CHAPTER 1
Introduction

Giuseppe Micali, Maria Rita Nasca, and Maria Letizia Musumeci
Department of Dermatology, University of Catania, Italy

Hidradenitis suppurativa (HS), also known as acne inversa, is a chronic, inflammatory, recurrent, debilitating skin disease of the hair follicle that generally appears after puberty (second and third decades). HS affects apocrine gland-bearing areas of the body, most commonly the axillae, inguinal, and anogenital regions, presenting with painful, deep-seated and inflamed nodules/abscesses, sinus tracts, purulent discharge, and scarring.

The term "hidradenitis" is derived from the Greek words ἱδρώς (hidrós = sweat) and ἀδήν (adén = gland) plus -ῖτις (-itis = inflammatory disease) [1]. Historically, HS has been considered a primary inflammatory disease of the apocrine glands. Its first clinical description dates back to 1839, when Alfred Armand Louis Marie Velpeau (1795–1867), an anatomist and surgeon from Paris, reported a suppurative disorder, which he named "phlegmon tubériforme" or "érysipélateux", characterized, respectively, by formation of indurated or painful abscesses occurring in the axillary, submammary, and perianal areas. Although he believed that this disorder was primarily the result of sebaceous follicles irritation precipitated by mechanical traumas and poor hygiene, fifteen years later, in 1854, one of his colleagues, also from Paris, Aristede Auguste Stanislas Verneuil (1823–1895), related the disease to the apocrine sweat glands and coined the term "hidrosadénite phlegmonuse" [2]. During the following decades, investigators paid relatively little attention to this neglected disorder. Nevertheless, its pathogenesis raised a controversial debate among different eminent clinicians, who denied its existence as a separate disease and favored the hypothesis of a link with "acne inversa", a disorder at that time thought to be acne vulgaris localized in the folds, often encompassing both diseases in a complex nosological framework along with "dissecting cellulitis of the scalp" [3]. Interestingly, in the same span of time the German philosopher Karl Marx (1818–1883) might have been suffering from HS, as he reported in his private correspondence between 1862 and 1874. In a letter to Friedrich Engels in June 1867 he wrote: "The bourgeoisie will remember my carbuncles until their dying day" [4]. In 1956, Pillsbury, Shelley, and Kligman published an article describing the disease's main characteristics, and coined the name "acne triad" to describe the condition characterized by the association of HS with acne conglobata and dissecting cellulitis of the scalp [5]. In 1975, Plewig and Kligman, following Pillsbury's proposal, added another element to acne triad, pilonidal sinus, modifying the "acne triad" in "acne tetrad" [6]. A key change to the previous point of view on pathogenesis was the definitive discard of the hypothesis of a primary apocrine sweat gland involvement. Actually, HS is essentially considered a disease triggered by follicular occlusion from keratinous material, with subsequent follicle rupture, secondary inflammation, superimposed infection, and destruction of skin appendages, including the apocrine glands. It has also been suggested that HS, rather than merely an acneiform disease, should rather be considered as a "dissecting terminal hair folliculitis" affecting exclusively fully developed hair follicles, especially in adults [7].

Over the years, HS has been named by different and sometimes inappropriate terms, which have not always achieved a common and universal acceptance in the scientific audience, including acne conglobata, apocrine acne, apocrinitis, pyoderma fistulans, fox-den disease, Verneuil's disease, and some other outdated denominations. Strictly speaking, HS should not be considered as a type of acne, as it shows no comedones and, unlike acne, it is the outcome of the involvement of the deep, rather than of the superficial, part of the follicle.

Establishing a validated glossary of terms allowing the best possible description of lesions observed in HS patients has also represented a major research target [8]. The classical dermatologic semantics do not fit some of the lesions observed in HS, and many of them cannot be straightforwardly assigned to a definite morphologic definition. This finding negatively impacts on the assessment of clinicopathological correlations and the objective evaluation of treatment response. For this reasons, many efforts are currently done to achieve a common glossary of terms useful for scientific communication and feedback in clinical research. Recently, a few new terms, so far not included in the classic dermatologic terminology, which may also be useful in
describing other skin diseases, such as “tunnel”, “cord”, and “bridge”, have been introduced [8].

A valid, accurate, as well as easy to use scoring system for the assessment of HS severity is also strictly needed. Although numerous different assessment tools have been proposed and described in the literature, no gold standard has yet been identified. Indeed, the ideal scoring system should be easy to use both in the clinical practice and in trial settings, as well as capable of providing evidence about worsening or treatment-related improvement. Since all the proposed clinical assessment tools share both advantages and limitations, the integration of parameters gained by objective instrumental examination, such as ultrasound, may be desirable in order to obtain a better classification of HS severity and to adequately monitor treatment response.

These are only some of the multiple unsolved challenges recently put forward by this troublesome and disabling disorder. Without intervention, the natural history of HS is chronic and progressive. Clinicians should take into account that HS is known to be associated with several and often undiagnosed comorbidities and burdened with many and various complications. Therefore, they should suggest appropriate screening and treatment. An early diagnosis is crucial to a correct management and to improve both physical and psychological impairments.

**References**

7. Chen W, Plewig G. Should hidradenitis suppurativa/acne inversa best be renamed as “dissecting terminal hair folliculitis”? Expl Dermatol 2016 [Epub ahead of print]