

New Approach in Combined Therapy of Perifolliculitis Capitis Abscedens et Suffodiens

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Perifolliculitis capitis abscedens et suffodiens (PCAS) is a rare scalp disease seen mostly in young males of Afro-American descent. The disease is characterized by perifollicular pustules, suppurative nodules and fluctuating abscesses, as well as by intercommunicating sinus tracts on the scalp. We report here an elderly Caucasian woman who fulfilled the clinical and histological criteria for diagnosis of PCAS.

The patient was treated with initial prednisolone, systemic acitretin, topical tacrolimus and oral zinc. After 6 months, her condition was stable.

CASE REPORT

An 86-year-old woman reported having had purulent scalp lesions for approximately one year. She denied trauma or a preceding herpes zoster. Topical corticosteroids, as well as systemic amoxicillin/clavulanic acid, produced no improvement. A large draining boggy mass evolved on her occiput.

Clinically, a 5 × 5 cm weeping, ulcerated nodule, with a raised border and numerous sinus tracts, was seen (Fig. 1A). In addition, pustules were scattered over her scalp; some of which had evolved into fluctuant nodules with overlying alopecia.

Laboratory results showed a leukocyte count of 10.7/nl (normal range 4.0–10.0/nl) and a C-reactive protein concentration of 2.8 mg/dl (normal: <0.5 mg/dl). In addition, the blood level of zinc was slightly decreased (8.9 µmol/l; normal 9.2–18.4 µmol/l). The remaining laboratory parameters were within the normal range.

A search for circulating autoantibodies specific for pemphigus and pemphigoid by ELISA was negative. The virus-specific antibody counts for varicella-zoster and herpes simplex virus were IgG positive and IgM negative. PCR failed to detect *Treponema pallidum*, *Bartonella*, *Mycoplasma* and *Mycobacteria*.

A skin biopsy of the occipital region showed dissecting dermatitis with abscess formation (Fig. 2). The dermis was dominated by a neutrophilic inflammatory infiltrate. Terminal hair shafts were variably destroyed and entered into a sinus

tract. In the upper dermis there was a suppurative inflammation with abscess material and hair shaft fragments being discharged via a draining sinus. Periodic acid-Schiff (PAS) staining for fungi, as well as immunohistochemical staining with anti-*Mycobacterium bovis* (BCG) and for *T. pallidum* was negative. Direct immunofluorescence of the perilesional scalp skin was negative.

Microbiological examinations failed to identify pathogenic organisms.

Following the diagnosis of a PCAS, the patient was treated with 10 mg acitretin and 30 mg prednisolone daily as an inpatient. The prednisolone was reduced to 5 mg per day during the course of treatment. Because of the decreased level of zinc, 100 mg zinc aspartate was given daily. In addition, topical glucocorticoids and tacrolimus 0.1% were alternated.

This regimen produced almost total healing within 11 days (Fig. 1B). The systemic corticosteroids were further tapered and the topical corticosteroids were stopped.

Six months after discharge the patient's skin condition was stable on a regimen of 10 mg acitretin and 100 mg zinc aspartate daily, supplemented by topical therapy with tacrolimus 0.1%.

DISCUSSION

PCAS was described for the first time in 1903 by Spitzer (1) and named by Hoffmann (2) in 1908. It is a disease of unknown aetiology that mainly affects young Afro-American males (3, 4). While the disease has been observed in Caucasian women in the age range 20–40 years (4, 5), to the best of our knowledge this is the first report of PCAS in an elderly Caucasian woman. The diagnosis was made on the basis of clinical and histopathological characteristics and after excluding other possible differential diagnostic considerations.

The characteristic symptoms of PCAS are perifolli-



Fig. 1. Skin condition before treatment. (A) Weeping, ulcerated nodule with a raised border. (B) Skin condition after treatment: nearly total healing.

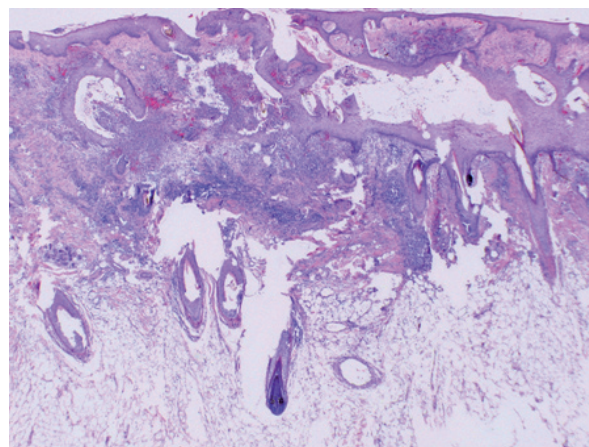


Fig. 2. Occipital skin biopsy with destruction of hair follicles, abscess formation and a draining sinus tract. Hematoxylin & eosin staining × 40.

cular pustules, suppurative nodules and fluctuating abscesses, as well as extensive interconnecting sinus tracts that may drain pus and blood. The disease occurs most often on the vertex and occipital region (4, 5).

