CASO REPORT

Follicular occlusion tetrad: Clinical case

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ABSTRACT

Follicular occlusion tetrad encompasses four clinical entities with similar pathophysiology, including suppurative hidradenitis, acne conglobata, dissecting cellulitis of the scalp and pilonodal sinus. Although the exact pathogenesis of this group of pathologies is unknown, all the evidences suggest that they share the same pathological process initiated by follicular occlusion in areas bearing apocrine glands. The case of a 22-year-old patient manifesting typical clinical characteristics is presented.

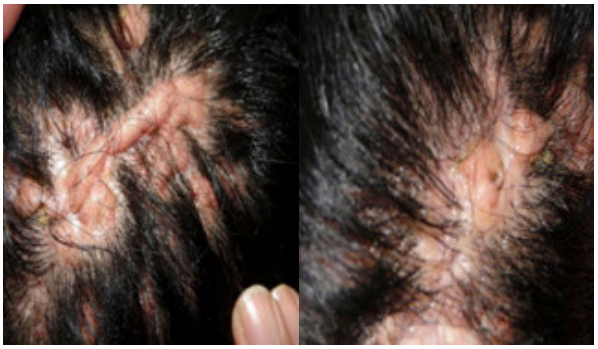
INTRODUCTION

Suppurative hidradenitis, acne conglobata, dissecting cellulitis of the scalp and pilonodal sinus integrate a set of inflammatory conditions of common clinical features and a similar pathogenic process taking place within terminal hair follicles. It is a rare disorder which commonly affects men ages 18 to 40 and black patients.[1]

Described in 1956 by Pillsbury, Kligman and Shelley, who argued that the main pathogenic event was the hyperkeratinization and dilation of the follicular walls with occlusion and corneal retention, as well as secondary bacterial infection, resulting in a chronic inflammatory process of the pilosebaceous duct, apocrine glands or both, along with suppuration, sinus and scarring.[2]

CLINICAL CASE

A 22-year-old male patient, with no hereditary family history, but a history of pilonodal cyst treated by proctology 2 years ago. No smoking and occasional alcohol intake. He presented with a 7-year-old clinical picture: dermatosis of the scalp, parieto-occipital region, nape of the neck, consisting of extensive alopecic plaques, keloid scars, and erythematous papules and nodules in the posterior cervical region (Figures 1, 2 and 3). On both buttocks, cysts of various sizes, hyperpigmented excavated scars (figure 4). On the back, cystic nodule lesions with necrotic areas (figure 5). General tests included complete blood count, liver profile test, blood sugar levels, dehydroepiandrosterone sulfate, free testosterone, androstenedione, within normal parameters, triglycerides 321 mg/dl, rest of normal lipid panel.



Figures 1 and 2. Scalp with alopecic plaques, keloid scars, on the vertex and active lesions on the nape of the neck.

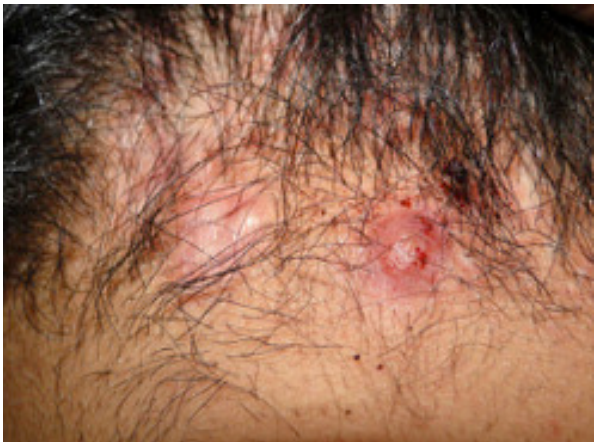


Figure 3 Erythematous papules and nodules on posterior cervical region.

