CASE REPORT

A teenage girl with extensive form of Hidradenitis suppurativa of sternal, submammary region and both axilla
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Abstract:
Hidradenitis suppurativa is a rare non-contagious, chronic, relapsing suppurativa cicatrizing skin disease that most commonly affects areas of the body bearing apocrine sweat glands or sebaceous glands, such as the underarms, breasts, inner thighs, groin and buttocks, which manifests itself as a clusters of chronic abscesses, sinuses, fistulas or multifocalized infections. This physically, psychologically, and socially disabling disease is extremely painful to touch and may persist for years with occasional to frequent periods of inflammation, culminating in drainage, often leaving open wounds that will not heal. As it is considered as a rare disease the incidence rate is not well known, but estimated in a range between 1:24(4.1%) and 1:600(0.2%) in which the post-pubertal females are more affected than males. The exact cause of hidradenitis suppurativa remains unclear. What is understood is that the condition is a disorder of follicular occlusion. With genetic predisposition, obesity, hormonal influences etc contribute to the causation of the disease. There is no cure for hidradenitis suppurativa. But early treatment can help to manage the symptoms and to prevent new lesions from developing. Long term antibiotics are the treatment of choice in early stage, but relapsing and recurrent cases may need surgical interventions. Here we present an 18 year old girl suffering from this disabling disease in both axillas, sternal and submammary region for six years. She was treated inadequately with oral antibiotics and surgical intervention in the form of incision and drainage done previously. We treated her with wide excision of the diseased skin and defect reconstructed with local advancement flap. Histopathological study of the excised skin revealed Hidradenitis suppurativa.

Key word : Epidermis/surgery: vitiligo/therapy: transplantation autologous

Introduction:
Hidradenitis suppurativa (from the Greek hidros, sweat, and aden, glands) also known as “Verneuil’s disease”, “Pyoderma fistulans significa,” and “Acne inversa” is a chronic, debilitating, inflammatory skin disease of the apocrine glands.1 It is non-contagious and recurrent; size ranging from a small pea to as large as baseball, typically manifesting as a progression from single boil-like, pus-filled abscesses, or hard sebaceous lumps, to painful, deep-seated, often inflamed clusters of lesions with chronic seepage (suppuration --- hence the name) involving significant scarring.2,3 It commonly occurs on apocrine sweat gland-bearing skin affecting inverse areas of the body (those places where there is skin-to-skin contact - armpits, groin, breasts, etc.).18 The prevalence has not been accurately determined. The disease is found more commonly in the white population than in the black population and is rarely observed in the Asian population. Post pubertal women are more affected than men. It is most common in the third decade of life, but the untreated disease may persist into the seventh decade.4,5.

Leper provided the first descriptions of hidradenitis suppurativa in 1839, when he noticed peculiar localization of abscesses within the axillary and the perianal skin. Verneuil first described the apocrine gland involvement in 1854.6 Clinical features of the disease were described by Lane and Brunstig.7 Experimental
reproduction was achieved by Shelly and Cahn in 1955, which helped to establish the pathogenesis of the disorder. Further work by Conway, Paletta, Pollock, Lettermann, and others in establishing the surgical management of the disease has been noteworthy.

The pathology of hidradenitis suppurativa is disputed, for unknown reasons; people with hidradenitis develop plugging or clogging of their apocrine glands. Follicles into which the apocrine glands open are plugged by keratin comedones and infection occurs following stasis. (Keratin comedones--- Occlusion of the apocrine ducts--- Superimposed inflammation and infection--- Abscess formation--- Chronic infection and spread--- Indurations and sinus and fistula formation)

The exact cause of hidradenitis suppurativa remains unclear. Obesity, Tight clothing, Smoking, Deodorants, depilation products, shaving of the affected area, oral contraceptives and lithium, are thought to play a role in the development of hidradenitis suppurativa. Genetic factors and endocrine factors are also responsible for the occurrence of the disease. Hidradenitis suppurativa is diagnosed clinically based on its appearance. There are no lab tests or biopsies that establish the diagnosis. Streptococci, staphylococci, and Escherichia coli have been identified in the early stages of the disease; however, in the chronic relapsing stages, anaerobic bacteria and Proteus species have more commonly been isolated. Whether the bacteria are the cause or the result of the disease has not been determined. There is no cure for hidradenitis suppurativa. However, several treatment options are available, including preventive, medical, surgical, and psychological strategies, early treatment can help manage the symptoms and to prevent new lesions from developing. Mild to moderate cases can be treated with self-care measures, including warm compression and regular washings with antibacterial soap, and topical or oral medications. However, recurrent, severe cases requires surgical intervention.

