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Case Report

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Hidradenitis Suppurativa, Sapho and Bariatric Surgery

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Abstract

Hidradenitis suppurativa (HS), also known as hidradenitis or acne inversa, is a chronic inflammatory dermatosis, characterized by recurrent nodules, abscesses and fistulous tracts, mostly affecting the anatomical folds. This condition may present as outbreaks and evolve towards a chronic inflammatory status with the development of fibrous tracts and hypertrophic scars.

Several cases have been published over the past few years, suggesting a likely association between this disease and rheumatic diseases, among which SAPHO Syndrome stands out. Most rheumatologists believe that this is a variant of Spondyloarthritis (SpA), while other consider it part of Psoriatic Arthritis; however, this fact becomes relevant since in some cases la hidradenitis suppurativa develops in the gluteal and perianal region and may be mistaken by the perianal variant of inflammatory bowel disease (Enteropathic Arthritis).

This clinical case analyzes the characteristics of a patient with hidradenitis suppurativa Hurley stage I, who following a sleeve gastrectomy advanced to Hurley stage III, with failed anti-TNF response, in addition to presenting SAPHO-type manifestations.

Keywords: *Hidradenitis, SAPHO, Microbiota, Spondyloarthritis*

Introduction

HS is a chronic inflammatory process, with a prevalence affecting around 1% of the population. Its onset is usually after puberty, commonly developing early in the third decade of life, and frequently remains active during the third and fourth decades of life. It has been found that in women, there is often an improvement with the onset of menopause, so it is usually males who continue with the active disease after 50 years old [1,2].

With regards to gender distribution, HS is more frequent in females than in males. Based on the information from published trials, the female to male ration is approximately 3:1, with ranges according to the most relevant studies from 2.6:1 and 3.3:1 [2-4].

From the epidemiological point of view, HS has been associated to multiple comorbidities, as shown in Table 1, with SAPHO as one of the most outstanding [2].

Dermatological diseases
Pyoderma gangrenosum (PASH syndrome)
Pityriasis rubra pilaris
Acanthosis nigricans
Panniculitis (erythema nodosum)
Fox-Fordyce disease
Cushing's disease
Acromegaly
Thyroid diseases
Follicular occlusion syndrome

Acne conglobata
Dissecting cellulitis of the scalp
Pilonidal sinus
Genetic disorders associated with follicular occlusion
Congenital pachyonychia
Dowlin-Degos disease
Steatocystoma multiplex
Rheumatological diseases
Spondyloarthritis (Spa) **Psoriatic Arthritis (PsA)** **SAPHO Syndrome**
Psychiatric diseases
Depression
Anxiety
Alcohol or drug abuse
Neoplasms
Cutaneous epitheliomas (epidermoid carcinoma)
Lymphomas
Gastrointestinal Diseases
Crohn's disease **Ulcerative Colitis (UC)**
Kidney Diseases
Nephrotic syndrome
Acute interstitial nephritis
Other
Anemia
Amyloidosis
Polycystic Ovary Syndrome
Behçet's disease
Sjögren syndrome
PAPA syndrome
Down's syndrome
Keratitis-ichthyosis-deafness syndrome

Table 1: HS differential diagnosis.

HS is currently considered an inflammatory disease of the pilosebaceous follicle with an underlying immune system imbalance, in a genetically predisposed individual. The assumption is that this predisposed patient is under the influence of certain exogenous triggering or aggravating disease-modifying factors. Approximately 40% of the patients have family members affected by the disease. The most frequently observed hereditary pattern is autosomal dominant. The genes involved are localized in the 1p21.1-1q25.3 locus [2,4].

Deactivating mutations have been described in the presenilin I genes (PSENI), presenilin II enhancer (PSENEN) and nicastrin NCSTN in families presenting with severe and atypical manifestations of the disease. These genes encode for 3 of the 4 subunits of γ-secretase, involved in the Notch receptors pathway. These mutations have been associated in mice trials with epidermal and follicular dysfunction, with absent or disturbed formation of the sebaceous glands [1,4].

