ONLINE EXCLUSIVE

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Systemic lupus erythematosus

THE COMPARISON

A A 23-year-old White woman with malar erythema from acute cutaneous lupus erythematosus. The erythema also can be seen on the nose and eyelids but spares the nasolabial folds.

B A Black woman with malar erythema and hyperpigmentation from acute cutaneous lupus erythematosus. The nasolabial folds are spared.

C A 19-year-old Latina woman with malar erythema from acute cutaneous lupus erythematosus. The erythema also can be seen on the nose, chin, and eyelids but spares the nasolabial folds. Cutaneous erosions are present on the right cheek as part of the lupus flare.

Systemic lupus erythematosus (SLE) is a chronic autoimmune condition that affects the kidneys, lungs, brain, and heart, although it is not limited to these organs. Dermatologists and primary care physicians play a critical role in the early identification of SLE (particularly in those with skin of color), as the standardized mortality rate is 2.6-fold higher in patients with SLE compared to the general population.[1] The clinical manifestations of SLE vary.

Epidemiology

A meta-analysis of data from the Centers for Disease Control and Prevention National Lupus Registry network including 5417 patients revealed a prevalence of 72.8 cases per 100,000 person-years.[2] The prevalence was higher in females than males and highest among females identifying as Black. White and Asian/Pacific Islander females had the lowest prevalence. The American Indian (indigenous)/Alaska Native–identifying population had the highest race-specific SLE estimates among both females and males compared to other racial/ethnic groups.[2]

Key clinical features in people with darker skin tones

The diagnosis of SLE is based on clinical and immunologic criteria from the European League Against Rheumatism/American College of Rheumatology.[3,4] An antinuclear antibody titer of 1:80 or higher at least once is required for the diagnosis of SLE, as

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long as there is not another more likely diagnosis. If it is present, 22 additive weighted classification criteria are considered; each criterion is assigned points, ranging from 2 to 10. Patients with at least 1 clinical criterion and 10 or more points are classified as having SLE. If more than 1 of the criteria are met in a domain, then the one with the highest numerical value is counted.[3,4]

Aringer et al[3,4] outline the criteria and numerical points to make the diagnosis of SLE. The mucocutaneous component of the SLE diagnostic criteria[3,4] includes nonscarring alopecia, oral ulcers, subacute cutaneous or discoid lupus erythematosus,[5] and acute cutaneous lupus erythematosus, with acute cutaneous lupus erythematosus being the highest-weighted criterion in that domain. The other clinical domains are constitutional, hematologic, neuropsychiatric, serosal, musculoskeletal, renal, antiphospholipid antibodies, complement proteins, and SLE-specific antibodies.[3,4]

The malar ("butterfly") rash of SLE characteristically includes erythema that spares the nasolabial folds but affects the nasal bridge and cheeks.[6] The rash occasionally may be pruritic and painful, lasting days to weeks. Photosensitivity occurs, resulting in rashes or even an overall worsening of SLE symptoms. In those with darker skin tones, erythema may appear violaceous or may not be as readily appreciated.[6]

Worth noting

- Patients with skin of color are at an increased risk for postinflammatory hypopigmentation and hyperpigmentation (pigment alteration), hypertrophic scars, and keloids.[7,8]
- The mortality rate for those with SLE is high despite early recognition and treatment when compared to the general population.[1,9]

Health disparity highlight

Those at greatest risk for death from SLE in the United States are those of African descent, Hispanic individuals, men, and those with low socioeconomic status,[9] which likely is primarily driven by social determinants of health instead of genetic patterns. Income level, educational attainment, insurance status, and environmental factors[10] have far-reaching effects, negatively impacting quality of life and even mortality. JFP

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