Case History:
An 18-year old girl was admitted in the plastic surgery unit of Shaheed Sohrawardi Medical College Hospital, Dhaka with the complaints of swelling and pain in the sternal region for the last six years which subsequently extended to the sub-mammary region, these swelling manifested as spontaneous bursting with purulent to serosanguinous discharge. She was treated with inadequate antibiotics and chronicity developed. Surgical intervention in the form of incision and drainage was done 4 years back which led to formation of sinus and fistula. She also developed similar painful swelling in both axilla 4 years back. She gave no history of fever, anorexia, cough, bony pain, significant weight loss, and bowel or bladder abnormalities. She has no history of diabetes mellitus, hypertension, jaundice or bronchial asthma.

Fig-1: Hydradenitis suppurativa. Lesion on Pre-sternal region

On general examination the patient was conscious, co-operative, her pulse, blood pressure and respiratory rate was normal; there was no accessible palpable lymph nodes, no engorged neck gland or neck vein. On local examination there were multiple discharging sinus in both axilla, sternal and sub-mammary region with...
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Fig. 2: Hydradenitis suppurativa. Lesion on pre-sternal and submammary region.

Fig. 3: Hydradenitis suppurativa. Lesion on both axillas.

Fig. 4: Marking of the excision of the lesions.

a broad scar on pre-sternal region, which were tender on touch and indurated, there were serosanguinous discharge through the sinuses. The local temperature of the overlying skin was normal. All other systemic examination reveals no abnormality.

She was first operated under general anesthesia on May 2010 for the sternal and sub-mammary region. Wide excision of the skin and involved sub-cutaneous tissue and the defect was closed primarily by local advancement flap, a drain was kept in situ for 7 days, on June 2010 her both axillary lesions were excised and defects were closed in a similar manner with local advancement

Fig. 5: Defects following excision of the lesion on pre-sternal and submammary region.

Fig. 6A: Defects were reconstructed with local advancement flap.
Discussion:
Hidradenitis suppurativa is a physically, psychologically, and socially disabling, inflammatory skin disease that is characterized by recurrent boil-like lumps (abscesses) that culminate in pus-like discharge, difficult-to-heal open wounds and scarring. The disease has a propensity to become chronic and indolent because of subcutaneous extension leading to indurations, sinus, and fistula formation. Because this is considered a rare disease, its incidence rate is not well known, but has been estimated as being between 1:24 (4.1%) and 1:600 (0.2%).

Women are affected by hidradenitis suppurativa three times as often as men; the reason for this is unknown. The condition most commonly occurs between 20-40 years and coincides with the post-pubertal increase in androgen levels, but the untreated disease may persist into the seventh decade. Disease onset rarely occurs before puberty and after menopause. Flare-ups have been linked with menses; shorter menstrual cycles and longer duration of menstrual flow are associated with the disease. Genetic component may be an important aetiological factor in the causation of disease. In one study of 110 patients, 38 percent reported to have a family history of the disease. This may reflect a familial form with autosomal dominant inheritance.

As this disease is poorly studied, the causes are controversial, but there are certain triggering and pre-disposing factors that should be taken into consideration, as it is advisable to avoid such triggers.

