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Introductory Chapter: Recent Advances in Systemic Lupus Erythematosus

Sophia Lionaki and Ioannis Boletis

1. Introduction

Systemic lupus erythematosus is the prototype of autoimmune disorder, which is typically manifesting in multiple organ systems and running a chronic course, affecting primarily females. It is associated with significant public health impact in affected individuals, with highly heterogeneous presentation and progression. During the last two decades, substantial progress has been made in our knowledge on systemic lupus erythematosus incidence, pathogenesis, therapeutic interventions, and long-term outcomes. However, it remains a challenging area of research, especially considering the genetic, epigenetic, and environmental factors that have been found to play a crucial role in disease prognosis [1]. Estimates on the worldwide incidence and prevalence of lupus revealed that the highest incidence was found in North America, while the lowest rates have been reported in Northern Australia [2]. Interestingly, a registry from the island of Crete reported that the overall age-adjusted/sex-adjusted incidence is growing among males [3]. Factors such as age, gender, ethnicity, genetics, hormonal status and environmental factors appear to have a central function in the development of the disease [2].

2. The new era of systemic lupus erythematosus

Despite the significant improvements which have been achieved in the field of lupus, including the overall management and immunosuppressive agents used for therapy, mortality rates of affected patients remain three times higher than those in the general population [2]. In particular, patients with lupus nephritis, who end up in end-stage kidney disease, incur a huge burden of morbidity, related not only to the dialysis procedure but also to the inflammatory background, the impact of cumulative immunosuppression, and the phenomenon of accelerated atherosclerosis which results in cardiovascular death [4]. Still the performance of each patient is variable. For instance, neurologic and psychiatric manifestations of systemic lupus erythematosus appear to have an increasing rate in recent reports although they are found in different frequencies across lupus cohorts, depending on the methodology used to define the related signs/symptoms and the screening practice [5]. Likewise, family planning becomes a crucial problem for women with systemic lupus erythematosus, considering the fact that females of reproductive age are the most frequently affected patients. Pregnancies in patients with active lupus and especially in those with renal involvement have been associated with significant morbidity and mortality for both the mother and the fetus [6]. Moreover, the interplay between

environmental factors and the genetic profile of each individual appear to be of great importance with respect to the onset and the progression of this disorder [7, 8]. Given these circumstances, we consider systemic lupus erythematosus a challenging field of research, which enquires continuing updating in order to illustrate all current knowledge regarding disease pathogenesis and provide guidelines for clinical practice employing all newer immunosuppressive agents.

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