IS PEDIATRIC IGG4-RELATED DISEASE DISTINCT FROM ITS ADULT COUNTERPART? A SINGLE CENTER CASE SERIES

Michael B Nelson[1], Daniella Schocken[2], Marietta De Guzman[3]

[1] Baylor College of Medicine, Department of Pediatrics, Immunology, Allergy and Rheumatology

[2] Baylor College of Medicine, Pediatrics, Resident

[3] Baylor College of Medicine, Pediatrics, Rheumatology

Background: IgG4-related disease (IgG4-RD) is a poorly understood and rarely diagnosed fibro-inflammatory condition in pediatrics. While myriad presentations of this disease have been described in the adult literature for centuries, only recently has this disease been recognized as a clinical entity in children. Indeed, consensus as to best practices in the diagnosis and treatment of pediatric IgG4-RD has not yet been established.

Materials/Methods: Retrospective chart review identified 4 patients who presented for evaluation by Pediatric Rheumatology for IgG4-RD at TCH from January 2012 to May 2019. Median age at presentation was 13.2 years (range 4 – 15.3). 3 patients presented with IgG4-related ocular disease, while one presented with sialadenitis. 3 of the 4 patients met the criteria for "histologically highly suggestive of IgG4-RD" by the consensus criteria outlined by Deshpande, et al (2012). The patient not satisfying criteria did not meet secondary to inadequate IgG4 per HPF but did satisfy the requirement of >40% IgG4 to IgG ratio. 2 of the 4 patients met the newly proposed ACR/EULAR IgG4-related disease classification.

Results: All 4 patients received treatment with oral and/or IV steroids. 3 patients received steroid-sparing therapy: 2 patients were treated with methotrexate, 1 with mycophenolate. All 3 of the patients receiving steroid sparing therapy also received rituximab. All of these patients were subsequently considered in remission after receipt of rituximab. Additional autoimmunity was noted in 1 patient, malignancy was diagnosed approximately 2 years after initial presentation in another patient, 1 has had clinical/imaging evidence of resolution of IgG4 disease but continues to have evidence of systemic inflammation, and 1 patient was recently diagnosed and evaluation remains ongoing.

Conclusions: IgG4-RD remains a rare diagnosis in pediatrics, especially when compared with adults. While classification criteria with excellent specificity and sensitivity have been developed for adult disease, it remains to be seen whether these criteria are entirely applicable to pediatric IgG4-RD. Moreover, treatment regimens suggested in adult IgG4-RD rely heavily on glucocorticoids, which may be both inadequate and unacceptable secondary to morbidity with long-term use. Additional research is needed to better define pediatric IgG4-RD, develop and validate classification criteria, and determine optimal therapeutic regimens.