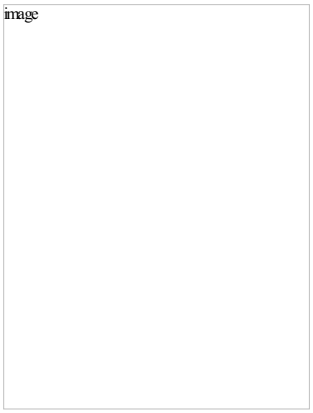
Lessons Learned from Bone Marrow Failure in Systemic Lupus Erythematosus: Case Reports and Review of the Literature

Erik Anderson, Bhakti Shah, Anne Davidson, Richard Furie



PII: S0049-0172(17)30383-9
DOI: https://doi.org/10.1016/j.semarthrit.2017.12.004
Reference: YSARH51282

To appear in: *Seminars in Arthritis and Rheumatism*

Cite this article as: Erik Anderson, Bhakti Shah, Anne Davidson and Richard Furie, Lessons Learned from Bone Marrow Failure in Systemic Lupus Erythematosus: Case Reports and Review of the Literature, *Seminars in Arthritis and Rheumatism,*doi:10.1016/j.semarthrit.2017.12.004

Title: Lessons Learned from Bone Marrow Failure in Systemic Lupus Erythematosus: Case Reports and Review of the Literature

Authors: Erik Anderson MD, Bhakti Shah MD, Anne Davidson MBBS, Richard Furie MD

Department/Institution Affiliation: Division of Rheumatology, Northwell Health, 865 Northern Boulevard, Great Neck, New York, 11021, United States of America

Keywords: aplastic anemia; autoimmune myelofibrosis; bone marrow failure; paroxysmal nocturnal hemoglobinuria; systemic lupus erythematosus

E-mail: Erik Anderson, eanderson22@northwell.edu; Bhakti Shah, Bshah4@northwell.edu; Anne Davidson, adavidson1@northwell.edu; Richard Furie, RFurie@northwell.edu

Corresponding Author: Erik Anderson MD
Mailing Address: Feinstein Institute for Medical Research, Clinical Research Service Room 1139, 350 Community Drive, Manhasset, New York 11030-3816
Telephone: (516)562-3641
E-mail: eanderson22@northwell.edu

1.1 Introduction

Systemic lupus erythematosus (SLE) is the prototypic systemic autoimmune disease with variable multisystem involvement. Hematologic abnormalities involving multiple cell lineages are common in SLE. Lymphopenia was present in 75% among a large cohort of SLE patients and occurred more frequently than any other criterion used to classify the disease.[1] Anemia is estimated to occur in about 50% of patients.[2] While anemia of chronic disease and iron deficiency are quite common, autoantibody- or complement-mediated autoimmune hemolytic anemia are occasionally encountered.[3] Autoimmune thrombocytopenia can result in severe thrombocytopenia and bleeding diatheses. Another less common cause of thrombocytopenia is thrombotic microangiopathy, a complication that also leads to a Coombs' negative hemolytic anemia owing to microvascular thrombosis.[4] Finally, neutrophil abnormalities may include neutropenia, excessive NETosis, and the appearance in the peripheral blood of a population of low density granulocytes.[5]

Whereas cytopenias in SLE are typically secondary to peripheral destruction, acquired bone marrow failure is a rarely reported mechanism of cytopenias in SLE. Myelodysplastic syndrome,[6] aplastic anemia (AA),[7] paroxysmal nocturnal hemoglobinuria (PNH),[8] autoimmune myelofibrosis (AIMF),[9] pure red cell aplasia[10] and hemophagocytosis[11] have all been described in SLE. Therefore, it is essential to determine the cause of the hematologic abnormality in the patient with SLE as treatments may vary dependent upon the underlying pathology.

In this review, we present two cases of AIMF in SLE as well as two cases of AA, one with features of PNH. We conducted a review of the current literature with the goal of more clearly characterizing cases of AIMF, AA and PNH associated with SLE. Specifically, we collected information regarding SLE serologies, hematologic manifestations, bone marrow findings, treatment, and outcomes. We discuss current theories on the pathophysiology of AIMF, AA and PNH in SLE and the challenge inherent in differentiating them from primary hematological disorders. We suggest diagnostic and therapeutic strategies for these disorders in SLE based on lessons learned from the present and previous cases.