Following factors are responsible for flaring of the disease:
- Obesity is an exacerbating rather than a triggering factors through mechanical irritation, occlusion, and maceration.
- Tight clothing and clothing made of heavy, non-breathable materials.
- Smoking tobacco products.
- Deodorants, depilation products, shaving of the affected area - their association with Hidradenitis suppurativa is still an ongoing debate amongst researchers.
- Drugs, in particular oral contraceptives and lithium.
- Genetic factors: an autosomal dominant inheritance pattern has been postulated.
- Endocrine factors: sex hormones, principally an excess of androgens, are thought to be involved, although the apocrine glands are not sensitive to these hormones. Women often have outbreaks before menstruation and post-pregnancy, and the disease usually remits during pregnancy and after menopause.

Hidradenitis suppurativa often starts at puberty with a single, painful bump that persists for weeks or months. The extent and severity of the disorder varies widely between individuals. Initially firm pea-sized nodule (0.5-1.5 cm diameter) resembling acne may appear on one site. These lesions may resolve spontaneously or within hours to days to rupture and ooze a pus-like discharge. These may heal without treatment but at a later time new lesions recur in the adjacent area. If uncontrolled, this leads to development of larger lesions (golf ball size), sinus tract formation, and involvement.
of multiple sites. Three distinct clinical stages have been defined for the condition.1,2

- Stage 1 – solitary or multiple, isolated abscess formation without scarring or sinus tracts
- Stage 2 – recurrent abscesses, single or multiple widely separated lesions, with sinus tract formation
- Stage 3 – diffuse or broad involvement, with multiple interconnected sinus tracts and abscesses.

Potential complications of hidradenitis suppurativa include dermal contraction, local or systemic infection resulting from the spread of micro-organisms, arthritis secondary to inflammatory injury, squamous cell carcinoma (in indolent sinus tracts), disseminated infection (rare), restricted limb mobility from scarring, lymphedema caused by lymphatic injury from inflammation and scarring, rectal or urethral fistulas, systemic amyloidosis, and anemia from chronic infection.28,29

There is no cure for hidradenitis suppurativa. However, several treatment options are available, including preventive, medical, surgical, and psychological strategies. Because of the variety of ways in which the disease can manifest and progress, treatment should be based on the patient’s presentation and circumstances. Obesity, incomplete removal and ongoing skin infections can increase the chances that hidradenitis suppurativa returns, even after surgical treatment. All patients also should be offered reassurance and psychosocial support. In one study,22,30 24 percent of patients found nothing to help their condition, despite an average disease duration of almost 19 years. Deodorants, shaving and depilation have not been supported as a cause for this discrepancy in at least one study.26 However; these should be avoided if they cause irritation. Warm compression, topical antiseptics, and antibacterial soap may help in patients with folliculitis. Patient should be reassured that hidradenitis suppurativa is not caused by poor hygiene and is not contagious.30 As excessive underarm adiposity advances the progression of hidradenitis by creating an ideal environment for bacterial growth and friction. Therefore, weight loss may prevent the progression. Friction from clothing increases pain and discomfort, and patients should avoid wearing tight, synthetic clothing near the affected areas. Heat and humidity also have been associated with flare-ups, and prolonged exposure to hot, humid climates should be avoided if possible. Stress management methods may be useful because the disease can be aggravated during times of increased psychosocial stress.30

