REVIEW

Standard classification and pathophysiology of rosacea: The 2017 update by the National Rosacea Society Expert Committee

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In 2002, the National Rosacea Society assembled an expert committee to develop the first standard classification of rosacea. This original classification was intended to be updated as scientific knowledge and clinical experience increased. Over the last 15 years, significant new insights into rosacea's pathogenesis and pathophysiology have emerged, and the disorder is now widely addressed in clinical practice. Growing knowledge of rosacea's pathophysiology has established that a consistent multivariate disease process underlies the various clinical manifestations of this disorder, and the clinical significance of each of these elements is increasing as more is understood. This review proposes an updated standard classification of rosacea that is based on phenotypes linked to our increased understanding of disease pathophysiology. This updated classification is intended to provide clearer parameters to conduct investigations, guide diagnosis, and improve treatment. (J Am Acad Dermatol http://dx.doi.org/10.1016/j.jaad.2017.08.037.)

Key words: comorbidity; erythema; ocular; papules; pathophysiology; phenotypes; phymas; pustules; rosacea; telangiectasia.

Rosacea is well established as a chronic cutaneous syndrome, encompassing various combinations of potential signs and symptoms that manifest primarily on the convexities of the central face (cheeks, chin, nose, and central part of forehead) and are often characterized by repeated remissions and exacerbations. Rosacea is commonly diagnosed in certain demographic groups, and in epidemiologic studies of whites, its prevalence has been 10% or higher.[1-4] Although it has been most frequently observed in patients with fair skin, rosacea has also been diagnosed in Asians, Latin Americans, African-Americans, and Africans (Figs 1 to 4).[5-8] The disorder is identified more often in women than in men, and although it may occur at any age, the onset typically occurs at any time after age 30.[1,2]

Abbreviations used:

NRS: National Rosacea Society
RosaQoL: Rosacea Quality of Life Index

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In 2002, the National Rosacea Society (NRS) assembled an expert committee to develop a classification system to provide standard criteria essential for performing research, analyzing results, and comparing data from different sources, as well as to serve as a diagnostic reference in clinical practice.[9] The classification system also established standard terminology to facilitate clear communication among a broad range of basic, clinical, and other researchers; practicing dermatologists; primary care physicians; ophthalmologists; other medical specialists; regulatory authorities; health and insurance administrators; patients; and the general public. Because the etiology and pathogenesis of rosacea were unknown at that time and there were no histologic or serologic markers for the disease, the system was based on morphologic characteristics alone to provide a framework that could be readily updated as new discoveries were made. The standard classification was proposed as a provisional system, and it was hoped that with increased scientific knowledge and experience in clinical practice, the definition of rosacea would ultimately be based on pathophysiology.

CAPSULE SUMMARY

- In 2002, the National Rosacea Society assembled an expert committee to develop the first standard classification of rosacea.
- New scientific investigations have yielded significant insights into rosacea's pathogenesis and pathophysiology.
- The current update provides clearer parameters to conduct investigations while offering a more meaningful guide to diagnosis and treatment.

Over the 15 years since the classification's development, scientific investigations have yielded significant insight into rosacea's pathogenesis and pathophysiology, whereas clinical experience has suggested a need for an updated approach to diagnosis, classification, and treatment.[10,11] To fulfill the directive of the original authors of the 2002 classification system, the NRS convened a committee to update the standard classification of rosacea, including new parameters for its diagnosis, assessment, and common presentations. The revised standard diagnostic features were adopted on the basis of their positive predictive value as supported by rosacea experts.[12] In addition, new scientific evidence has shown that rosacea's diverse features may be part of a continuum of inflammation that may not be clinically visible but is detectable histologically and biochemically.[13-16] This suggests that the common presentations of flushing and fixed centrofacial erythema may progress to include papules and pustules and potentially lead (in a small proportion of cases) to subsequent development of phymas.[17,18] This is further supported by the observation that in patients who have been treated successfully for their papules and pustules, the erythema and flushing often remain.[18]

The following updates have been developed by the committee and reviewed by a panel of 21rosacea experts worldwide and are intended tobe useful to clinicians and researchers byproviding clearer and more meaningful parameters to conduct new investigations, as well as to build on existing findings while offering a more meaningful guide to diagnosis and treatment.

DIAGNOSTIC CRITERIA

The original standard classification of rosacea identified the most common patterns or groupings of signs and symptoms and designated them as follows: subtype 1, erythematotelangiectatic; subtype 2, papulopustular; subtype 3, phymatous; and subtype 4, ocular. Although didactically successful, the subtype designations were widely utilized individually and construed as distinct disorders, ignoring the frequent simultaneous occurrence of more than 1 subtype and the potential progression from one subtype to another.[18] Moreover, the focus on subtypes tended to limit consideration of the full range of potential signs and symptoms that may occur in individual patients and that, in some cases, may confound the assessment of severity.

Because rosacea can encompass a multitude of possible combinations of signs and symptoms, the following updated classification system is based on phenotypes—observable characteristics that can result from genetic and/or environmental influences—to provide the necessary means of assessing and treating rosacea in a manner that is consistent with each individual patient's experience (Table I). The phenotypes and diagnostic criteria are largely in agreement with those recommended by the global rosacea consensus panel in 2016, and at least 1 diagnostic or 2 major phenotypes are required for the diagnosis of rosacea.[12]

Diagnostic phenotypes

A diagnosis of rosacea may be considered in the presence of 1 of the following diagnostic cutaneous signs.[12]