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Editorial

A special European Journal of Rheumatology issue on systemic sclerosis: What and why?

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Recently, the pace of new knowledge on systemic sclerosis (SSc) has been overtaking most other immune-mediated inflammatory diseases (IMIDs). SSc is now mentioned in the pipeline of most of the major pharmaceutical industries, and several biotechnology companies have targeted SSc as one of the main connective tissue diseases (CTDs). This is a dramatic change from the last decade when only a few clinical trials were conducted on SSc.

In previous years, the breadth of knowledge acquired on SSc ranged from the fundamental mechanisms of the disease to the importance of clinical domains in contributing to patient outcomes in clinical management and clinical trials. Thus, it is becoming challenging to follow the distinct aspects in the literature.

To provide up-to-date knowledge on SSc, we collaborated with the European Journal of Rheumatology to publish this comprehensive but succinct special issue. The key opinion leaders worldwide were invited to review most of the aspects that are rapidly changing in SSc.

A hallmark and a major distinct aspect of SSc compared with other IMIDs is the fibrosis of tissues and vessels. Perhaps as the most "connective tissue" centered CTD, the pathogenesis of SSc goes beyond autoimmunity and includes a deep change in the functions of endothelial and mesenchymal cells. This issue includes articles on endothelial cells and their progenitors (1), epigenetics (2), and mesenchymal transition (3) that are deeply interlinked, giving a complementary view on this peculiar aspect of SSc.

As we move closer to the clinic, we are re-learning how to characterize SSc and raising the awareness about SSc being a family of conditions. This notion challenges the historical classification of SSc (4); it is supported by the knowledge acquired from the multi-omic studies in the previous decade (5) and the heterogeneity in the clinical relevance of biomarkers (6). The emerging role of autoantibodies (7) and microbiome (8) adds to this complexity.

As a result of this ever-changing horizon, we also reconsider how to analyze the standard clinical approaches from the way we assess the severity the Raynaud's phenomenon (9) or the cardiovascular involvement (10) or progressively earlier screening of pulmonary artery hypertension (11).

We could not issue an SSc-focused supplement without reviewing the learning points of our therapeutic approaches. Therefore, we offer 2 complete, balanced, and informative reviews on the use of the most classic immunosuppressive agent (12, 13) and the most recent successes and failures in clinical trials (14).

While co-editing this issue, we were convinced that it will serve a great purpose for the readers by providing a detailed and balanced review of the most crucial and debated aspects of SSc in 2020.

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