes or disorders, including rheumatoid arthritis, Still's disease, rheumatic fever, Lyme disease, multiple sclerosis. A doctor will make a diagnosis of SLE based on symptoms, medical history, physical exam, and blood test for antinuclear antibodies.

General Criteria for Diagnosing System Lupus Erythematosus (SLE)

- Characteristic rash across the cheek
- Discoid lesion rash
- Photosensitivity
- Oral ulcers
- Arthritis
- Immunologic abnormalities
- Positive antinuclear antibody (ANA)
- Evidence of severe neurologic disease

Conclusion

Systemic lupus erythematosus (SLE) is an autoimmune disorder. Systemic Lupus Erythematosis is not easy to say, and even more difficult to diagnose. SLE is a chronic disease, primarily affecting skin, joints, kidneys, heart, lungs, nervous system, blood elements, and serosal membranes. The patient and medical provider's goal is to find the most effective treatment plan for each individual patient to manage the symptoms with which they are presenting. Many ocular complications and disease deterioration are preventable, treatable, or even curable, thus inter-department collaboration is highly important. This evolves over time through working at maintaining a balance between preventing flares and the potentially life-threatening organ damage they can cause. Prompt treatment with high dose systemic corticosteroid and immunosuppressive therapy are necessary in many circumstances. People who develop an autoimmune disease may have a genetic predisposition that is triggered by some environmental factor such as sunlight, stress hormones, or viruses.

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