Hidradenitis suppurativa with complete tetrad

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Introduction

Hidradenitis Suppurativa (HS)

Synonyms: Apocrinitis, Hidradenitis Axillaries, Acne Inversa

It was first described in 1885 by French physician Alfred Verneuil for its characteristic intertriginous distribution [1]. It is a recurrent and suppurative disease with an insidious onset. It is characterized by deep furuncles, abscesses, fistulas, sinus tracts and scarring and has a prevalence rate of 0.3% to 4% of the general population [2]. The average age of onset is 23 years but it may fall anywhere between post-pubertal age to middle age [3]. HS is more prevalent in women than men by a ratio of between 2:1 and 5:1, with genito-femoral lesions found more often in women and anogenital lesions found more often in men [3,4]. The most commonly affected site, the axilla, is involved in equal proportion of men and women. Other affected area may include the areola, infra-mammary folds, periumbilical skin, scalp, zygomatic and malar face, buttocks, thighs, popliteal fossa, ear canal and eyelids [2,5]. Patients with active disease generally develop 2 furuncles per month with average disease duration of 19 years [5].

Case report

A 42yr old male presented to dermatology department with complaint of generalized, bilaterally symmetrical, mildly pruritic, reddish tender lesions on the face,
shoulder and back since last 5 years. The patient also complained of patchy hair loss following the appearance of painful lesions on the scalp since last 4 years. There was history of large painful pus filled lesions with purulent discharge in the axillary and anogenital regions since last 3-4 years for which patient had been consulting a general surgeon. He had taken multiple courses of several antibiotics for the lesions which improved partially, only to recur later. He had undergone incision and drainage for the lesions around the anus following which pus discharge, pain and tenderness decreased after 2 days, but relapse occurred after 15 days. He gave a history of a dimple at the lower end of the spine. Patient had, however, never ever taken any treatment for the lesions on scalp and the back. He was a chronic smoker since last 20 years. There was no history of joint pain or joint swelling. He had no history of exposure to chemicals due to occupation or otherwise. There was no history of diabetes. Family history for similar disease was negative.

**Physical examination:**

The patient was overweight but otherwise healthy. Vitals were normal.

Dermatological examination revealed few tender papules with patchy areas of scarring alopecia (on the vertex) on the scalp. Patient also had male pattern hair loss. There were generalized bilaterally symmetrical multiple follicular papules, follicular pustules and 2 to 3 cm fluctuating nodules on the face, shoulder and back. There were multiple atrophic scars on the face and more on the back. There were multiple, intermingled, 2 to 6 cm, at places linear hypertrophic and puckered scars and dermal contractures in the axillae and the anogenital region.

Scarring, fibrosis and sinus tracts were seen. The lesions were painful and had a foul odour.

**Investigations:**

Cultures of the lesions did not grow out organisms. Haemoglobin was 10.5 gm%, Total Leucocyte Count was 10,500/ mm3, Differential Leucocyte Count was Neutrophil 73%, Lymphocyte 23%, Eosinophils 2% and Monocyte 2%. Erythrocyte Sedimentation Rate was 30 mm at the end of first hour. Total Serum Protein was 7 gm%, Albumin 4 gm% and Globulin 3 gm%. Repeated cultures of pus, Rheumatoid Factor, Lupus Erythematosus cell phenomenon, Mantoux test were negative. Urine and stool tests were normal. Patient was non reactive for Human Immuno Deficiency Virus and Hepatitis B Surface Antigen (HBsAg).

In the face of typical clinical picture the diagnosis of follicular occlusion tetrad was made which includes four entities namely Acne Conglobata, Hidradenitis Suppurativa, Dissecting Cellulitis of scalp and Pilonidal Sinus.

**Treatment:**

Patient was started on topical clindamycin lotion twice daily and oral clindamycin and rifampicin 300 mg each twice daily for 12 weeks. Patient was reviewed every fortnight. At the end 3 months of above therapy patient responded partially. So keeping in view his clinical presentation of Follicular Occlusion Tetrad (FOT) along with male

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pattern baldness he was started on tab finasteride 5 mg daily while continuing the topical therapy. The patient started improving on this treatment. At the end of three months the lesions became quiescent and there was no relapse till 1 year thereafter.