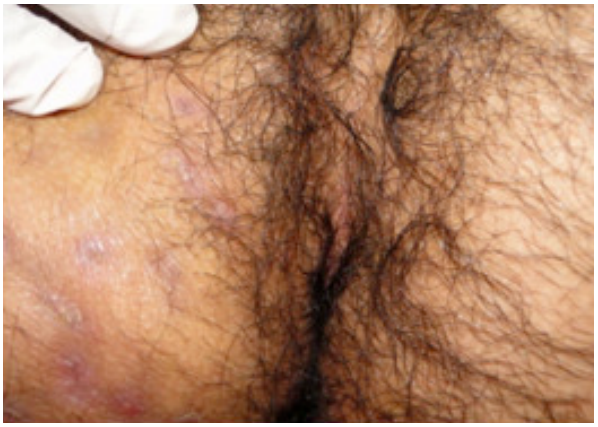


Figure 4. In the gluteal region, cysts of various sizes, hyperpigmented excavated scars.

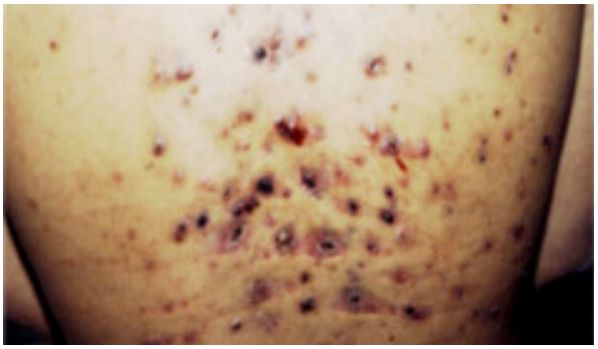


Figure 5. In the back region, nodules and cysts with some necrotic areas.

DISCUSSION

Follicular occlusion syndrome is composed of hidradenitis suppurativa, acne conglobata, pilonidal sinus, and dissecting cellulitis of the scalp, thus constituting the follicular occlusion tetrad.

First suggested in 1956 by Pillsbury, Kligman and Shelley who, by the anatomical, pathophysiological and clinical similarities of this multiple inflammatory event, described the follicular occlusion triad (abscedans cellulitis, acne conglobata and hidradenitis suppurativa). Later in 1975, Plewig and Kligman added another entity, the pilonidal sinus, naming this clinical picture, follicular occlusion tetrad.[2,3]

In 2007, Dr. Ana Kaminsky suggested the name follicular occlusion syndrome of the terminal follicles (FOST),[4] which respects the individuality of the entities that make up this table and identifies the site where the pathological process develops, taking into account that there are three types of structure of the pilosebaceous unit:

- **Villous follicle:** short, fine hair and small sebaceous glands.
- **Sebaceous follicle:** medium-sized hair and large multilobed sebaceous glands.
- **Terminal follicle:** long, thick hair and a large sebaceous gland. The terminal follicles of the axilla, groin, anal fold, pubis, and scalp are often associated with apocrine sweat glands, which make

up the so-called apocrine pilosebaceous unit, consisting of a follicle with sebaceous appendage and an apocrine sweat gland, whose function is related to genetic factors and circulating hormones, such as androgens, responsible for the increase in size of the sebaceous glands and the excretion of sebum.[4]

Suppurative hidradenitis

Hidradenitis suppurativa is a chronic and recurrent inflammatory skin condition that initially presents as tender subcutaneous nodules coalescing into deep dermal abscesses. Lesions most often occur in skin areas with apocrine glands such as the axillae, inguinal, perianal, perineal, inframammary, and buttocks.[5]

This condition manifests after puberty, with a peak incidence in the second and third decades of life. It's three times more frequent in females.

Its etiology considers genetic predisposition, hormonal factors, repeated trauma. A highly significant association with smoking and being overweight has been documented.[6]

The initial lesion is follicular plugging with occlusion, secondary infection and subsequent inflammatory response, destruction of the pilosebaceous-apocrine apparatus with extension to the adjacent subcutaneous tissue.[7]

The typical characteristics of the lesions, topography, chronicity and recurrence establish the diagnosis. Ultrasound is useful for staging, and the differential diagnosis, as well as in demonstrating the existence of subclinical inflammation and inapparent fistulous tracts.[8]

The comprehensive approach to this disease begins with the management of possible triggering and exacerbating factors. Oral corticosteroids can be useful in outbreaks in short-course regimens. Prednisone at doses of up to 0.5-1mg/kg/day. On the one hand, prolonged antibiotic treatments depend on the reduction of the bacterial load in the lesions (microbiome), understanding that these low-pathogenic microorganisms are a stimulus for the immune response, and on the elimination of the biofilm. On the other hand, some antibiotics have been shown to have some anti-inflammatory effect. The combined use of clindamycin and rifampicin (300mg of each drug every 12h for 10 weeks) has positioned itself as one of the main therapeutic alternatives in moderate-severe cases.[9,10]

Acne conglobata

It is the most serious and rare form of acne vulgaris, characterized by the formation of comedones, nodules, abscesses, cysts, fistulas and scars.

