Case Report

A patient with multiple recurrent abscesses in the axillae and buttocks

患者腋下及臀部多發性複發性膿腫

AWM Au 區慧明, WYM Tang 鄧旭明, WY Lam 林永賢

A 31-year-old man presented with multiple recurrent painful abscesses over both axillae and buttocks for more than 15 years. He also suffered from acne conglobata and scarring alopecia over the scalp. Clinical features and skin biopsy confirmed the diagnosis of follicular occlusion tetrad.

31歲男性患者15年來於雙側腋下及臀部出現復發性痛性膿腫。患者亦同時患上疥性痤瘡及頭皮疤痕性斑禿。經臨床及皮膚病組織檢查確診為毛囊閉塞性四聯徵。

Keywords: Follicular occlusion tetrad, Hidradenitis suppurativa

關鍵詞：毛囊閉塞性四聯徵，化膿性汗腺炎

Introduction

Follicular occlusion tetrad is a condition including hidradenitis suppurativa, acne conglobata, dissecting cellulitis of scalp and pilonidal sinus. The exact pathogenesis of this group of disease is unknown but evidence suggests that they share the same pathological process initiated by follicular occlusion. Hidradenitis suppurativa (HS) is an uncommon disease affecting the axillae and anogenital region. It is a chronic condition characterised by recurrent painful abscesses and draining sinus tracts with subsequent scarring. It is notoriously resistant to treatment and can cause significant debilitation.

Case report

A 31-year-old Chinese man presented with recurrent painful abscesses over both axillae and buttocks since age 14 to 15, and had been treated with short courses of antibiotics in the
private sector. He also developed nodulocystic acne over the face and chest in his later teenage years. He did not seek medical advice again until recently, when the abscesses over the axillae were too painful to bear. Incision and drainage was done by an orthopaedic surgeon.

On physical examination, the patient was obese and both axillae revealed wounds after incision and drainage with inflammation, purulent discharge and scarring (Figure 1). Multiple tender inflammatory nodules with comedones and varying degrees of scarring were also found over the back, chest and sacral area (Figure 2). In addition, multiple inflammatory nodules and cystic acne were present over the face (Figure 3) with scarring alopecia over scalp (Figure 4). The differential diagnoses included hidradenitis suppurativa, infected epidermal cyst, and multiple skin abscesses.

An incisional biopsy was done over the left sacral region. Histology showed dermal scarring with inflammation and overlying granulation tissue and reactive epidermis, which could be present in all of the above differential diagnoses. Subsequently, a second biopsy over a new nodule over the left shoulder was performed. This biopsy showed nodular lesions with underlying suppurative folliculitis, ruptured hair

Figure 1. A wound with scarring and discharge in the axilla.

Figure 2. Eroded nodular lesions at the buttock.

Figure 3. Acne conglobata over the face and neck.

Figure 4. Inflammatory nodules over the occipital scalp with scarring alopecia.
folicles, adjacent dermal abscesses, granulomatous and fibrous perifolliculitis (Figures 5 & 6). No fungal elements were seen. These features were compatible with hidradenitis suppurativa and the constellation of features was consistent with the follicular occlusion tetrad.

Discussion

In 1956, Pillsbury, Shelly and Kligman used the term follicular occlusion triad for the common association of hidradenitis suppurativa, acne conglobata and dissecting cellulitis of the scalp. They proposed that the pathological event unique to each disease was follicular hyperkeratinization. In 1975, the presence of a pilonidal sinus was added to the triad, producing the term follicular occlusion tetrad. The term acne inversa is sometimes used as an inclusive term for this group of disorders.

Hidradenitis suppurativa (HS) usually starts at or soon after puberty. Women are three times more commonly affected than men. The pathogenesis and aetiology are unknown, but is thought to originate from poral occlusion of the pilosebaceous units. Subsequent occlusive spongiform infundibulofolliculitis weakens the infundibular wall. Thereafter, ruptured follicles spill their contents, including keratin and bacteria, into the surrounding dermis. This excites a vigorous chemotactic response and abscess formation. Epithelial strands are generated, possibly from ruptured follicular epithelia, and form sinus tracts.

Hidradenitis suppurativa is thought to be related to hyperandrogenism since HS is seldom seen before puberty. Childhood cases have been associated with precocious puberty. However, reports show that female patients with HS have no evidence of biochemical hyperandrogenism. Therefore, the role of androgen in HS probably lies in end-organ sensitivity rather than with plasma levels of androgen. Patients who are obese or overweight tend to have more severe disease. A familial form with autosomal dominant inheritance has been described.