**Discussion**

The initial symptoms of HS are localized erythema, pruritus and hyperhidrosis followed by the spontaneous development of painful and tender red nodules and abscesses with incessant purulent drainage, open comedones, sinus tracts and scarring [2,4]. This was present in our patient.

Obesity tends to be more prevalent in patients with HS [6], although it is not been universally demonstrated in all studies [7]. It has been reported that the Body Mass Index (BMI) for men and women with HS is elevated at 27.8(+3) and 28.5(+5), respectively [8]. Obesity acts as a secondary factor, aggravating disease through mechanical trauma at skin folds. Our patient was overweight.

Our patient was a chronic smoker of 20 yrs. A high incidence of smoking (89% are active smokers compared with 46% of controls) occurs amongst HS patients [9,10]. While HS is not typically thought to be related to medications, it has been reported to be exacerbated by lithium [11,12] and sirolimus [13].

Family history for similar disease was negative. In women, HS occurs almost exclusively after menarche. Premenstrual flares are common in about 50% of all cases. HS tends to improve with pregnancy and rebounds after parturition. Signs of virilization have been seen in HS but most case series indicate no significant change in serum hormone levels in HS patients [14]. The free androgen index is increased because of low level of sex-hormone-binding globulin, which is influenced by body weight. Thus, the role of androgens in the pathogenesis of HS remains unclear and proved to be secondary rather than primary.

Familial forms of HS have been described, although no specific gene has been identified [15]. A recent genome-wide scan in a Chinese family showed that a disease gene of acne inversa is located on chromosome 1p21.1-1q25.3 [16].

HS has been associated with various dermatologic syndromes including Keratitis Ichthyosis Deafness [17] which was absent in our patient, Synovitis-Acne-Pustulosis-Hyperostosis-Osteitis syndrome [18], and Crohn disease [19] in addition to the follicular triad. Arthritis is commonly associated with HS and can worsen with a flare of HS. Arthropathy typically involves the large joints in the extremities, particularly the knee joints. There was no history of joint pain or joint swelling.

Chronic HS lesions have a higher risk of developing malignancy [20]. HS has a significant negative effect on quality of life that correlates with the disease severity and a more detrimental effect than other dermatologic conditions [21].

Hurley classification [22] separates HS into three stages: Stage I is limited to the presence of abscesses without evidence of sinus tracts or scarring. Stage II demonstrates sinus tracts and scarring with discrete recurrent abscesses. Stage III
represents diffuse involvement of interconnected sinus tracts, scars and abscesses. Prominent open comedones often with multiple orifices are seen. The more severe the disease, the more refractory it is to medical treatment and more likely it is to recur after surgical intervention. Our patient was in Stage II.

HS lesions are concentrated where apocrine hair follicles are typically found. The mechanism of disease for HS is not completely understood and has evolved from being a process centered on apocrine glands [23] to a process centered on hair follicles. While inflammation of the apocrine glands can be seen, the pathophysiology of HS is thought to be similar to acne vulgaris, with hair follicle occlusion being the initiating event. The disease begins with spongiosis of the infrainfundibular region; follicular hyperkeratosis and dilatation of follicular infundibula, leading to comedone formation and follicular rupture [24]. Follicular rupture induces the recruitment first of neutrophils, followed by a granulomatous infiltrate with foreign body giant cells [24]. The dermal abscesses then extends into the subcutaneous fat to involve the adenexal structures. Thus, the inflammation of the apocrine glands, or hidradenitis, is thought to be a secondary event [25]. The subsequent fibrosis and formation of sinus tracts are likely a result of tissue repair response to chronic inflammation, bacterial superinfection, and necrotic debris[26]. HS is thus a misnomer, because the pathogenesis does not involve sweat glands dysfunction as originally thought.

