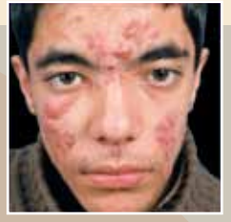
PART I

Disorders Presenting in the Skin and Mucous Membranes

SECTION 1

Disorders of Sebaceous and Apocrine Glands



Acne Vulgaris (Common Acne) and Custic Acne ■ ○ ➔ ◑

- An inflammation of pilosebaceous units, very common
- Appears in certain body areas (face, trunk, rarely buttocks)
- Most frequently in adolescents
- Manifests as comedones, papulopustules, nodules, and cysts
- Results in pitted, depressed, or hypertrophic scars

ICD-9:706.1 ◦ ICD-10:L70.0

Epidemiology

Occurrence. Very common, affecting approximately 85% of young people.
Age of Onset. Puberty—10 to 17 years in females, 14 to 19 in males; however, may appear first at 25 years or older.
Sex. More severe in males than in females.
Race. Lower incidence in Asians and Africans.
Genetic Aspects. Multifactorial genetic background. Familial predisposition: majority of individuals with cystic acne have parent(s) with a history of severe acne. Severe acne may be associated with XYY syndrome.

Pathogenesis

Key factors are follicular keratinization, androgens, and *Propionibacterium acnes* (Image 1-1).

Acne results from a change in the keratinization pattern in the pilosebaceous unit, with the keratinous material becoming more dense and blocking secretion of sebum. These keratin plugs are called comedones and represent the "time bombs" of acne. Linoleic acid, which regulates keratinocyte proliferation, is decreased in acne. Comedonal plugging and a complex interaction between androgens and bacteria (P. acnes) in the plugged pilosebaceous units lead to inflammation. Androgens (qualitatively and quantitatively normal in the serum) stimulate sebaceous glands to produce larger amounts of sebum. Bacteria contain lipase, which converts lipid into fatty acids, and produce proinflammatory mediators, [interleukin 1, tumor necrosis factor TNF]. Fatty acids and proinflammatory mediators cause a sterile inflammatory response to the pilosebaceous unit. The distended follicle walls break, and the contents (sebum, lipids, fatty acids, keratin, bacteria) enter the dermis, provoking an inflammatory and foreign-body response (papule, pustule, nodule). Rupture plus intense inflammation lead to scars.

Contributory Factors. Acnegenic mineral oils, rarely dioxin and others.
Drugs. Lithium, hydantoin, isoniazid, glucocorticoids, oral contraceptives, iodides, bromides and androgens (e.g., testosterone), danazol.
Others. *Emotional stress* can definitely cause exacerbations. *Occlusion* and *pressure* on the skin, such as by leaning face on hands, very important and often unrecognized exacerbating factor (*acne mechanica*). Acne is not caused by chocolate or fatty foods or, in fact, by any kind of food.

Clinical Manifestation

Duration of Lesions. Weeks to months.
Season. Often worse in fall and winter.
Symptoms. Pain in lesions (especially nodulocystic type).
Skin Lesions. *Comedones*—open (blackheads) or closed (whiteheads); *comedonal acne* (Fig. 1-1). *Papules* and *papulopustules*—i.e., a papule topped by a pustule; *papulopustular acne* (Fig. 1-2). *Nodules* or *cysts*—1–4 cm in diameter (Fig.

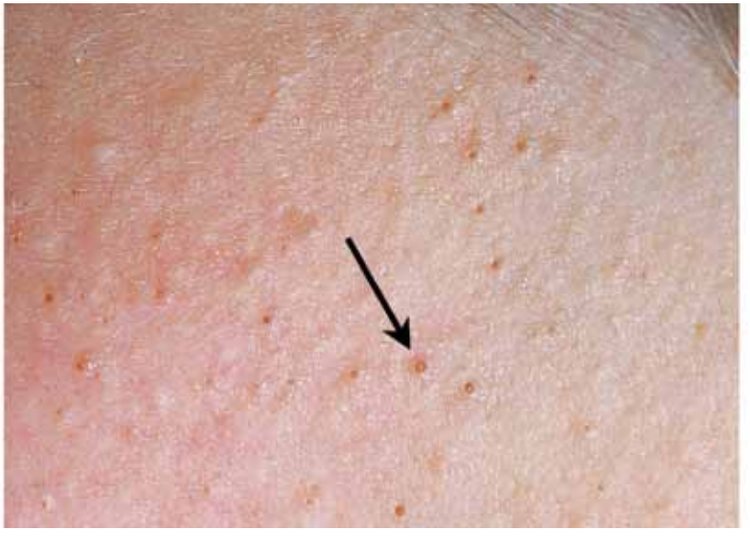


Figure 1-1. Acne vulgaris: comedones Comedones are keratin plugs that form within follicular ostia, frequently associated with surrounding erythema and pustule formation. Comedones associated with small ostia are referred to as closed comedones or "white heads"; those associated with large ostia are referred to as open comedones or "black heads." Comedones are best treated with topical retinoids.