The clinical features are mainly inflammatory nodules and sterile abscesses in the axillae, groin, perianal and inframammary areas. These lesions are very painful and tender. Sinus tracts and hypertrophic scars may develop with time. Discharged fluid is often a mixture of serous exudates, blood and pus, and is frequently malodorous. Complications include anaemia, secondary amyloidosis, hypoproteinaemia as a

Figure 5. Suppurative folliculitis, follicular rupture and granulomatous folliculitis. (H&E, Original magnification x 100)

Figure 6. Dermal abscess consequent to destruction following suppurative folliculitis. (H&E, Original magnification x 40)
result of chronic inflammation, fistula into urethra, bladder and peritoneum, arthropathy, malaise, depression, and lymphatic obstruction resulting in lymphoedema of limbs and genitalia. Squamous cell carcinomas, sometimes with metastasis have also been reported. 

The differential diagnoses include those conditions that produce recurrent abscesses and sinus tract formation with a characteristic distribution in the intertriginous skin, namely follicular pyodermas, granuloma inguinale, Crohn's disease and acne. Histological findings include spongiform infundibulofolliculitis, occlusion of the infundibular portion of the follicle, dilation of the follicle with stasis in apocrine glands, cystic epithelium-lined structures containing hair shafts, abscesses formation leading to destruction of pilosebaceous units, epithelial strands forming sinus tracts and subcutaneous abscesses and inflammation with extensive fibrosis.

Treatment of hidradenitis suppurativa is difficult and is at present non-curative. The goals of treatment are to reduce existing lesions and prevent development of new lesions. So far, there has not been any large-scale randomised controlled trial comparing the treatment modalities, and no single treatment has been found to be effective for all patients. General measures include reducing further trauma to the involved area by avoiding heat and humidity, friction from clothing, and sweating. Weight reduction and smoking cessation should be advised. Use of antibacterial cleansers and antiperspirants may be helpful but should be avoided if irritation occurs. Systemic antibiotics such as tetracycline, and dapsone have been used with some effects. However, the condition usually recurs after withdrawal of treatment. Anti-androgens have been used with variable effects. Combination of cyproterone acetate and ethinyl estradiol has shown some effects in women. Finasteride 5 mg daily has also shown promising results. Since finasteride is a teratogen, it should not be used in pregnant women. Immunosuppressants such as systemic steroid and cyclosporine have been used for control of an acute flare. Their use is limited because of significant side effects and the lack of long term benefits. Intralosomal triamcinolone has been used in isolated painful lesions but is not appropriate for extensive disease. Retinoids reduce sebum production and regulate cell proliferation and differentiation, and its use is hoped to correct the follicular abnormality that initiates the disease. Isotretinoin (0.5-1.2 mg per kg per day) has been used in multiple case series with variable results.

Tumour necrosis factor-alpha (TNF-α) inhibitors have been used in patients with Crohn's disease and other chronic inflammatory diseases. Promising results have been reported with infliximab in a case of hidradenitis suppurativa with coexisting Crohn's disease. Additional case series reporting the use of infliximab and etanercept have demonstrated favourable results. However, more studies would be needed to determine long term effects and side effects.

Surgical treatment includes incision and drainage, unroofing and debriding cysts and wide excision. Among these surgical modalities, wide excision is the most effective treatment for chronic and extensive disease. Other modalities such as carbon dioxide laser, zinc, photodynamic therapy, botulinum toxin A, cryotherapy and methotrexate were also used in isolated case reports.

References
Follicular occlusion tetrad


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Announcement

Application for Annual / Exit Assessment, December 2008
Specialty Board of Dermatology & Venereology
Hong Kong College of Physicians

Please be reminded that the application for the Annual / Exit Assessment, December 2008 is now open to the eligible candidates, who should be:

1. Registered trainees in Dermatology & Venereology, Hong Kong College of Physicians
2. Qualified for / will be able to qualify of the Annual / Exit Assessment by 30 March 2009

Those who wish to attend the Assessment should complete the Higher Physician Training (HPT) Annual Assessment Application Form / Higher Physician Training (HPT) Exit Assessment Application Form plus Testimonial, to the Examination Co-ordinator of the Specialty Board of Dermatology & Venereology in **July 2008**. Late applicant will not be able to sit for the assessment.

Dr. TANG Yuk-ming
Chairman
Specialty Board (Dermatology & Venereology)
Hong Kong College of Physicians