SPECIAL REPORT

Standard classification of rosacea: Report of the National Rosacea Society Expert Committee on the Classification and Staging of Rosacea

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Rosacea is well recognized as a chronic cutaneous disorder primarily of the convexities of the central face (cheeks, chin, nose, and central forehead), often characterized by remissions and exacerbations. Based on present knowledge, it is considered a syndrome, or typology, encompassing various combinations of such cutaneous signs as flushing, erythema, telangiectasia, edema, papules, pustules, ocular lesions, and rhinophyma.[1] In most cases, some rather than all of these stigmata appear in any given patient.

Rosacea appears to be quite common, and in an epidemiologic study in Sweden its prevalence was 10%.[2] It has been most frequently observed in patients with fair skin, but has also been diagnosed in Asians and African Americans. Rosacea occurs in both men and women and, although it may occur at any age, the onset typically begins at any time after age 30.[3]

Despite its apparent high incidence, the nosology of rosacea is not well established, and the term "rosacea" has been applied to patients and research subjects with a diverse set of clinical findings that may or may not be an integral part of this disorder. In addition to the diversity of clinical manifestations, the etiology and pathogenesis of rosacea are unknown, and there are no histologic or serologic markers.

Therefore, the National Rosacea Society assembled a committee to develop a standard classification system that can serve as a diagnostic instrument to investigate the manifestations and relationships of the several subtypes and potential variants of rosacea. Standard criteria for diagnosis and classification of patients are essential to perform research, analyze results and compare data from different sources, and may further serve as a diagnostic reference in clinical practice. The standard terminology will also facilitate clear communication among a broad range of basic, clinical, and other researchers; practicing dermatologists, primary care physicians, ophthalmologists and other specialists; health and insurance administrators; and patients and the general public.

The committee based the standard classification system on present scientific knowledge and morphologic characteristics. This avoids assumptions on pathogenesis and progression, and provides a framework that can be readily updated and expanded as new discoveries are made. As knowledge increases, it is hoped that the definition of rosacea may ultimately be based on causality, rather than on morphology alone.

The following provisional classification system describes the primary features of rosacea and defines 4 subtypes and 1 variant. Evolution from one subtype to another may or may not occur, and research to investigate this process may provide important insight into the pathogenesis of rosacea. Regardless of subtype, however, each individual characteristic may progress from mild to moderate to severe. Early diagnosis and treatment are therefore recommended.

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The opinions set forth in this report are those of the committee members and do not represent the Food and Drug Administration in any way.

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DIAGNOSTIC CRITERIA

Primary features

Rosacea typically affects the convexities of the central face. The presence of one or more of the following signs with a central face distribution is indicative of rosacea. These signs are commonly transient, and each may occur independently. Many patients may present with more than one of these diagnostic features.

- Flushing (transient erythema). A history of frequent blushing or flushing is common.
- Nontransient erythema. Persistent redness of the facial skin is the most common sign of rosacea.
- Papules and pustules. Dome-shaped red papules with or without accompanying pustules, often in crops, are typical. Nodules may also occur. Although patients with concomitant acne may exhibit comedones, comedones should be considered part of an acne process unrelated to rosacea.
- Telangiectasia. Telangiectases are common but not necessary for a rosacea diagnosis.

Secondary features

The following signs and symptoms often appear with one or more of the primary features of rosacea, but in some patients can occur independently.

- Burning or stinging. Burning or stinging sensations with or without scaling or dermatitis may occur, especially on malar skin.[4]
- Plaque. Elevated red plaques without epidermal changes in the surrounding skin may occur.
- Dry appearance. Central facial skin may be rough and scaling so as to resemble dry skin and suggest an eczematous dermatitis, and may often include the coexistence of seborrheic dermatitis. This "dryness" may be associated with burning or stinging sensations, and may be caused by irritation rather than the disease process.
- Edema. Edema may accompany or follow prolonged facial erythema or flushing. Sometimes soft edema may last for days or be aggravated by inflammatory changes. Solid facial edema (persisting hard, nonpitting edema) can occur with rosacea, usually as a sequel of the papulopustular type, and also independently of redness, papules and pustules, or phymatous changes.
- Ocular manifestations. Ocular manifestations are common, and range from symptoms of burning or itching to signs of conjunctival hyperemia and lid inflammation. Styes, chalazia, and corneal damage may occur in many patients with rosacea in addition to cutaneous stigmata. The severity of ocular manifestations may not be proportional to those of the skin.

