

## Hidradenitis suppurativa, a rare skin disease

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### Abstract

Hidradenitis Suppurativa (HS) is a rare, chronic and recurrent skin disease involving folliculopilosebaceous unit. It is a debilitating disease due to its chronicity, painful relapses and cosmetic outcomes. It affects the patient's personal, family, social and professional life. It is often diagnosed late during its course, due to lack of awareness and knowledge among general practitioners. Management is symptomatic, as ultimate treatment is latest and has limitations due to cost and availability issues. Reported here is a case of HS, being treated at Hearts International Hospital, Rawalpindi, Pakistan.

**Keywords:** Hidradenitis suppurativa, Folliculopilosebaceous unit, Chronic, Relapses.

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### Introduction

HS is a relapsing and remitting, chronic skin disease, primarily affecting apocrine gland rich areas of the body.<sup>1</sup> It's a genetic disease, and the diagnosis is clinical. The data shows an average time for diagnosis of approximately 7 years.<sup>2</sup> It is difficult to treat, causing frequent relapses. We report a case of HS, after informed written consent from the patient. Patient was being treated in the out patient department of our hospital, with regular follow-ups. This disease has significantly affected his quality of life.

### Case Report

A 44-year-old male presented to us at Hearts International Hospital, Rawalpindi, with 10 years history of eruptive skin lesions. He was a known diabetic (T2DM) and obese with BMI of 35. He had no history of hypertension and his lipid profile was within normal range. Lesions involved the back of the neck, armpits, perianal and groin regions. Patient's brother and 18-year-old son recently started developing similar lesions.

Lesions were painful nodules and papules, with pus and blood discharge, followed by development of hard

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**Figure-1** (a-f): Lesions in different stages of eruptive phase with pus discharge.

crusts and scars. Lesions were aggravated by hot and humid climate, and dried up with medications temporarily, then relapsed again. He has no history of antiperspirants use.

He had been to many local doctors during all these years but his condition could not be diagnosed. Now, he is under treatment in the out patient department of our hospital and has been diagnosed as a case of HS on clinical grounds. He is been managed with intermittent antibiotics, both topical (polymyxin B and bacitracin) and oral (clarithromycin, metronidazole, amoxicillin, clauvulonate, quinolones), oral trypsin/chymotrypsin formulation, apple cider vinegar, a course of oral prednisolone, permethrin and ivermectin (for concurrent scabies), oral itraconazole (antifungal), alprazolam (benzodiazepine for anxiety) and amitriptyline (TCA for depression). Treatment succeeded in controlling relapses and remissions with symptomatic relief during flare ups, and as a result his quality of life has improved but he still awaits ultimate solution.



**Figure-2** (a-f): Lesions in remitting phase with dried up crust and scar formation.

Patient was also advised to maintain a good glycaemic control and adopt life style modifications for weight loss. Regular counselling sessions for the patient and his family members were also carried out to improve the quality of life.

## Discussion

HS, also known as acne inversa or Verneuil disease, is a rare skin disease involving folliculopilosebaceous units,<sup>1</sup> with a prevalence of 1% in European population, and between 0.05% to 0.20% in American population.<sup>2</sup> It is more common in females, with female:male ratio reported as 3:1 to 4:1,<sup>1</sup> especially in second and third decades of life.<sup>2</sup>

Etiology is genetic, inheritance being autosomal dominant with 100% penetrance,<sup>2</sup> with variance in phenotypic expression.<sup>3</sup>

Triggering factors include cigarette smoking, obesity/metabolic syndrome,<sup>2</sup> (risk for HS increases by 1.12 for each unit increase in BMI<sup>4</sup>, irritation from antiperspirants use or possible trauma to hair follicles while shaving axillary hair.<sup>5</sup> Diseases that cause follicular occlusion (acne vulgaris, acne conglobata, dissecting cellulitis of the scalp), pilonidal cyst and Crohn's disease are reported to be associated with comorbidities.<sup>6</sup> HS is not a contagious disease.<sup>7</sup>

It is a chronic, relapsing, inflammatory skin disease, affecting the patient physically, psychologically as well as socially. It involves apocrine glands primarily of axilla, groin, perineum and inframammary regions, causing recurrent painful skin papules, nodules, scarring, sinus tract formation, abscess, secondary bacterial infections,<sup>2</sup> mucopurulent discharge, hyperkeratosis and hypertrophic scarring.<sup>3</sup> Ruptured lesions may become infected, mostly by streptococcus species (*s. viridians*, *s. aureus*, *s. epidermidis*), peptostreptococcus, bacteroides, coryneform bacteria, and gram-negative bacteria, including *escherichia coli*, *klebsiella* and *proteus* species.<sup>5</sup> Patients are at risk of developing stress and depression.<sup>2</sup>

Diagnosis is clinical. Diagnostic criteria include typical appearance of the lesions, their topographic distribution, chronicity and recurrence.<sup>8</sup> Four staging systems, Hurley staging (HS), Physician's Global Assessment (PGA), Modified Sartorius Score (MSS) and HS Severity Index (HSSI) are used for assessing severity of the disease.<sup>2</sup> Biopsies, cultures, and laboratory tests are of limited help in the diagnosis.<sup>9</sup>

Complications include fibrotic changes, scarring, lymphatic obstruction and squamous cell carcinoma (scc) in case of longstanding HS.<sup>5</sup>

Treatment of HS is challenging both on the part of the physician and the dermatologist for control of relapses. It is also demanding on the patient's part with regards to adherence to follow ups due to its chronic recurrent course and difficulty in coping up with the sequelae. Treatment includes medical, surgical, symptomatic during flare ups and management of comorbidities. Patients are advised to avoid skin trauma, especially while shaving, refrain from tight fitting clothings, and keep their skin cool as heat and sweating causes flare ups.<sup>7</sup>

Treatment strategies include topical clindamycin as the most effective modality, systemic clindamycin-rifampicin in mild cases, oral rifampicin-moxifloxacin-metronidazole combination for refractory cases to control relapses and ertapenem to decrease the disease severity, are all reported to be beneficial. Use of oral tetracycline as compared to topical clindamycin is less effective. Benefits of the use of acitretin, cyclosporin A (an immunosuppressant), dapsone (diaminodiphenyl sulfone, an antibiotic of sulfone family) and isotretinoin (13-cis-retinoic acid) is limited and controversial. Use of intralesional or systemic corticosteroids is an option to reduce inflammation, long term use is limited due to its systemic side effects.<sup>5</sup> Latest use of adalimumab and

infliximab (monoclonal antibodies against TNF) has shown to have promising results.<sup>2</sup> Nd:YAG LASER (neodymium-doped yttrium aluminum garnet) and intense pulse light shows significant improvement, whereas, carbon dioxide laser has controversial results.<sup>10</sup>

Surgical options include wide local excision, primary closure, skin grafts, flaps, and vacuum assisted closure (VAC therapy) which can be used depending on the types of lesions and their severity.<sup>16</sup>

Primary take away lesson is for junior doctors and general practitioners, to seek expert opinion if a patient comes up with a disease that is rare and diagnostically challenging as in this case. Misdiagnosis can cause long term physical, cosmetic, social and psychological problems.

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