Clinical Case

Female, 35-year old patient with a history of obesity grade III and metabolic syndrome, with no family history of autoimmune disease. The patient visited the dermatology clinic since she was 25 years old, because of mixed inflammatory lesions in the axillar and perineal regions, that improved with the use of antibiotics (ATB) and corticosteroid creams (CS), with 6 years of mild and controllable evolution of the underlying disease. Due to the history of metabolic syndrome and obesity grade III, the patient was offered a sleeve gastrectomy (bariatric surgery) when she was 30-years old. 24 months after the bariatric surgery, the axillary and perineal lesions evolved to Hurley stage II (few fistulous tracts and dystrophic scars); however, at 33 years old, the patient presented with multiple abscesses, fistulous tracts and dystrophic scars (Hurley III). Systemic therapy (CS and ATB) was initiated at that point, with later addition of dapsone and colchicine-type anti-fibrotic agents (Figure 1). Since no improvement was achieved, the patient underwent surgical procedures

(incision with drainage, moving on to Deroofing and marsupialization).

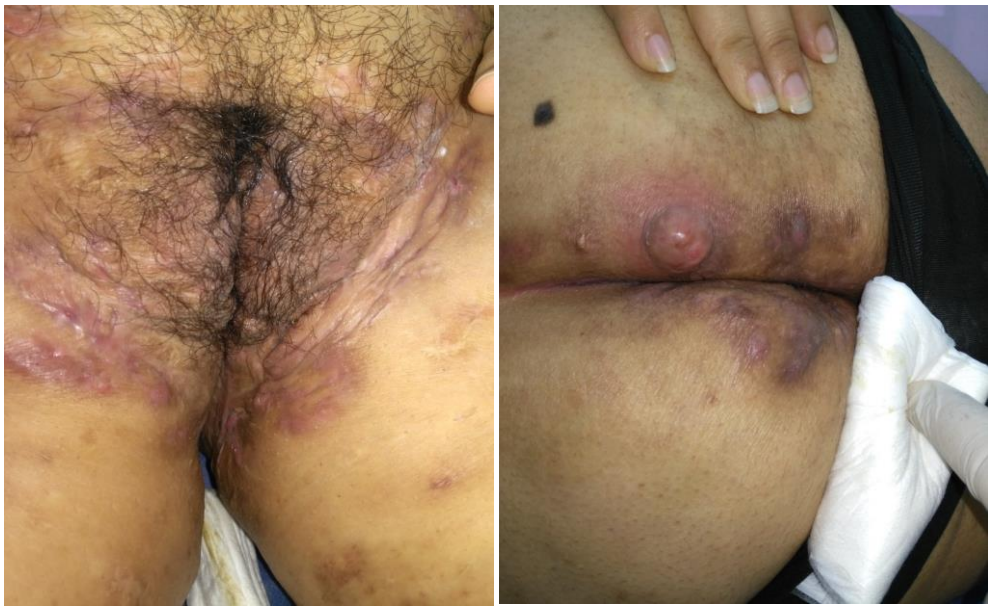

Figure 1: Before Adalimumab treatment (week 1).

The patient was re-assessed at 34 years old, because of persistent multiple abscesses with dystrophic scars; a work plan was set in motion to rule out granulomatous vs. autoimmune disease. The intradermal reaction tests were negative (PPD, Sporotrichin, Candidin), the lung x-rays were normal and ANA, ANCA, rheumatoid factor and ASCA IgA were all negative. However, a gastroenterology assessment was requested to rule our inflammatory bowel disease (IBD). An upper and lower GI video-endoscopy was conducted, with the following results: traces of semiliquid stools, the ileocecal valve, the appendix and the cecum were identified, with normal mucosa and vascular pattern. The exploration of the ascending, transverse, descending and sigmoid colon and rectum, revealed a normal mucosa and vascular pattern.