Table I. Guidelines for the diagnosis of rosacea

Presence of one or more of the following primary features:
Flushing (transient erythema)
Nontransient erythema
Papules and pustules
Telangiectasia
May include one or more of the following secondary features:
Burning or stinging
Plaque
Dry appearance
Edema
Ocular manifestations
Peripheral location
Phymatous changes

- Peripheral location. Rosacea has been reported to occur in other locations,[5] but the frequency and occurrence of this are ill-defined. Rosacea in peripheral locations may or may not be accompanied by facial manifestations.
- Phymatous changes. These can include patulous follicles, skin thickening or fibrosis, and a bulbous appearance. Rhinophyma is the most common form, but other phymas may occur (Table I).

SUBTYPES

The primary and secondary rosacea features described above often occur together. The most common patterns or groupings of signs are provisionally designated as specific subtypes of rosacea and are described here (Table II). Each subtype includes the fewest signs sufficient to make a diagnosis of the subtype (though not necessarily limited to these), and patients may have characteristics of more than one rosacea subtype at the same time.

Subtype 1: Erythematotelangiectatic rosacea

Erythematotelangiectatic rosacea is mainly characterized by flushing and persistent central facial erythema. The appearance of telangiectases is common but not essential for a diagnosis of this subtype. Central facial edema, stinging and burning sensations, and roughness or scaling may also be reported. A history of flushing alone is common among patients presenting with erythematotelangiectatic rosacea.

Subtype 2: Papulopustular rosacea

Papulopustular rosacea is characterized by persistent central facial erythema with transient papules or pustules or both in a central facial distribution. However, papules and pustules also may occur perioriificially (that is, they may occur in the perioral, perinasal, or periocular areas). The papulopustular

Table II. Subtypes and variants of rosacea and their characteristics

	Characteristics
Subtype	
Erythematotelangiectatic	Flushing and persistent central facial erythema with or without telangiectasia.
Papulopustular	Persistent central facial erythema with transient, central facial papules or pustules or both.
Phymatous	Thickening skin, irregular surface nodularities and enlargement. May occur on the nose, chin, forehead, cheeks, or ears.
Ocular	Foreign body sensation in the eye, burning or stinging, dryness, itching, ocular photosensitivity, blurred vision, telangiectasia of the sclera or other parts of the eye, or periorbital edema.
Variant	
Granulomatous	Noninflammatory; hard; brown, yellow, or red cutaneous papules; or nodules of uniform size.

subtype resembles acne vulgaris, except that comedones are absent. Rosacea and acne may occur concomitantly, and such patients may have comedones as well as the papules and pustules of rosacea. Burning and stinging sensations may be reported by patients with papulopustular rosacea.

This subtype has often been seen after or in combination with subtype 1, including the presence of telangiectases. The telangiectases may be obscured by persistent erythema, papules, or pustules, and tend to become more visible after successful treatment of these masking components.

Subtype 3: Phymatous rosacea

Phymatous rosacea includes thickening skin, irregular surface nodularities, and enlargement. Rhinophyma is the most common presentation, but phymatous rosacea may occur in other locations, including the chin, forehead, cheeks, and ears. Patients with this subtype also may have patulous, expressive follicles in the phymatous area, and telangiectases may be present.

This subtype has frequently been observed after or in combination with subtypes 1 or 2, including persistent erythema, telangiectases, papules, and pustules. In the case of rhinophyma, these additional stigmata may be especially pronounced in the nasal area.

