Correspondence

Juvenile systemic lupus erythematosus in Bahrain. *A tertiary referral center experience*

To the Editor

I read the interesting article by Al-Mosawi et al[1] on juvenile systemic lupus erythematosus in Bahrain. A tertiary referral center experience. I have 2 comments considering that article.

First, the diagnosis of systemic lupus erythematosus (SLE) in the studied patients was based on fulfilling at least 4 of the 1997 revised American College of Rheumatology classification criteria for SLE.[2] These criteria have inherent limitations, including bias towards more severe and longer duration disease, equal weighting of features that vary in clinical significance, and exclusion of patients with SLE from research because they do not meet 4 criteria.[3] Accordingly, I presume that many patients with SLE who did not fulfill the 4 criteria of the American College of Rheumatology classification criteria were misdiagnosed and cannot be included in the study. This might alter the results and conclusions presented by the authors. Therefore, revised criteria for SLE are needed and establishment of new classification is crucial to facilitate researches and guide clinical practice.

Second, the Al-Mosawi et al[1] study added additional cases of sicklers with coexistent SLE to the list of sporadic cases previously published. Patients with sickle cell disease (SCD) present with a defective activation of the alternate pathway of the complement system that increases the risk of capsulated bacterial infection and failure to eliminate antigens, predisposing these patients to autoimmune diseases.[4] The immunological background of the coexistence of SLE with SCD might be explained by the observation that up to 23% of patients with SCD have antinuclear antibodies.[5,6] Pediatricians should be knowledgeable to the possible coexistence of SLE in patients with SCD, and appropriate actions are undertaken.

Mahmood D. Al-Mendalawi
Department of Paediatrics
Al-Kindy College of Medicine
Baghdad University
Baghdad, Iraq

Reply from the Author

We are grateful for valuable comments of Dr. Al-Mendalawi on our article Juvenile systemic lupus erythematosus in Bahrain. A tertiary referral center experience.[1] In order to study cases with SLE and compared it with other study, we need to refer to an updated criteria that the other studies rely on to diagnose SLE. The diagnosis of SLE is in general made based on American College of Rheumatology criteria as commonly used by the rheumatologists although there were no specific criteria developed as diagnostic criteria yet. Since our study retrospectively analyzed the clinical and serological features, no place was left for including the uncompleted or latent cases of SLE. In other words, including cases diagnosed at presentation may lead to over diagnosis of SLE. Midline search in the last 4 decade for making diagnosis of SLE revealed many studies and different results. Historically Cohen et al published the first ACR criteria in 1971,[7] these criteria were updated in 1982 and later revised in 1997 where antiphospholipid antibody and lupus anticoagulant were added to 1882 criteria.[8,9] Clough et al[10] (1984) had described weighted criteria (WC) for the diagnosis of SLE and Costenbender et al[11] (2001) had modified these criteria. They concluded that modified weighted criteria had better overall psychometric properties than the ACR criteria. However WC was more sensitive but less specific than ACR criteria as shown; sensitivity and specificity were 90.3% and 60.4 versus 86.5% and 71.9%, respectively.[10,11] In their conclusion, authors admitted that the weighted criteria may capture more lupus patients for clinical or interventional studies than the ACR criteria, but some of these patients may have no lupus according to experienced rheumatologist. Those patients who were diagnosed clinically but who did not meet the ACR criteria are different from those who met the criteria; they are sometime described as having incomplete or latent lupus.

Based on the specificity of ACR criteria, we included patients who fulfilled the ACR criteria. The aim was to exclude the suspected cases and to compare the clinical and the serological manifestations accurately with other international studies. Regarding SLE in SCD patients, the SLE diagnosis was given to these patients based on fulfilling the ACR criteria and not only on the presence of autoantibody. Definitely, knowing such co-existence is important to minimize any delay in diagnosis of SLE in SCD patients.

Zakiya Al-Mosawi
Department of Pediatrics
Salmaniya Medical Complex
Manama, Kingdom of Bahrain

References

1. Al-Mosawi Z, Al-Hermi BE, Al-Saad KK, Farid EM, Makki HA. Juvenile systemic lupus erythematosus in Bahrain. A tertiary referral center experience. ***Saudi Med J*** 2009; 30: 667-672.
2. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. ***Arthritis Rheum*** 1997; 40: 1725.
3. Smith EL, Shmerling RH. The American College of Rheumatology criteria for the classification of systemic lupus erythematosus: strengths, weaknesses, and opportunities for improvement. ***Lupus*** 1999; 8: 586-595.
4. Appenzeller S, Fattori A, Saad ST, Costallat LT. Systemic lupus erythematosus in patients with sickle cell disease. ***Clin Rheumatol*** 2008; 27: 359-364.
5. Baethge BA, Bordelon TR, Mills GM, Bowen LM, Wolf RE, Bairnsfather L. Antinuclear antibodies in sickle cell disease. ***Acta Haematol*** 1990; 84: 186-189.
6. Khalidi NA, Ajmani H, Varga J. Coexisting systemic lupus erythematosus and sickle cell disease: a diagnostic and therapeutic challenge. ***J Clin Rheumatol*** 2005; 11: 86-92.
7. Cohen AS, Reynolds WE, Franklin EC et al. Preliminary criteria for the classification of systemic lupus erythematosus. ***Bull Rheum Dis*** 1971; 21; 643-648.
8. Tan EM, Cohen AS, Fries JF, Masi AT, McShane DJ, Rothfield NF, et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum. 1982; 25: 1271-1277.
9. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. ***Arthritis Rheum*** 1997; 40: 1725.
10. Clough JD, Elrazak M, Calabrese LH, Valenzuela R, Braun WB, Williams GW. Weighted criteria for the diagnosis of systemic lupus erythematosus. ***Arch Intern Med*** 1984; 144: 281-285.
11. Costenbader KH, Mandl LA, Liang MH, Improving the utility of ACR criteria for clinical trial. ***Lupus*** 2001; 10: S117.

Illustrations, Figures, Photographs

Four copies of all figures or photographs should be included with the submitted manuscript. Figures submitted electronically should be in JPEG or TIFF format with a 300 dpi minimum resolution and in grayscale or CMYK (not RGB). Printed submissions should be on high-contrast glossy paper, and must be unmounted and untrimmed, with a preferred size between 4 x 5 inches and 5 x 7 inches (10 x 13 cm and 13 x 18 cm). The figure number, name of first author and an arrow indicating "top" should be typed on a gummed label and affixed to the back of each illustration. If arrows are used these should appear in a different color to the background color. Titles and detailed explanations belong in the legends, which should be submitted on a separate sheet, and not on the illustrations themselves. Written informed consent for publication must accompany any photograph in which the subject can be identified. Written copyright permission, from the publishers, must accompany any illustration that has been previously published. Photographs will be accepted at the discretion of the Editorial Board.