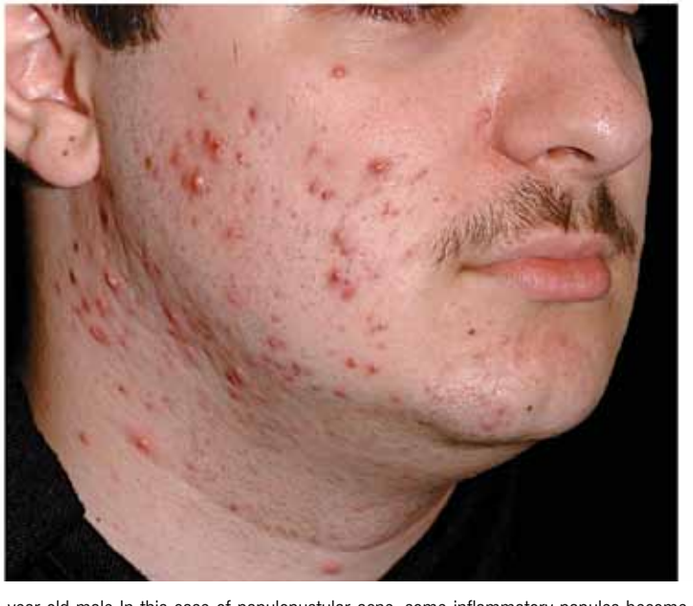


Figure 1-2. 20-year-old male In this case of papulopustular acne, some inflammatory papules become nodular and thus represent early stages of nodulocystic acne.

1-3); nodulocystic acne. Soft nodules result from repeated follicular ruptures and reencapsulations with inflammation, abscess formation, and foreign-body reaction. Cysts are actually pseudocysts as they are not lined by epithelium but represent fluctuating abscesses (Image 1-1). Round isolated single nodules and cysts coalesce to linear mounds and sinus tracts (Fig. 1-4). Sinuses: draining epithelial-lined tracts, usually with nodular acne. Scars: atrophic depressed (often pitted) or hypertrophic (at times, keloidal). Seborrhea of the face and scalp often present and sometimes severe. For more clinical pictures, see acne picture gallery on online version.

Sites of Predilection Face, neck, trunk, upper arms, buttocks.

Special Forms

Acne Conglobata. Severe cystic acne (Figs. 1-4 and 1-5) with more involvement of the trunk than the face. Coalescing nodules, cysts, abscesses, and ulceration; occurs also on buttocks. Spontaneous remission is long delayed. Rarely, acne conglobata seen in XYY genotype (tall males, slightly mentally retarded, with aggressive behavior) or in the polycystic ovary syndrome.

Acne Fulminans. Teenage boys (ages 13 to 17). Acute onset, severe cystic acne with concomitant suppuration and always ulceration; also present are malaise, fatigue, fever, generalized arthralgias, leukocytosis, and elevated erythrocyte sedimentation rate.

SAPHO Syndrome. Synovitis, acne, acne fulminans, palmoplantar pustulosis, hidradenitis suppurativa, hyperostosis, and osteitis. Rare.

PAPA Syndrome. Sterile pyogenic arthritis, pyoderma gangrenosum acne. An inherited autoinflammatory disorder; very rare.

Tropical Acne. Flare of acne, usually with severe folliculitis, inflammatory nodules, and draining cysts on trunk and buttocks in tropical climates; secondary infection with Staphylococcus aureus.

Acne with Facial Edema. Associated with recalcitrant, disfiguring midline facial edema. Woody induration with and without erythema.

Acne in the Adult Woman. Persistent acne in an (often) hirsute female with or without irregular menses needs an evaluation for hyper-secretion of adrenal and ovarian androgens: total testosterone, free testosterone, and/or dehydroepiandrosterone sulfate (DHEAS) (e.g., in the polycystic ovary syndrome).