Subtype 4: Ocular rosacea

The diagnosis of ocular rosacea should be considered when a patient's eyes have one or more of the following signs and symptoms: watery or bloodshot appearance (interpalpebral conjunctival hyperemia), foreign body sensation, burning or stinging, dryness, itching, light sensitivity, blurred vision, telangiectases of the conjunctiva and lid margin, or lid and periocular erythema. Blepharitis, conjunctivitis, and irregularity of the eyelid margins also may occur.[6] Meibomian gland dysfunction presenting as chalazion or chronic staphylococcal infection as manifested by hordeolum (stye) are common signs of rosacea-related ocular disease. Some patients may have decreased visual acuity caused by corneal complications (punctate keratitis, corneal infiltrates/ulcers, or marginal keratitis).[7] Treatment of cutaneous rosacea alone may be inadequate in terms of lessening the risk of vision loss resulting from ocular rosacea, and an ophthalmologic approach may be needed.[8]

Ocular rosacea is most frequently diagnosed when cutaneous signs and symptoms of rosacea are also present. However, skin signs and symptoms are not prerequisite to the diagnosis, and limited studies suggest that ocular signs and symptoms may occur before cutaneous manifestations in up to 20% of patients with ocular rosacea. Approximately half of these patients experience skin lesions first, and a minority have both manifestations simultaneously.[9]

VARIANTS

Variants of rosacea, which do not represent morphologic patterns or combinations as seen in rosacea subtypes, may occur. To date, the committee has recognized one such variant.

Granulomatous rosacea

Granulomatous rosacea is characterized by hard, yellow, brown, or red cutaneous papules or nodules that may be severe and lead to scarring. These lesions tend to be less inflammatory than papules and pustules and sit upon relatively normal-appearing skin. They can vary in size among patients but are monomorphic in each individual patient, and typically appear on the cheeks and periorificial areas. Granulomatous rosacea may occur in locations other than those in which the phymas are observed. The presence of other rosacea signs is not needed for a diagnosis of the granulomatous rosacea variant.

EXCLUSIONS

The committee noted that certain disorders may have been prematurely identified as associated with rosacea or as a variant of rosacea, and for clarity should be recognized at this time as separate entities. There is insufficient basis at present to include the following conditions as types of rosacea.

Rosacea fulminans

Popularly known as pyoderma faciale, the grouping of this disorder as a type of rosacea is premature. It is characterized by the sudden appearance of papules, pustules, and nodules, along with fluctuating and draining sinuses that may be interconnecting. The condition appears primarily in women in their 20s, and intense redness and edema also may be prominent.

Steroid-induced acneiform eruption

Steroid-induced acneiform eruption is not a variant of rosacea and can occur as an inflammatory response in any patient during or after chronic corticosteroid use. The same inflammatory response may also, of course, occur in patients with rosacea.

Perioral dermatitis

Although rosacea papules may appear in the perioral area, as noted earlier, perioral dermatitis without rosacea symptoms cannot be classified as a variant of rosacea. Perioral dermatitis is characterized by such stigmata as microvesicles, scaling, and peeling.

FUTURE

This investigational instrument is intended to set the stage for a better understanding of rosacea and its subtypes among researchers and practitioners by fostering communication and facilitating the development of a research-based classification system. As a provisional standard classification system, it is likely to require modification in the future as the pathogenesis and subtypes of rosacea become clearer, and as its relevance and applicability are tested by investigators and clinicians. The committee welcomes reports on the usefulness and limitations of these criteria.

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The National Rosacea Society is a 501(c)(3) nonprofit organization whose mission is to support rosacea research, including the awarding of research grants, and to provide educational information on rosacea to physicians, patients, and the public. Reports or inquiries should be directed to the National Rosacea Society, 800 S Northwest Hwy, Suite 200, Barrington, IL 60010; telephone 847/382-8971; E-mail: rosaceas@aol.com.

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