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Sjögren's disease, not syndrome

To the Editor:

Sjögren's syndrome should henceforth be known as Sjögren's disease. Our call for this change is based on the precedent for such name changes in rheumatology, as well as important differences between syndrome and disease. In addition, the struggle faced by Sjögren's patients to gain recognition for this serious autoimmune disease is difficult when some describe it as a collection of nuisance symptoms.

Sjögren's is a multisystemic disease that is characterized by its targeting of the salivary and lacrimal glands, leading to impaired secretion of saliva and tears and occasional salivary gland enlargement or recurrent sialadenitis. In 1933, the Swedish ophthalmologist Henrik Sjögren published an analysis of 19 patients with a dry eye disease that he termed "keratoconjunctivitis sicca" (1). The disease had been recognized earlier, but the eponym was earned based on Sjögren's comprehensive description and continued study of the disease throughout his lifetime.

Sjögren's is a distinct autoimmune disease with characteristic autoantibodies, glandular histopathology, and a pattern of systemic involvement. Akin to other systemic rheumatic diseases, it has defined genetic susceptibility traits and pathogenetic pathways. Accurate diagnosis using protocol-driven labial salivary gland biopsy and interpretation, as well as measures of dry eye and salivary hypofunction, is essential in differentiating this disease from dryness symptoms present in individuals who do not have an underlying autoimmune disease. Accurate diagnosis is also a prerequisite for the development of targeted disease-modifying therapies.

It is against this background that we call for the abandonment of "Sjögren's syndrome" in favor of "Sjögren's disease," or simply "Sjögren's." Are the differences between "syndrome" and "disease" sufficient to warrant this name change? We believe that they are. A syndrome denotes an aggregate of symptoms and signs that are associated with a morbid process, independent of pathogenesis (2,3). "Flu-like syndrome" is a good example. Mikulicz disease (4) and sicca syndromes (5) have been used synonymously with Sjögren's in the past, but each is now recognized as having a broad differential diagnosis. As the understanding of the etiology and/or pathogenesis of a particular condition improves, the term "syndrome" is replaced by "disease." In rheumatology, an example is Kawasaki syndrome, which is now properly known as Kawasaki disease (KD) (2).

We appreciate the counterargument that rheumatic diseases are often heterogeneous and specific etiologies may be identified in the future for some subsets. This has been exemplified by the recent identification of monogenic causes for variants of polyarteritis nodosa (6) and relapsing polychondritis (7), as well as the recognition of coronavirus disease 2019 as a potential proximate cause of KD (8).

However, the adoption of "Sjögren's disease" in lieu of "Sjögren's syndrome" would solidify the concept that it is a disease for which targeted therapies are being actively developed. For those affected, it also emphasizes that this is a distinct entity deserving of accurate diagnosis, careful study, and comprehensive management.

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