The culture of axillary and perianal secretions was able to isolate Proteus mirabilis, sensitive to Levofloxacin, Imipenem, Gentamicin, and Meropenem. After antibiotic therapy, and with negative cultures on two occasions, methotrexate 20 mg/week was initiated, in addition to anti TNF therapy (Adalimumab) 40mg/week. The lesions improved over the course of 8 to 10 months (Figure 2). At 12 months the patient developed rachialgia of inflammatory characteristics, which initially improved with NSAIDs; at 36 years of age, the patient presents morning stiffness lasting more than one hour, in addition to feet arthralgia, and the decision was made to undergo a rheumatology assessment.

The osteoarticular examination showed evidence of Shöber 13 cm, Achilles enthesitis, lateral fibula with synovitis of both subtalar joints, in addition to positive sacroiliac maneuvers (Patrick's sign with sacroiliac pain). An MRI of the sacroiliac joints reported synovitis of the right SI, one single focus of bone edema with no fatty lesions or erosions (Figure 3). A new immune-rheumatological profile including ANA, HLA B27, HLA Cw6, and HLACw7 was negative.

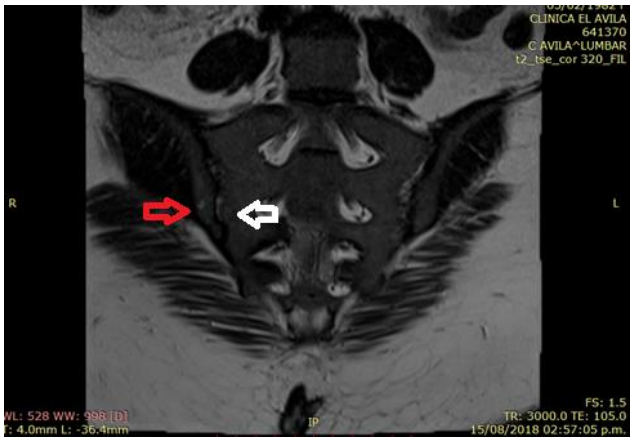

Figure 2: Sacroiliac MRI:
- Red arrow Bone Edema.
- White Arrow Synovitis

Since the patient was on anti-TNF therapy (Adalimumab) and one Methotrexate-like DMARD, it was replaced by sulfasalazine, which the patient received for 12 weeks with no pain relief. They even considered a swap to ustekinumab (Anti IL-12/23), but its use was not approved by the regulators in the country, so the decision was made to switch over to another Anti TNF (Infliximab). The starting dose was 5mg/kg with an induction dose o week 0, 2, 6 and then every 8 weeks. By week 12 all abscesses and fistula had cleared, but there were still dystrophic scars undergoing maintenance therapy every 8 weeks.

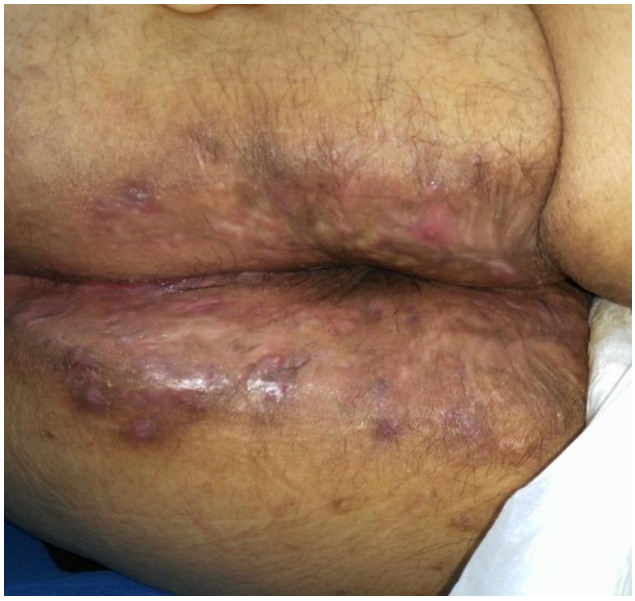

Figure 3: After Anti-TNF treatment (week 16).

