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SYSTEMIC LUPUS ERYTHEMATOSUS: DIAGNOSIS AND CLINICAL MANAGEMENT

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Abstract:

A worldwide chronic autoimmune disorder, systemic lupus erythematosus terribly affects each organ and tissue. Disease development and activity may be influenced by a few factors including environmental triggers, hormonal abnormalities, and genetic variation. The heterogeneity is existed in the clinical pattern and between the patients which fluctuate treatment over time in every patient. The complex pathology of SLE includes the immune complexes, abrupt regulation of various cytokines, and disturbance in the clearance of nucleic acid followed by cell death. The aim of treatment is to achieve a lower degree of activity through the immunomodulators, immunosuppression, and stave off the organ from the harmful effect of active lupus. Diminish the secondary diseases and treatment to overcome accelerating atherosclerosis and decrease the major reasons that lead to death. Commence the treatment at an early stage as soon as possible after the diagnosis is very essential to bring back the patient to normal life.

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INTRODUCTION:

The multi-systemic inflammatory auto-immune disorder, systemic lupus erythematosus is an impaired self-sustained autoimmune process with complexities in apoptotic clearance, up-regulation of the innate and adaptive immune system, and tissue inflammation. The relapse manifestations develop a spectrum of clinical complexities from ruinous life-threatening. Many pathogenic mechanisms of systemic lupus erythematosus can be linked with clinical phenotype. [1] The heterogeneity is existed in the clinical pattern and between the patients which fluctuate over time in every patient. This is the main reason, makes the diagnosis more difficult or impeded and based on immunological and clinical findings by keen expertise. Clinically, SLE is a challenge for clinicians, while it affects many organs and tissues with varying degrees of severity. [2] A research proposed the diagnosis and management of SLE through the secondary and tertiary centers have experience in the disease, to make sure exact initiation of therapy and recognized the early and control of flares to maximize the medical care over the disease course. [3] However, we will discuss the classification criteria and evidence-based treatments.

Epidemiology:

Epidemiological studies elucidate the prevalence and incidence of SLE according to age, sex, and ethnicity. Globally, lupus disease is a striking inclination for women of childbearing age. The prevalence ratio is 13:1 for female and males aged 15 to 44 years old and for children is 2:1. In Europe and the United States, frequently found in people of African descent and more common in African-Americans with worse consequences. [4] Noticeably, African-American women are 3 times more sensitive to develop lupus with a high mortality rate than Caucasian-Americans. According to an estimated report of the Center for Disease Control and Prevention, on prevalence is about 321,000 cases with SLE, a large number in African-American, American, Indians and Alaska Natives. [5]

Clinical Diagnosis:

In the segment of clinical diagnosis of this disorder, we elucidate key organ manifestations and important advancement in lupus diagnosis.

Cutaneous Lupus:

Generally, 90% of patients of SLE have must experience cutaneous lupus including acute and sub-acute cutaneous lupus, and chronic cutaneous lupus (discoid lupus, lupus profundus, chilblain lupus, and lupus tumidus. The majority forms of cutaneous lupus exhibit the same histological results as interface dermatitis with perivascular inflammation and at the junction of the dermo-epidermal junction have immunoglobulin. [6] In cutaneous lupus, a biopsy is considered a fundamental part of the diagnosis. The photo-sensitive (polymorphous light eruption or rosacea) rashes are the distinct characteristics of Cutaneous lupus erythematosus. Discoid lupus can be converted into permanent scarring lupus, smoking may enhance the risk of discoid lupus with ongoing activity and diminish the benefit of hydroxychloroquine. [7]

Musculoskeletal Involvement:

Commonly, arthralgia and synovitis have found in 90% of cases of lupus. Fibromyalgia, Monoarthritis are potently associated with musculoskeletal pain. The difference in active inflammatory disease can be carried out through the articular ultrasound and magnetic resonance. Patients with lupus usually experienced depression in cases of musculoskeletal pain. [8]

Renal disease:

In lupus disease 50% of the patient must experience the renal-related disorders and nephritis is the main cause of morbidity and mortality in SLE, impede the diagnosis associated with end-stage renal disorder. Proteinuria provided the clue to diagnosis renal disorder. A biopsy is deemed a key for renal diagnosis in patients with lupus disorder. The high level of urine protein (above 500mg/24) is the link with lupus nephritis and prompt biopsy recommended. [9] other renal diseases include tubulointerstitial nephritis, vascular disease (such as thrombotic microangiopathy, vasculitis), and collapsing glomerular sclerosis. [10]