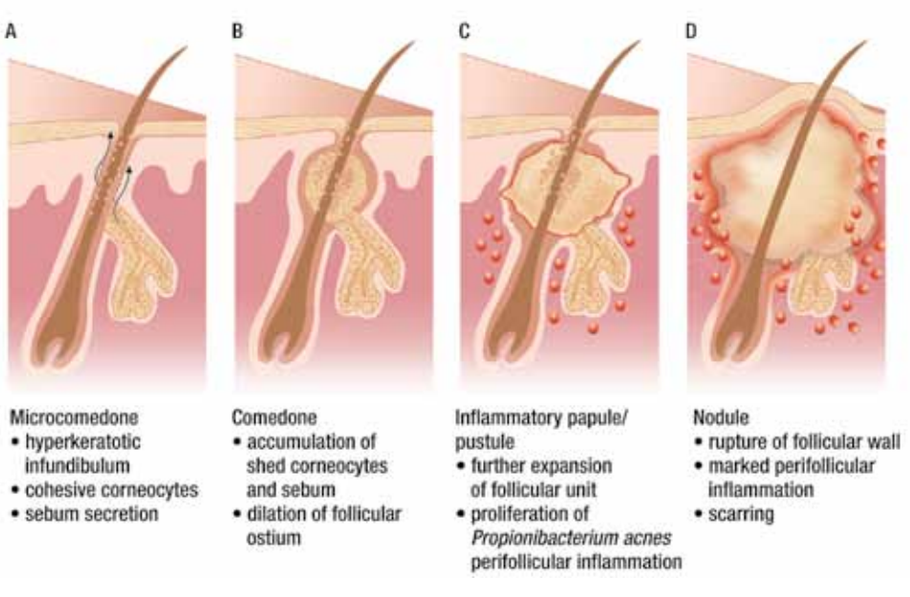


Image 1-1. Acne pathogenesis [From Zaenglein AL et al. Acne vulgaris and acneiform eruptions, in Wolff K et al (eds): *Fitzpatrick's Dermatology in General Medicine*, 7th ed. New York, McGraw-Hill, 2008.]

Recalcitrant Acne. Can be related to congenital adrenal hyperplasia (11- or 21- hydroxylase deficiencies).

Acne Excoriée. Mild acne, usually in young women, associated with extensive excoriations and scarring due to emotional and psychological problems (obsessive compulsive disorder).

Neonatal Acne. On nose and cheeks in newborns or infants, related to glandular development; transient.

Occupational Acne. Due to exposure to tar derivatives, cutting oils, chlorinated hydrocarbons (see "Chloracne," below). Large comedones, inflammatory papules and cysts; not restricted to predilection sites of acne but can appear on other (covered) body sites.

Chloracne. Due to exposure to chlorinated aromatic hydrocarbons in electrical conductors, insecticides, and herbicides. Sometimes very severe due to industrial accidents or intended poisoning (e.g., dioxin).

Acne Cosmetica. Due to comedogenic cosmetics.

Pomade Acne. On the forehead, usually in Africans applying pomade to hair.

Acne Mechanica. Flares of preexisting acne in face, because of leaning face on hands, or on forehead, from pressure of football helmet.

Acne-Like Conditions

Steroid Acne Following systemic or topical glucocorticoids. Monomorphous folliculitis—small erythematous papules and pustules without comedones.

Drug-Induced Acne Monomorphous acne-like eruption due to phenytoin, lithium, isoniazid, high-dose vitamin B complex, epidermal growth factor inhibitors (see Section 22), halogenated compounds. No comedones.

Acne Aestivalis Papular eruption after sun exposure ("Mallorca acne"). Usually on forehead, shoulders, arms, neck, and chest. No comedones. Pathogenesis unknown.

Gram-Negative Folliculitis Multiple tiny yellow pustules develop on top of acne vulgaris as a result of long-term antibiotic administration.

Diagnosis and Differential Diagnosis

Note: Comedones are required for diagnosis of any type of acne. Comedones are not a feature of acne-like conditions (above) and of the conditions listed below.

Face S. aureus folliculitis, pseudofolliculitis barbae, rosacea, perioral dermatitis.

Trunk Malassezia folliculitis, "hot-tub" pseudomonas folliculitis, S. aureus folliculitis, and acne-like conditions (see above).

Laboratory Examination

No laboratory examinations required. If there is suspicion of an endocrine disorder, free testosterone, follicle-stimulating hormone, luteinizing hormone, and DHEAS should be determined to exclude hyperandrogenism and polycystic ovary syndrome. Note: In the overwhelming majority of acne patients, hormone levels are normal.