Whereas the role of infection is not clearly understood, though multiple species of bacteria are isolated from HS lesions. At times, cultures are routinely negative. Thus bacterial infection is likely to be secondary to chronic sinus tracts and moisture rather than of primary etiologic importance. When present, bacterial cultures are often polymicrobial. Staphylococcus aureus, S epidermidis, Streptococcus milleri and S hominis have all been isolated from aspiration of deep lesions about 49% of the time [27]. Peptostreptococcus was the most common anaerobic organism. Enterococcus, Enterobactericiae, diptheroids, Bacillus cereus, Propionibacterium acnes, Lactobacillus and Bacteroides have also been described [28].This case is being reported for the complete Tetrad of HS (Fig. 1), Dissecting Cellulitis (Fig. 2), Acne Conglobata (Fig. 3), And Pilonidal Sinus (Fig. 4), are known as the Follicular Occlusion Tetrad.
Fig 2: Dissecting Cellulitis

Fig 3: Acne Conglobata

Fig 4: Pilonidal Sinus
Treatment:

Treatment of HS is challenging. Although there are various treatment options for HS, none of the traditional therapies is wholly satisfactory or uniformly effective. Therapy varies based on the on the clinical severity of the disease, although there is considerable overlap between treatment groups. Mainstays of treatment are non-steroidal anti-inflammatory drugs for pain and inflammation, antiseptics, antibacterial soaps, warm compresses, hydrotherapy, and antibiotics.

Medical treatments can reduce inflammation and associated tenderness and drainage, but usually do not halt disease progression. Medical therapy can be divided into topical and systemic. Clindamycin 1% solution twice daily for 12 weeks helps in resolving abscesses and pustules [29]. Systemic medications can be further divided into antibiotics, retinoids, hormonal modulators, and immunosuppressants. Antibiotics can be helpful, particularly if specific organisms can be demonstrated. However, antibiotics require fairly long courses of therapy. Oral tetracyclines, clindamycin, rifampicin and minocycline have been tried. In a case series of 14 patients receiving oral clindamycin 300 mg twice daily and rifampicin 300 mg twice a day for 10 weeks achieved remission in 8 of the 14 patients. Remission was induced in 2 additional patients when minocycline was substituted for clindamycin[30]. Results from systemic retinoids, particularly isotretinoin, have proven to be disappointing. A prospective trial with long term follow-up showed that only 23% of HS patients benefit from isotretinoin 0.5 to 1 mg/kg and only 16% of patients showed durable remission after treatment[31].

Antiandrogen therapy has been studied in HS. Cyproterone acetate and spironolactone need further study to define their role in treating HS. Finasteride, a 5-alpha reductase inhibitor, 5 mg once a day showed improvement by 8 weeks and remissions from 8 to 18 months [32].

Immunosuppressive therapy has been used in HS particularly in acute inflammatory stage with varying degrees of success. Dapsone, cyclosporine, azathioprine, mycophenolate mofetil, tacrolimus have been tried.

Biologics like TNF-alpha inhibitors (infliximab, etarnecept, adalimumab) and T-cell specific agents (alefacept and efalizumab) for the treatment of HS have thus far proved to have variable long-term results and are laden with significant adverse effects. However, the use of three anti-TNF biologic agents might be considered a potentially life-altering maneuver [33,34,and35].

Non-surgical modalities like cryotherapy, carbon-dioxide laser and photodynamic therapies have been tried but none of them have proved to be satisfactory. Radiotherapy with x-rays has been reported to be useful in control of the disease [36].

Surgical excision is the treatment of choice for early disease. Surgical techniques for excision and repair vary, depending on extent of disease, location of the lesions, existing comorbidities, and chronicity of disease [37].
Differential Diagnosis:

Possible misdiagnoses are numerous, and vary with the stage and site. Axillary and inguinal lesions that have ulcerated may be confused with scrofuloderma. Inguinal lesions may simulate actinomycosis, granuloma inguinale or lymphogranuloma venereum. When only one or a few nodules and sinuses are present in the anogenital region, pilonidal sinus, sigmoidal diverticulitis and Crohn's disease must be excluded.

In atypical cases, the diagnosis may be difficult. The characteristic comedones should be sought and the non-specific histological changes may be negatively useful.

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