Initial treatment of hidradenitis suppurativa can begin with conservative measures such as warm baths, hydrotherapy, and topical cleansing agents to reduce bacterial load.13 Nonsteroidal anti-inflammatory drugs may alleviate pain as well as inflammation. Antibiotics, although not proven to be effective, are the mainstay of medical treatment, especially for lesions suspected of being superinfected. Empiric antibiotic treatment may be given when conservative measures with several days’ observation have not improved symptoms. Treatment can begin with topical or systemic antibiotics, or both. The only topical antibiotic that has been proven effective in a randomized controlled trial is clindamycin.8 Antistaphylococcal agents are best for axillary disease, and more broad-spectrum coverage is better for perineal disease. Dicloxacillin (1 to 2 g daily), erythromycin (1 g daily), tetracycline (1 g daily), and minocycline (1 g daily) have been used. Cephalosporins may be helpful for concurrent active cellulitis.31 For severe, recurrent disease, anecdotal evidence suggests that two months or more of antibiotic therapy may be needed to prevent progression and worsening of concomitant infection.22 Other medical treatment options include oral contraceptive agents that contain a high estrogen-to-progesterone ratio and low androgenicity of progesterone.32 Two patients with severe, long-standing disease benefited from finasteride (Proscar) at a dosage of 5 mg daily.33 In Europe, the antiandrogen cyproterone acetate (Cyprostat) has been successful in some studies,34 but it currently is not approved for use in the United States.
Oral retinoids, which work by inhibiting sebaceous gland function and abnormal keratinization, also have been used. Pre-treatment with isotretinoin (Accutane) at a dosage of 0.5 to 1.0 mg per kg daily for a few months before surgery has been recommended to reduce the inflammatory components. No consensus on the dosing and duration of isotretinoin therapy has been reached by the few studies that have investigated it as a possible therapeutic option. Side effects of isotretinoin remain a major issue and include birth defects, hepatotoxicity, pseudotumor cerebri, and aggression.

Corticosteroids and immunosuppressants are other treatment possibilities. Topical triamcinolone (Aristocort) may be an option, but insufficient research has been conducted for it to be recommended routinely. Oral cyclosporine (Sandimmune) has shown some benefit, but chronic treatment can cause serious toxicity.

For early, limited disease that presents with a fluctuant abscess, incision and drainage may be a good first option. When hidradenitis sinus tracks are well established but relatively superficial, they can be unroofed or laid open. Because these tracks are lined by epithelium, the floor of the track can be preserved; this facilitates the rapid healing and minimizes scarring.

Early, rather than delayed, wide excisional therapy has been recommended by some experts as the treatment of choice because repeated failed treatments lead to the disease being more widespread and severe at presentation, making surgical options more difficult. Patients should be advised that surgery treats only the disease that is present at the site of the excision; recurrence at a new site is possible. In one study 39 of 82 patients treated with wide excision, recurrence rates were zero percent for perianal disease, 3 percent for axillary disease, and 37 percent for inguinoperineal disease. Obesity, insufficient excision, significant skin maceration, and chronic skin infection may increase the incidence of recurrence. In another study, the overall complication rate was 17.8 percent; most complications were minor, such as suture dehiscence, postoperative bleeding, and hematoma. The rate of recurrence in this study was 2.5 percent and was related to the severity of the disorder.

Radiotherapy has been investigated as a potential treatment option. In a study the effects of radiotherapy in 231 patients, 38 percent had complete relief, and 40 percent showed clear improvement of symptoms. However, the possibility of long-term side effects must be discussed thoroughly with the patient. Cryotherapy also has been considered. In one small study, 10 patients who did not respond to systemic antibiotics were given one cycle of cryotherapy; eight patients reported improvement. However, patients also experienced significant pain, prolonged healing time (average, 25 days), and post-treatment infection. A carbon dioxide laser used in conjunction with second-intention healing provided relief for a few patients.

In addition to treating the physical illness, it is crucial that physicians acknowledge and treat the psychological burden associated with the disease. Because of the areas of the body that are affected, the malodorous discharge, the chronic discomfort, and the general unsightliness of the disease, hidradenitis suppurativa poses many challenges for patients in their personal life. Sexuality can be negatively affected. Unforgiving societal attitudes regarding inappropriate body odor (especially for those who choose not to wear deodorants), as well as years of inadequate treatment, may lead to frustration, depression, and isolation. For patients at increased risk for these outcomes, early surgical intervention should be strongly considered.

Conclusion:

Hidradenitis suppurativa remains a challenging disease for patients and physicians. Because there has been no significant research comparing treatment options, the choice of therapy should depend on the patient’s circumstances and preferences, the outcome of previous treatments, the experience of the physician, local expertise
A teenage girl with extensive form of Hidradenitis suppurativa of sternal, submammary region and both axilla (e.g., a surgeon or dermatologist who specializes in treatment of the disease), and the chronicity and severity at presentation. Further research should be conducted not only on the etiology of this disease but also on the optimal treatment regimen.

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