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Review Article

Hidradenitis suppurativa: a review

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ABSTRACT

Hidradenitis suppurativa (HS) is a chronic inflammatory disorder that is characterized by recurrence, as well as the characteristic location of skin lesions. Patients usually develop very painful inflammatory nodules that generally end in the formation of multiple abscesses and fistulas that typically occur in the skin of the axillary, inguinal, buttock, and perianal folds. It significantly affects the quality of life of patients, leaving physical, economic and psychological sequelae. There is a wide therapeutic arsenal available, but each patient must be individualized and the best possible treatment determined. Early assessment and intensive treatment of the disease can prevent and even avoid significant sequelae and permanent deformities.

Keywords: HS, Chronic inflammatory disorder, Skin lesions

INTRODUCTION

Hidradenitis suppurativa (HS), also known as acne invertia, is a chronic inflammatory disorder that is generally associated with systemic manifestations. HS is characterized by its chronicity, recurrence, as well as the characteristic location of skin lesions. Patients usually develop very painful inflammatory nodules that generally end in the formation of multiple abscesses and fistulas that typically occur in the skin of the axillary, inguinal, buttock, and perianal folds.[1,2] HS usually occurs in young patients and no predisposition by sex has been observed, it significantly affects the quality of life of patients, leaving physical, economic and psychological sequelae because due to the fetid/purulent behavior of some lesions, patients tend to isolate themselves, having a profound negative effect on their daily lives.[3,4]

EPIDEMIOLOGY

Worldwide there are no clear reports as it varies widely depending on the method of data collection. Registry-based studies report a low prevalence of <0.1%, while studies based on self-reported disease, diagnosed using validated questionnaires, demonstrate a substantially higher prevalence in the range of 1 to 2%, although

recent reports from the USA and Norway have reported an increasing incidence of HS.[5,6]

PATHOPHYSIOLOGY

Some authors have identified a hereditary family h/o in ~30% of patients with HS.[7] In several Chinese families, mutations have been identified in genes encoding γ-secretase, which is protease complex with multiple intramembrane subunits. γ-secretase cleaves several transmembrane proteins that are involved in different signaling cascades.[8] So far, about 40 mutations in different components of γ-secretase have been described in patients with HS. Most of these mutations appear to alter γ-secretase activity.[9] Experimental genetic studies in mice have identified that alteration in γ-secretase results in transformation of hair follicles into cysts. [10] However, most HS patients with positive family history do not harbor γ-secretase mutations, so genetic characteristics that contribute to development of disease in these patients are still unknown.[1]

RISK FACTORS

Smoking has been identified as one of the main risk factors since nicotine induces epidermal hyperplasia, in addition to influencing bacterial propagation and biofilm formation, as well as increase the production of IL-10.[11-13] 50% of HS patients are obese, and ~40% of HS patients have metabolic syndrome. Obesity could contribute to the pathogenesis of HS through subclinical inflammation, metabolic changes, and increased friction in skin folds. Shaving and the use of chemical depilatories, deodorants, and talcum powder have been shown to play no role as causative factors in the pathogenesis of HS.[1]

DIAGNOSIS

The clinical diagnosis of HS is based on the nature and location of the lesions, the course of the disease, which can be persistent for at least 6 months or recurrent >2 skin lesions within 6 months. Typical lesions include inflammatory nodules, abscesses, swollen and draining sinus tracts/fistulas, scarring and presence of multiple types of lesions simultaneously is frequent.[14] The most frequently affected locations are the armpits and the inguino-genito-femoral region. Most cases will be associated with numerous follicular papules and pustules (folliculitis)/epidermoid cysts. HS can progress through flare-ups, alternating phases of clinical activity and phases of latency, or progress to a chronic inflammatory state with the formation of fistulous tracts, dermal fibrosis, and characteristic scars.[15]

Diagnosis is exclusively clinical, there is no specific diagnostic test and 3 concise premises must be met: Location of lesions in intertriginous areas, predominantly in armpits/groin, objectify presence of any of lesions considered typical, including painful nodules/ abscesses/ suppurative sinuses/scars and that course of disease presents multiple recurrences.[16,17] Differential diagnosis established with: local pyodermatitis such as folliculitis, boils, simple abscesses, erysipelas, cellulitis, neoplasms, lymphogranuloma venereum, actinomycosis.[15]

TREATMENT

Most limiting and most influential symptom is pain. First line in treatment of pain in HS includes topical anti-inflammatory drugs and non-steroidal anti-inflammatory drugs/oral paracetamol. In some cases, pain can be controlled with application of ice packs and, topically, with 5% lidocaine, 4% microsomal lidocaine/1% diclofenac gel. Some patients report immediate pain relief when abscesses are drained.[18]

Ultrasound-guided intralesional corticosteroid injections can improve inflammatory lesions in HS patients in long-term, not only for inflammatory nodules and abscesses but also draining fistulas. However, in order to achieve this effect, high-potency corticoid such as triamcinolone acetonide should be chosen and infiltration should be placed appropriately. For this, use of ultrasonography is crucial.[19-21] Intralesional corticosteroid infiltrations can have continued beneficial effect on abscesses and simple small to medium draining fistulas in HS patients, evidence level 2b.[22]

Surgery is indicated in isolated nodules, localized fistulas and in extensive severe cases that do not respond to medical treatment.[23] There are several surgical techniques: a) incision and drainage, b) deroofing ("unroofing") and marsupialization, c) localized/ extensive extirpation. The most widely used technique in primary care and emergency centers is simple incision and drainage. It is simple procedure that can be performed in the office under local anesthesia and usually produces rapid pain relief from isolated nodules, although recurrence is the norm.[15]

CONCLUSION

Cause of HS is multi-pathogenic with complex etiopathogenesis, with numerous systemic associations in which it affects patients in all directions. There is wide therapeutic arsenal available, but each patient must be individualized and best possible treatment determined. Early assessment and intensive treatment of disease can prevent and even avoid significant sequelae and permanent deformities.

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