Central nervous system disease:

Neuropsychiatric has a wide association with SLE. Although merely a few of them are specified for SLE diagnosis. It may involve seizures, psychosis, mononeuritis multiplex, myelitis, peripheral or cranial neuropathy, and an acute confusional state. [11] The essential diagnostic process includes magnetic resonance imaging (MRI) and cerebral spinal fluid analysis. The detection of chronic microvascular changes, infarcts, hemorrhages, cortical atrophy, edema, abscesses, transverse and longitudinal myelitis can be conducted by the central nervous system MRI. Active neuropsychiatric lupus can identify by abnormal cerebrospinal fluid IgG and oligoclonal bands. The majority of patients of SLE show cognitive impairment at the time of diagnosis that can be detected through psychometric testing and psychiatric analysis. Depression is the most

prominent cognitive impairment in SEL patients with a central nervous abnormality. [12]

Management:
The aim of treatment is to achieve a lower degree of activity through the immunomodulators, immunosuppression, and stave off the organ from the harmful effect of active lupus. Diminish the secondary diseases and treatment to overcome accelerating atherosclerosis and decrease the major reasons that lead to death. Commence the treatment at an early stage as soon as possible after the diagnosis is very essential to bring back the patient to normal life. The rationale few treatments are given below.

Hydroxychloroquine:
Merely this medication ameliorates the survival of an individual with lupus. Interestingly, reduced the lupus flares, protect the organ from such cardiovascular related issues, reduced the seizures, and overcome the risk associated with neuropsychiatric lupus. In a clinical study, hydroxychloroquine lower the skin manifestation, control the insulin resistance, and abolish the probability of infection and malignancy. [13]

Vitamin D:
The immunomodulatory and anti-fibrotic effect of vitamin D encourages to recommend in all patients with SLE. Vitamin D deficits are linked with the enhancement risk of multi-organ fibrosis including the lungs and kidneys. [14] The findings of multiple studies concluded that in patients with SLE, vitamin D deficits associated with a high risk of thrombosis and intensify the disease activities and fatigue. Vitamin D supplementation ameliorates the global disease activity in SLE, abolish the proteinuria, and routinely monitoring is necessary to assess the proper absorption and dosing. [15]

Dehydroepiandrosterone (DHEA):
It is known as an adrenal hormone regulator and acts as a precursor of estrogen and androgen. On the base of randomized clinical trials on women, DHEA has an efficacious response in disease improvement along with the cytokine and bone density profile. Importantly, women with lupus have experienced a low level of androgen and high estradiol. However, its use is prohibited in women with menopause as it may develop hormone-sensitive malignancies while no evidence was found for DHEA for men use. [16]

Other medication use for the treatment of SLE may include corticosteroids, cyclophosphamide, azathioprine, methotrexate, mycophenolate, and calcineurin inhibitors.

Lifestyle:
The living style also has an effect to improve the disease condition. Patients should protect themselves through the sun's rays using protective cloths and sunscreens. Proper exercise, stretching will help to reduce fatigue, pain from fibromyalgia, and cognitive dysfunction. [17]

Prevention of comorbidities:
Lupus has increased 2-3 time mortality cases and a huge number of deaths in lupus is because of cardiovascular disorder, infections, and the renal and respiratory complication of lupus. Therefore, vigorous management for traditional causes namely, smoking, obesity, diabetes, and hypertension, lupus activity, and to minimized the cardiovascular risk factor to prevent untimely death. [18]

DISCUSSION:

- An attempt to study the Systemic Lupus Erythematosus (SLE) concluded that 55.5% of patients with the cutaneous disorder, 20% with renal dysfunction and 10% with central nervous disorder were found during the study. The proper medical required for a patient's diagnosis with this disease as can build many other complexities and more worsen the disease. [19]
- The findings of another clinical trial base on the diagnosis of systemic lupus erythematosus indicate the existence of lupus erythematosus cells in 76.7% of patients, skin biopsies in 5%, and renal biopsies in 1.3%. Myalgia was found in 35% of patients while no history was detected with cutaneous involvement. Renal abnormalities were noticed in 28% of patients. [20]

CONCLUSION:
To overcome the difficulty in the way to ameliorate the treatment of SEL, cooperation from multi-discipline specialists is needed at each level of medical care including primary, secondary, and tertiary which help to overcome the disease burden at the community level.

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