The disease is usually chronic, but may wax and wane with periods of inactivity. Sometimes healing with hypertrophic or keloidal scarring occurs. PCAS can also lead to scarring alopecia. Rarely squamous cell carcinoma or secondary amyloidosis can develop in chronic inflammatory skin lesions. PCAS is also associated with acne inversa (3–6).

In histopathology, a folliculitis and perifolliculitis with a heavy infiltrate of neutrophils, leading to abscess formation in the dermis, can be seen. Draining sinuses develop in later lesions going along with a destruction of follicles. Both direct immunofluorescence and microbiological examinations are negative (3, 4).

PCAS is often mistaken for folliculitis decalvans. In elderly patients, erosive pustular dermatosis of the scalp should be excluded. Both diseases show neither abscesses nor sinus tracts. Thus they can be distinguished histologically and clinically from PCAS (7, 8).

In addition, the vegetative form of the pyoderma gangraenosum is a differential diagnostic consideration. The histopathology of this disease is characterized by a pseudoepitheliomatous hyperplasia, dermal abscesses, sinus tracts and a palisading granulomatous reaction. However, a pseudoepitheliomatous hyperplasia and a palisading granulomatous reaction were absent in our case. In addition, no preceding traumas or bacterial infections were found in an immunocompromised patient (9). Thus, we preferred the diagnosis of PCAS.

PCAS is best treated systemically. Isotretinoin has often proven helpful (3–5). Ljubojevic et al. (4) reported low-dose treatment regimen for PCAS, starting with an initial dose of 30 mg of isotretinoin and continuing with 10 mg daily over 10 months. There are many reports describing the use of topical, intralesional and systemic corticosteroids, especially for acute flares (3, 5).

In addition, there are reports of successful treatment with doxycycline (10), ciprofloxacin (11), rifampicin (12), dapson (3), and adalimumab (13) as well as infliximab (14). Berne et al. (15) reported healing after zinc substitution therapy for at least 3 months.

As far as we know, there has not been a case reported that was treated successfully with maintenance therapy combining topical tacrolimus with oral low-dose aci-

tretin and zinc. The new approach in combined therapy may be an option for the treatment of PCAS.

REFERENCES

1. Spitzer L. Dermatitis follicularis et perifolliculitis conglobata. *Dermatol Z* 1903; 10: 109–120.
2. Hoffmann E. Perifolliculitis capitis et suffodiens: case presentation. *Dermatol Z* 1908; 15: 122–123.
3. Bolz S, Jappe U, Hartschuh W. Successful treatment of perifolliculitis capitis abscedens et suffodiens with combined isotretinoin and dapson. *J Dtsch Dermatol Ges* 2008; 6: 44–47.
4. Ljubojevic S, Pasic A, Lipozencic J, Skerlev M. Perifolliculitis capitis abscedens et suffodiens. *J Eur Acad Dermatol Venereol* 2005; 19: 719–721.
5. Karpouzis A, Giatromanolaki A, Sivridis E, Kouskoukis C. Perifolliculitis capitis abscedens et suffodiens successfully controlled with topical isotretinoin. *Eur J Dermatol* 2003; 13: 192–195.
6. Curry SS, Gaither DH, King LE Jr. Squamous cell carcinoma arising in dissecting perifolliculitis of the scalp. A case report and review of secondary squamous cell carcinomas. *J Am Acad Dermatol* 1981; 4: 673–678.
7. Sperling LC. Scarring alopecia and the dermatopathologist. *J Cutan Pathol* 2001; 28: 333–342.
8. Mastroianni A, Cota C, Ardigò M, Minutilli E, Berardesca E. Erosive pustular dermatosis of the scalp: a case report and review of the literature. *Dermatology* 2005; 211: 273–276.
9. Papadopoulos AJ, Schwartz RA, Kapila R, Samady JA, Ruszczak Z, Rao BK, Lambert WC. Pyoderma vegetans. *J Cutan Med Surg* 2001; 5: 223–227.
10. Moscatelli P, Ippoliti D, Bergamo F, Piazza P. Guess what. Perifolliculitis capitis abscedens et suffodiens. *Eur J Dermatol* 2001; 11: 155–156.
11. Onderdijk AJ, Boer J. Successful treatment of dissecting cellulitis with ciprofloxacin. *Clin Exp Dermatol* 2010; 35: 440.
12. Georgala S, Korfitis C, Ioannidou D, Alestas T, Kylafis G, Georgala C. Dissecting cellulitis of the scalp treated with rifampicin and isotretinoin: case reports. *Cutis* 2008; 82: 195–198.
13. Navarini AA, Trüeb RM. 3 cases of dissecting cellulitis of the scalp treated with adalimumab: control of inflammation within residual structural disease. *Arch Dermatol* 2010; 146: 517–520.
14. Brandt HR, Malheiros AP, Teixeira MG, Machado MC. Perifolliculitis capitis abscedens et suffodiens successfully controlled with infliximab. *Br J Dermatol* 2008; 159: 506–507.
15. Berne B, Venge P, Ohman S. Perifolliculitis capitis abscedens et suffodiens (Hoffman). Complete healing associated with oral zinc therapy. *Arch Dermatol* 1985; 121: 1028–1030.