

An Atypical Giant Form of Hidradenitis Suppurativa of the Pubic Region

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Abstract

Hidradenitis suppurativa (HS) is a chronic, inflammatory follicular cutaneous disease characterized by recurrent skin nodules, sinus tracts and scarring affecting mainly the axilla, groin, and other parts of the body that contain apocrine glands. We report a rare case of HS in a 29 years lady, who presented with a large swelling, insidious in onset, progressive in nature and measuring approximately 25 x 25 cm in the pubic region along with multiple small lesions, scarring and sinuses in axillae and groins for 6 months duration. The lesions were also associated with offensive discharge. The swelling was excised under general anesthesia. The diagnosis was confirmed by histopathologic examination. The post-operative period was uneventful and she is doing well on follow-up.

Keywords: Hidradenitis Suppurativa; Pilosebaceous Unit; Sinuses; Abscesses; Excision.

Introduction

Hidradenitis suppurativa (HS) is a chronic inflammatory cutaneous disease with abscesses, fistulas, and scarring affecting the groin, anogenital area, and axillae. HS has an estimated prevalence of 1%, and moderate-to-severe forms have a high disease burden [1,2].

The disease is a combination of homeostatic abnormalities of the pilosebaceous unit and dysregulation of the skin immune responses. Pathogenesis involves an interaction between a person's genetic background and environmental triggers, among which are the skin microbiome, cigarette smoking, and obesity [3].

Case Report

A 29 years old lady, presented with a large nodulo-pustular skin lesion in pubic region, progressively increasing in size and associated with pus discharge and multiple smaller lesions in axillae and groins of 6 months duration. She was recently diagnosed with diabetes mellitus, hyper-insulinemia and hypothyroidism and on medications for the same. Her body mass index was 29. A thorough clinical examination was carried out and the skin lesion was recorded of size approximately 25 x 25 cm (Figure 1). Due to persistent discomfort, large size of swelling and interference in her daily activities, decision was taken to excise the swelling. After complete preoperative work up, she was taken up for the surgery. Complete excision of swelling was done and the tissue was sent for histopathologic examination (HPE). HPE's gross description- Single skin covered tissue measuring 25x24x11.5 cm. Skin shows multiple greyish white areas of discoloration. Cut section was greyish white throughout the specimen. Microscopic examination- skin covered tissue piece with formation of multiple microabscesses with entrapped hair follicles (Figure 2). The dermis and subcutaneous tissue shows dense collagenisation, fibrosis, and multiple foci of acute-on-chronic inflammation with lymphoid aggregates surrounded by foreign body type of giant cell reaction. She was discharged with Rifampicin and Clindamycin.

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Post-operative period was uneventful and she recovered well with good response.



Fig. 1: Hidradenitis suppurativa in pubic region



Fig. 2: The histopathology of the lesion shows ruptured microabscess (blue arrow) and follicular epithelium (black arrow)

Discussion

The pathogenesis of hidradenitis suppurativa is not clear. However, it is considered to be a multifocal disease, in which atrophy of the sebaceous glands is followed by an early lymphocytic inflammation and hyperkeratosis of the pilosebaceous unit. This is followed by hair follicle destruction and granuloma formation [4,5]. Bacterial infection from microorganisms like Staphylococci, Streptococcus and Escherichia coli is considered to be a secondary event in the pathogenesis of HS [5].

The diagnosis of HS is generally made on clinical grounds. The patients with HS have characteristic inflamed and non-inflamed nodules with discharging abscesses in the axillary, inguinal and anogenital regions. The nodules are located in the deeper dermis and are rounded rather than having the pointed, purulent appearance of simple boils [6].

To make a diagnosis of hidradenitis suppurativa, the patient usually has one of the following:

- Active Disease with one or more primary lesions in a designated site along with a history of three or more discharging or painful lumps in designated sites since puberty.
- Inactive Disease with a history of five or more draining or painful abscess-like lumps in designated sites since onset of puberty, in the absence of concurrent primary lesions [7].

Assessment of the severity of the disease is generally based on the Hurley staging system [4]:

Stage I: Single or multiple abscesses without sinus tract formation or scarring (Figure 1). Stage 1 disease is most common.

Stage II: Recurrent abscesses with one or more sinus tracts and scarring widely separated by normal skin. Most diagnoses of hidradenitis suppurativa occur at this stage and referral to a surgeon is often indicated.

Stage III: The most devastating clinical stage. Diffuse involvement with multiple sinus tracts and no intervening normal skin. At this stage, scarring and oozing lesions are common. Because remission is unlikely at this stage, surgery is often recommended, although other treatments may be considered and tried [7]. About 1% of patients progresses to stage III disease [4].

Patients with stage I disease are managed with topical therapy, while those with stage III disease are managed with systemic therapy. Surgery is indicated in patients with scarring. Topical therapy with Clindamycin 10mg/ml twice daily for three months has been reported to reduce the number of abscesses, nodules and pustules [4,8]. Intralesional injections of glucocorticoids for individual lesions have been also reported. Oral antibiotics are used in patients who do not respond sufficiently to topical therapy [4].

Recently, systemic immunosuppressive agents have been recommended for patients with severe disease. Infliximab, etanercept and adalimumab have been used with good results [4,8].

Surgery is used in patients who have extensive scarring and in patients with stage III disease. Incision and drainage is discouraged as this usually leads to recurrence. Surgery may be limited to localized excision of sinus tracts, cysts and roofs of abscesses with wounds left open to heal by secondary intention [4]. More extensive procedures involving wide excision of all hair-bearing skin in affected areas and subsequent wound cover by skin grafting gives better results but may necessitate severely mutilating procedures [4,9].

Conclusion

Hidradenitis suppurativa is a disfiguring and a debilitating disease. Early diagnosis and prompt treatment is necessary to reduce the burden of the

disease, as it significantly affects the quality of life. There are several options for the treatment of HS with good outcome depending on the stage of the disease.

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