It is a rare pathology, which can develop from scratch or from mild forms of acne, the most serious presentation being acne vulgaris.

Males are more affected by the condition and it usually manifests more frequently between the ages 18-30.[11]

The primary cause remains unknown. However, reactivity to cutibacterimun acnes is considered a remarkable factor immersed in a complex process of self-inflammation.[12]

The most frequent compromised areas are the face, the upper region of the trunk, the buttocks, the abdomen, the limbs, the neck, the earlobes and the scalp; it is accompanied with seborrhea. It is characterized by the presence of grouped comedones, papules, pustules and nodules, which accompany deep inflammatory processes.[12,13]

Dissecting cellulitis of the scalp

Dissecting cellulitis of the scalp, also known as perifolliculitis capitis abscedens et suffodiens or Hoffman's disease, is a scarring alopecia. It has also been described as neutrophilic cicatricial alopecia of unknown origin. Initially described by Spitzer in 1903, the condition was given its eponymous name in 1908.

The primary pathologic event is obstruction of the pillar infundibulum by hyperkeratosis, accumulation of obstructed follicular products with rupture, and intense inflammatory reaction in the hair follicle and

bulb. This leads to the formation of pustules that develop into tracts that eventually coalesce into chronically inflamed tissue.[14]

Cellulitis dissecans of the scalp is found predominantly in men of Afro-Caribbean descent in early adulthood or middle age, although women and Caucasians have occasionally been reported to be affected. The cause is probably multifactorial.[14]

Pathologic examination may show a reduced number of follicles with evidence of follicular rupture and the presence of neutrophils.

Diagnosis is clinical. There are a wide variety of treatments that have been reported, including antibiotic suppression therapy, zinc sulfate, isotretinoin, corticosteroids, antiandrogens, biologic agents, laser therapy, aminolevulinic acid photodynamic therapy, radiation therapy, limited surgical therapy, and surgical resection. radical (scalpectomy).[15,16]

Pilonodal sinus

Pilonodal sinus is a disease reported by Anderson et al. as hair detected in a sacrococcygeal ulcer in 1847, first defined by Hodges et al. as a pilonodal sinus and found primarily in the sacrococcygeal area. In addition to this area, it can also be seen in rare locations such as the navel, forehead, scalp, clitoris, interdigital area, penis, abdomen, neck, and armpit.

Its incidence corresponded to 26 cases per 100,000, affecting men twice as often as women. It is more common in young adults. Males are thought to be at higher risk due to their hirsute nature. Pilonodal sinus is also associated with obesity, sedentary occupation, and local irritation or trauma.[17]

Pilonodal disease can appear as an acute abscess along with sinus tract formation or as chronic or recurrent abscesses with extensive and branching sinus tracts. The common form is an acute abscess characterized by the existence of a midline fossa in the natal cleft typically identified 4 to 8 cm from the anus. This primary tract leads to subcutaneous cavity, which contains granulation tissue and, usually, hair follicles present in two-thirds of cases in men and one-third of cases in women. It can be seen projecting from the opening of the skin.[18]

Treatment of pilonodal disease depends on its presentation and ranges from simple incision and drainage to wide excision with extensive reconstructive procedures.[19]

CONCLUSION

The entities grouped as a follicular occlusion tetrad: dissecting cellulitis, acne conglobata, hidradenitis suppurativa and pilonodal cyst can be considered in isolation or in various combinations, often 2 or 3 are found in the same patient, as in the clinical case presented, where clinical examination is essential for diagnosis. The prescribed treatment was Doxycycline 100 mg PO daily, Dapsone 100 mg PO daily. The lesions disappeared with this antibiotic association. Currently, the patient is evaluated for head and neck surgery to treat scalp lesions. Medical and surgical options for these disorders remain somewhat limited in their effectiveness.

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