Indian Journal of Pediatrics (December 2021) 88(12):1167–1168
https://doi.org/10.1007/s12098-021-03971-7

EDITORIAL COMMENTARY

Autoantibodies in Juvenile Dermatomyositis: *Need to Bridge the Bench and Bedside Gap*

Narendra Kumar Bagri[1]

Received: 13 September 2021 / Accepted: 16 September 2021 / Published online: 5 October 2021

Juvenile dermatomyositis (JDM) is an autoimmune disease, and is associated with various autoantibodies. These autoantibodies are classified as either myositis-specific antibodies (MSAs) or myositis-associated antibodies (MAAs). MSAs are exclusively detected in patients with idiopathic inflammatory myopathy, while MAAs are less specific and are observed in patients with myositis as a part of other connective tissue diseases [1]. The MSAs are associated with a specific disease phenotype; for example, anti-TIF-ϒ is associated with severe cutaneous disease including lipodystrophy, anti-MDA-5 is related to interstitial lung disease, whereas anti-SRP antibodies are associated with profound muscular weakness and Raynaud phenomenon [1, 2].

In this issue of the Journal, Sharma et al. have published their study describing the autoantibody profile in 34 children with JDM [3]. They detected either MSA or MAA in 23.5% (8/34) of children with JDM. These observations are in stark contrast to a large cohort from the UK reporting 59% (225/380) antibody positivity, and 50% of this cohort had MSAs [1]. The findings of the Indian cohort are also different from a Japanese study wherein 95.2% (20/21) were positive for MSAs [4]. In both UK and Japanese cohorts, anti-TIF-ϒ was the most common antibody, whereas none in the Sharma et al. study had anti-TIF-ϒ antibody [3]. These differences in the frequency and type of MSAs may be attributed to genetic and other study characteristics (e.g., children with overlap syndrome were excluded from the Indian cohort). The ongoing immunosuppression might also have contributed to these varying observations; however, this is mere speculation, as this study does not describe the treatment details and its correlation with MSA/MAAs. The most commonly observed antibody in the present study is anti-SRP antibody, which contrasts to the UK cohort where it was found only in 2% of the subjects. The characteristic muscular weakness is described in 3 out of 4 children with anti-SRP antibodies; however, the authors did not mention Raynaud phenomenon in these subjects, another characteristic finding reported in association with anti-SRP antibodies. The 3 children with anti-MDA-5 antibody did not have any clinical evidence of interstitial lung disease (a characteristic of anti-MDA-5–associated JDM); however, the chest CT findings have not been described in this study. Although the present study has described some clinical characteristics, the small sample size and lack of details about treatment and muscle enzyme preclude this study from making a meaningful conclusion regarding association of MSAs and clinical phenotype in Indian children. Despite these limitations, this study gives thoughtful insight about autoantibodies in JDM.

Presently MSAs do not constitute a part of classification/diagnostic evaluation for JDM; the emerging evidence suggests that autoantibodies in JDM correlate with specific disease phenotypes. Moreover, the uniqueness of MSA in an individual patient (i.e., usually, 1 patient shall have only 1 MSA) highlights the potential utility of MSAs in predicting the disease phenotype and guiding therapeutic plan for these patients. Therefore, large multicentric studies are needed to further delineate the clinical implicationsof MSAs in children with JDM before advocating them for routine clinical practice.

Declarations

Conflict of Interest None.

✉ Narendra Kumar Bagri
drnarendrabagri@yahoo.co.in

[1] Division of Pediatric Rheumatology, Department of Pediatrics, All India Institute of Medical Sciences, New Delhi 110029, India

References

1. Tansley SL, Simou S, Shaddick G, et al. Autoantibodies in juvenile-onset myositis: their diagnostic value and associated clinical phenotype in a large UK cohort. J Autoimmun. 2017;84:55–64.
2. Wu JQ, Lu MP, Reed AM. Juvenile dermatomyositis: advances in clinical presentation, myositis-specific antibodies and treatment. World J Pediatr. 2020;16:31–43.
3. Sharma A, Bhattarai D, Gupta A, et al. Autoantibody profile of children with juvenile dermatomyositis. Indian J Pediatr. 2021. https://doi.org/10.1007/s12098-021-03680-1.
4. Iwata N, Nakaseko H, Kohagura T, et al. Clinical subsets of juvenile dermatomyositis classified by myositis-specific autoantibodies: experience at a single center in Japan. Mod Rheumatol. 2019;29:802–7.