Laboratory examinations [transaminases (ALT, AST), triglycerides, and cholesterol levels] may be required if systemic isotretinoin treatment is planned (see below).

Course

Acne most often clears spontaneously by the early twenties but can persist to the fourth decade or older. Flares occur in the winter and with the onset of menses. The sequela is scarring (for clinical examples see, which should be avoided by proper treatment, especially with oral isotretinoin early in the course of the disease (see below).

Management

The psychological impact of acne (perceived cosmetic disfigurement) should be assessed individually in each patient and therapy modified accordingly. The goal of therapy is to remove the plugging of the pilar drainage, reduce sebum production, and treat bacterial colonization.

Mild Acne

- Topical antibiotics (clindamycin and erythromycin)
- Benzoyl peroxide gels (2%, 5%, or 10%)
- Topical retinoids (tretinoin, adapalene) require detailed instructions regarding gradual increases in concentration from 0.01% to 0.025% to 0.05% cream/gel or liquid. After improvement, medication is reduced to the lowest effective maintenance.

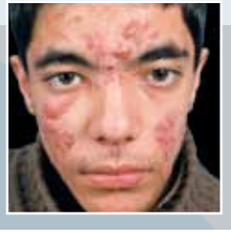
Improvement occurs over a period of months (2–5) but may take even longer for

PART II

Dermatology and Internal Medicine

SECTION 14

The Skin Signs in Immune, Autoimmune, and Rheumatic Disorders



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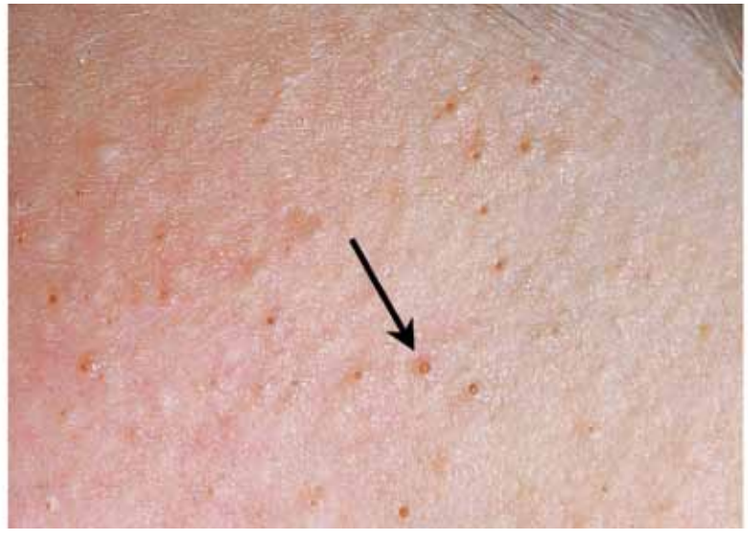

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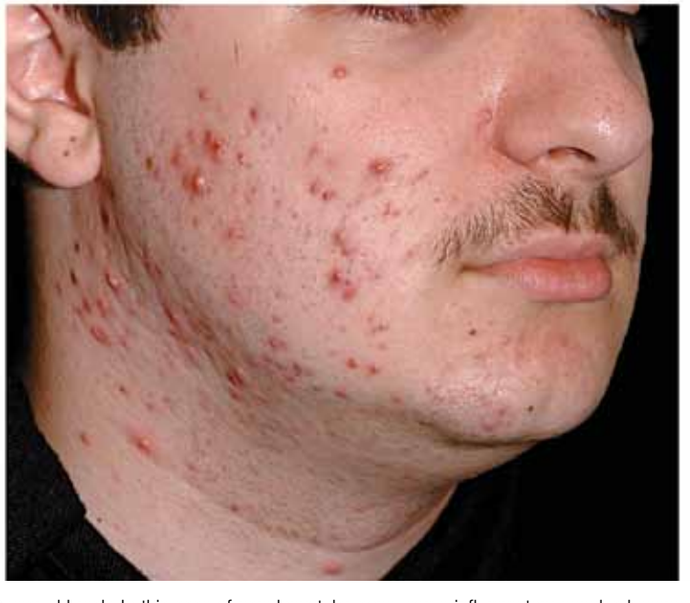

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PART III

Diseases Due to Microbial Agents

PART IV

Skin Signs of Hair, Nail, and Mucosal Disorders