

The performance of the EULAR/ACR 2019 classification criteria for monogenic lupus

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Objective

To evaluate the application of the European League Against Rheumatism/American College of Rheumatology (EULAR/ACR-2019) criteria to monogenic lupus patients and compare its performance against the Systemic Lupus International Collaborating Clinics (SLICC-2012) criteria.

Methods

In a multicenter retrospective cohort study, consecutive patients with monogenic lupus from three tertiary lupus clinics were enrolled. The diagnosis of monogenic lupus was based on the expert physician's opinion and fulfilling SLICC-2012 criteria. Also, all enrolled patients had genetic variants. A control group of sporadic childhood systemic lupus erythematosus (cSLE) and non-SLE patients including systemic idiopathic arthritis (sJIA) and juvenile dermatomyositis (JDM) were included. A descriptive data analysis was conducted, and the EULAR/ACR-2019 and SLICC-2012 criteria were applied to both groups.

Results

For these analyses, a total of 153 (100 females) patients were available. Forty-nine patients with monogenic lupus with a median age at diagnosis of 4.0 (IQR 2.0-8.0) years, and 104 controls (55 patients with cSLE and 49 non-lupus patients with a median age at diagnosis of 10.0 and 5.0 respectively) were included. Forty-three (87.8%) patients with monogenic lupus fulfilled EULAR/ACR-2019 with a mean score of 22.3±13.1. The most frequent domains were immunologic (93.9%), musculoskeletal and renal (each 57.1%), and mucocutaneous (55.1%). Fifty-four (98.2%) cSLE patients and six (12.2%) non-lupus patients met the criteria with a mean score of 22.8±8.1 and 14.7±10.2 respectively. The sensitivity of the 2019-EULAR/ACR criteria in monogenic lupus was 91.4% (95% CI: 0.8-0.9) while the specificity was 83.3% (95% CI: 0.5- 0.9).

Conclusion

This is the first and largest cohort of monogenic lupus patients testing the performance of the 2019-EULAR/ACR criteria. It efficiently classifies monogenic lupus patients, irrespective of the underlying genetic variants. Further studies are needed before these new criteria are adopted worldwide.