Response to: Caution the masqueraders of Good's syndrome on the thymoma with systemic lupus erythematosus

Arash Mollaeian¹ · Christopher J. Haas^{1,2}

Received: 25 December 2020 / Revised: 25 December 2020 / Accepted: 29 December 2020 / Published online: 9 January 2021
© International League of Associations for Rheumatology (ILAR) 2021

Dear Editor:

We appreciate Liu et al. for their correspondence [CLRHD-20-02326] and careful review of our case-based review on post-thymectomy systemic lupus erythematosus [1]. The point raised by Liu et al. regarding the possibility of Good syndrome in the reported case, considering the coexistence of thymoma and pure red cell aplasia, is well received and was considered as a differential in our case as well. Good syndrome is characterized by thymoma, hypogammaglobulinemia, and peripheral B cell lymphopenia [2]. Thymoma may precede or present simultaneously with hypogammaglobulinemia and lymphopenia; however, cases of Good syndrome diagnosed years after thymectomy have been reported in literature as well [3, 4]. In contrast to the predisposition to common infections in cases of Good syndrome [5], the patient in our report did not experience such opportunistic infections—sinopulmonary infections with encapsulated organisms, cytomegalovirus colitis, and mucocutaneous candida infections before, during, or after diagnosis of thymoma, and his infections were mostly multi-drug-resistant bacterial infections in the setting of neutropenia [1]. Unfortunately, no dedicated peripheral B cell or immunoglobulin studies are available at or around the time of thymoma diagnosis. Nevertheless, as part of further work-up for his severe anemia after thymectomy, a bone marrow biopsy and flow cytometry were performed, demonstrating a normal CD4:CD8 ratio and an absence of immunophenotypic evidence of monocytic population of B cells, abnormal T cells, or increased blasts. Furthermore, later in the course of disease, at the time

that patient developed autoimmunity and was diagnosed with systemic lupus erythematosus, serum protein electrophoresis (SPEP) revealed no evidence of monoclonal protein in the gamma region and diffuse patterns of gamma, alpha, mu, kappa, and lambda chains of immunoglobulins and a normal kappa:lambda ratio. While Good syndrome was certainly on the differential in our case, the lack of peripheral B cell depletion and hypogammaglobulinemia as evidence by bone marrow flow cytometry and SPEP, respectively, makes this diagnosis extremely unlikely.

Compliance with ethical standards

Disclosures None.

References

1. Mollaeian A, Haas C (2020) A tale of autoimmunity: thymoma, thymectomy, and systemic lupus erythematosus. *Clin Rheumatol* 39(7):2227–2234
2. Tarr PE et al (2001) Infections in patients with immunodeficiency with thymoma (Good syndrome). Report of 5 cases and review of the literature. *Medicine (Baltimore)* 80(2):123–133
3. Furukawa R, Yanagiya M, Matsumoto J, Hashimoto H, Horiuchi H, Usui K (2018) Good's syndrome with clinical manifestation after thymectomy: a case report. *Respir Med Case Rep* 24:89–91
4. Kelesidis T, Yang O (2010) Good's syndrome remains a mystery after 55 years: a systematic review of the scientific evidence. *Clin Immunol* 135(3):347–363
5. Kelleher P, Misbah SA (2003) What is Good's syndrome? Immunological abnormalities in patients with thymoma. *J Clin Pathol* 56(1):12–16

Publisher's note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

✉ Arash Mollaeian
arash.mollaeian@gmail.com

¹ MedStar Health Internal Medicine Residency Program, Department of Internal Medicine, MedStar Union Memorial Hospital, 201 E University Parkway, Baltimore, MD 21218, USA

² Georgetown University School of Medicine, Washington, DC, USA