Discussion

There is a link between the gut and spondyloarthropathies, was established back when a seronegative arthritis was described as Reactive Arthritis (ReA), which is triggered by enteritis due to gram-negative bacteria; then ankylosing spondylitis and peripheral arthritis were associated with IBD (Crohn's disease, ulcerative colitis). Scientific studies early in the 90's led several gastroenterology societies to report that an unexpectedly large number of patients undergoing ileocolonoscopy, presented with long-standing or chronic oligoarthritis or sacroiliítis [6]. The theory then was bacterial translocation as the cause of the Reactive Arthritis, so many patients were treated with antibiotics and even steroids. Inflammatory changes were also identified in the terminal ileum, the colon or both. These changes are characterized by acute intestinal inflammation or infection, but around one fourth of the patients presented chronic lesions, probably for early Crohn's disease.

Our study group (GRUVES) reported in the database of rheumatic patients receiving biologics (BIOBADAVEN), several patients (n=4; 0.08%) that after bariatric surgery (BS) developed HLA B-27 + associated SpA (n=3;75%) [7]. More recently, a Brazilian group reported nine cases of patients who claimed developing back pain or change in the pattern of pain intensity (intensity or night pain) after undergoing bariatric surgery (mean time 14.7 ± 18 months). Furthermore, 8 of them (88.9%) tested positive to the human leukocyte antigen (HLA-B27). All of the nine patients were classified according to the ASAS as ax-SpA and five (55.6%) patients were classified as AS, according to the mNY criteria (8).

Humans evolved within a microbial ecosystem that led to an interrelated physiology. The intestinal microbiota may send signals to the brain via the immune system, the vagus nerve, or other host-microbe interactions enabled by gut hormones, the tryptophan metabolism regulation and microbial metabolites – i.e. short-chain fatty acids (SCFA). Alterations at specific sites of the mucosa suggest that microbial factors may affect the mucosal immune response, and also play a relevant role in the pathogenesis of early arthritis (9). Variations in the composition of the microbiota and the abundance of microbiota, in other words, dysbiosis, may trigger a range of autoimmune and inflammatory diseases resulting from an imbalance in the T-cell population, such as Th1, Th2, Th17 and Treg cells [9-11].

During the last few decades, obesity has become a public health issue in many countries. Bariatric surgery was designed to fight comorbidities rather than promote vanity; however, the technique has improved over the years, decreasing mortality. Some meta-analysis published in the world literature show that hypertensive and diabetic patients have been able to reduce the use of anti-hypertensive and sugar-lowering agents, and some are even in remission of the disease (9). It is mandatory to properly classify patients that are suitable for this surgical technique, since we, rheumatologists, have observed over the past two decades, that while joint pain in patients with knee and hip osteoarthritis has improved, avoiding total hip replacement, we have also seen that in patients with rheumatoid arthritis class II-III receiving triple synthetic DMARDs therapy, after bariatric surgery they just need one DMARD [12-15]. Patients with RA who have received several biologic DMARDs with multiple treatment failures, go into remission after surgery [16-17]. However, there is

apparently, a subset of patients with seronegative arthritis or spondyloarthritis, in whom their symptoms get worse [18,19,20].

Take Home Point

- All patients with a personal or family history of autoimmunity should be evaluated by a rheumatologist.
- Patients with psoriasis, SAPHO, psoriatic arthritis or IBD seem to have greater susceptibility to microbiota changes.
- Use of probiotics as part of the treatment would seem a logical alternative in patients with IBD and SAPHO.
- In the case of patients not responding to Anti TNF, the change of molecules seems to be a better decision than to swap to other mechanisms of action such as Anti-Il.

Disclosure

The authors have no conflict of interests to disclose.

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