

Perifolliculitis capitis abscedens et suffodiens : a case report

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ABSTRACT :

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Perifolliculitis capitis abscedens et suffodiens (PCAS) is one of primary cicatricial alopecia. The common manifestation is perifollicular lesion with sinus tracts formation. Follicular hyperkeratosis with obstruction and bacterial infection play a role in the pathogenesis. The treatment options consist of medical and surgical treatments. Nevertheless, PCAS is usually difficult and often recalcitrant. In this report, we describe the patient with PCAS. A 28 year-old male with recurrent multiple painful erythematous papules, pustules, nodules and abscesses with hair loss on the scalp. Histopathological finding is compatible with Perifolliculitis capitis abscedens et suffodiens.

Key words: Perifolliculitis capitis abscedens et suffodiens, dissecting folliculitis, dissecting cellulitis, Hoffman's disease

บทคัดย่อ :

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**หน่วยโรคผิวหนัง ภาควิชาอายุรศาสตร์ คณะแพทยศาสตร์รามธิบดี มหาวิทยาลัยมหิดล

Perifolliculitis capitis abscedens et suffodiens เป็นโรคผมร่วงชนิดมีแผลเป็น อาการแสดงสำคัญ คือ พบการอักเสบรอบรูขุมขนและเกิดท่อเชื่อมต่อกับบริเวณหนังศีรษะ พยาธิกำเนิดเชื่อว่าเกิดจากการเพิ่มจำนวนของชั้นผิวหนังกำพร้าบริเวณรูขุมขนทำให้มีการอุดตันและเกิดการติดเชื้อแบคทีเรีย แนวทางการรักษาสามารถเลือกวิธีการใช้ยาหรือการผ่าตัด ผลการรักษาค่อนข้างได้ผลน้อยและมีแนวโน้มต่อการรักษา รายงานฉบับนี้เป็นการนำเสนอผู้ป่วยชายอายุ 28 ปี มาตรวจรักษาเรื่องผมร่วงร่วมกับมีการอักเสบเป็นตุ่มและฝีที่บริเวณหนังศีรษะ ผลตรวจทางจุลพยาธิวิทยาเข้าได้กับโรค

Perifolliculitis capitis abscedens et suffodiens

คำสำคัญ : โรคผมร่วงชนิดมีแผลเป็น

Case report

A 28 year-old Thai male patient from Bangkok presented with a 12-year history of recurrent multiple painful erythematous papules, pustules, nodules and abscesses with hair loss on the scalp. An examination revealed multiple discrete and confluent skin color and erythematous nodules, pustules with scarring alopecia at the occipital, vertex, frontal and bitemporal area with some sinus tracts (Figures 1). Hair pulling test was negative.

Histopathological finding showed irregular acanthosis with compact orthokeratosis of the epidermis. Dilated follicular infundibulum with intra- and perifollicular neutrophilic infiltration was found. There was a sinus tract formation surrounded by dermal fibrosis. There were perivascular and interstitial infiltration with mixed inflammatory cells (Figures 2). The provisional diagnosis was perifolliculitis capitis abscedens et suffodiens.

The patient was treated with oral isotretinoin 10 mg per day. At one month, he had good clinical response and at ten months follow-up visit, he had no active disease.



Figure 1 A,B Multiple discrete and confluent skin color, erythematous nodule, pustule, some sinus tracts with scarring alopecia

Discussion

Perifolliculitis capitis abscedens et suffodiens (PCAS) is also known as dissecting folliculitis or dissecting cellulitis or dissecting perifolliculitis of the scalp or Hoffman's disease. It was denominated by Hoffman in 1908.¹ It is an uncommon, chronic, progressive, inflammatory, suppurative scalp disorder. It represents 1-2% of the primary cicatricial alopecia that mainly affects African American males between 18-40 years of age.^{1,2}

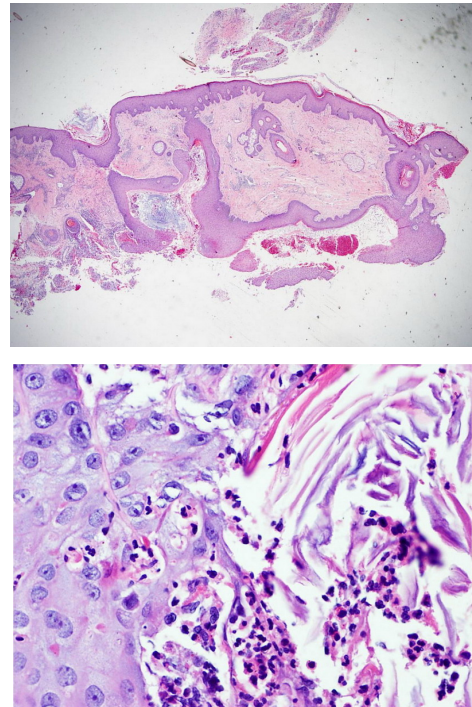


Figure 2 A,B Dilated follicular infundibulum with intra- and perifollicular neutrophilic infiltration and a sinus tract formation surrounded by dermal fibrosis

PCAS manifests with perifollicular pustules, nodules, abscesses interconnecting sinus tracts on the vertex and occipital scalp. It can wax and wane in severities for many years, eventually resulting in dense dermal fibrosis, sinus tract formation and finally, extensive scarring alopecia occurs.^{2,3} Unusual complications were reported that include osteomyelitis, squamous cell carcinoma and spondyloarthropathies.^{4,5}

PCAS may present as an isolated disease or within "the follicular occlusion triad", associated with hidradenitis suppurativa and acne conglobata and when also with pilonidal cysts or piloid cysts referred to as the follicular occlusion tetrad. All these diseases have the

same pathogenesis and the same histological changes. The pathogenesis is follicular hyperkeratosis with retention of follicular products and subsequent granulomatous response to keratin debris and occasional superimposed by bacterial infection usually with *Staphylococcus aureus* or *Staphylococcus epidermidis*. For now, the primary etiology of this disease remains inconclusive.^{2,3,6,7}

Diagnosis is based on the presence of fluctuated nodules and abscesses on the scalp, sometimes with sinus tract formation and seropurulent discharge. The course is often extended, and marked scarring alopecia may follow. The differential diagnosis includes neutrophilic cicatricial alopecias (folliculitis decalvans), lymphocytic cicatricial alopecias (lichen planopilalis, discoid lupus erythematosus) and mixed cell cicatricial alopecias (keloidalis nuchae). The histopathology of PCAS is well described, although the features are not diagnostic and depend on the stage of the disease. Early lesions will demonstrate follicular plugging; later, a perifolliculitis develops with dense infiltration of neutrophils, lymphocytes, and histiocytes. Foreign body giant cells from the ruptured follicles may be seen. In older or healing lesions, sinus tracts develop and extensive fibrosis replaces the inflammatory infiltration and normal dermal structures.⁸⁻¹⁰

The treatment options of PCAS consist of medical and surgical treatments. However, it is usually difficult and often recalcitrant. Medical therapies aimed to control the inflammatory process include: topical,

intralesional and systemic corticosteroids, topical and oral isotretinoin, oral antibiotics (ciprofloxacin, rifampicin, antimalarial drugs), dapsone, zinc supplements and some biological therapies such as infliximab, adalimumab. Surgical therapies include: incision and drainage with marsupialization, total scalp excision followed by skin grafting, laser treatments and radiation therapy are used when failure of medical therapy.^{1,6,8,11,12}

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