CICATRICIAL ALOPECIA – A CASE REPORT WITH A REVIEW OF LITERATURE

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ABSTRACT

Cicatricial alopecia encompasses a group of clinical entities that affect the hair unit leaving permanent destruction of the follicular ostia, skin atrophy and irreversible loss of hair follicles. Herein, a 43-year-old lady with erythematous follicular papules, resulting into atrophic scars with permanent loss of hairs, slowly progressing for more than 22 years, is presented. The patient has been consulted by numerous dermatologists and no exact histological diagnosis was verified. Upon proper clinico-pathological work-up, a diagnosis of lichen planopilaris was reached and treatment with intralesional corticosteroids and oral hydroxychloroquine was introduced. The patient improved significantly during the 3-month follow-up. Scarring alopecia should always be evaluated clinically and by scalp biopsy to accurately analyze the change in the follicular architecture and type, distribution and extent of the inflammatory infiltrate. This complex diagnostic approach is crucial for the diagnosis and management of all cases of cicatricial alopecias. Scr Sci Med. 2017;50(1):41-44

Keywords: *cicatricial alopecia, histopathological findings, differential diagnosis*

INTRODUCTION

Cicatricial alopecia describes a group of hair disorders that result in fibrous tissue replacement of the hair follicles (1). In addition to secondary scarring alopecias, caused by some dermatophytic infections, metastatic infiltrations and mechanical, physical or chemical triggers there are also primary ones, also named idiopathic, which probably result from certain defects of hair follicle structural proteins or an abnormal immune response. Other hypotheses suggest disruption of the mesenchymal/epithelial interaction or disturbance of the nutrition apparatus of the hair follicle. The process is usually chronic and slowly advancing, causing a great psychological burden to the affected individuals (2).

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Case REPORT

A 43-year-old woman presented with a history of erythematous patches with pityriasiform scaling, resulting in atrophic areas of missing follicular ostia, on the frontal and parietal scalp. The skin lesions persistently evolved in the past 22 years, slowly advancing, regardless the topical corticosteroid treatments, tacrolimus, revulsive lotions and some alternative therapeutic approaches such as acupuncture that were introduced. Severe burning and pruritus

were constant subjective symptoms. No histological examination has been done so far. At the time of the physical examination diffuse atrophic plaque with occasionally spared follicles was involving the parietal zone. Perifollicular erythematous papules with tiny peripheral scaling demarcated the lesional borders (Fig. 1).

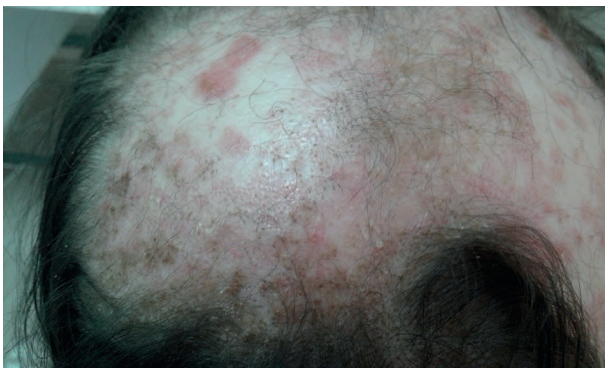

Fig. 1. Diffuse atrophic plaque with occasionally spared follicles and perifollicular erythematous papules with tiny peripheral scaling at the edge of the area involved

A 4-cm vertical punch biopsy showed compact hyperkeratosis, hypergranulosis, pseudoacanthosis, vacuolar degeneration of the basal keratinocytes at the sites of follicular ostia and prominent diffuse fibrosis with plenty of fibrous tracts, replacing the pre-existing follicular units (Fig. 2). The pathological changes were consistent with lichen planopilaris.

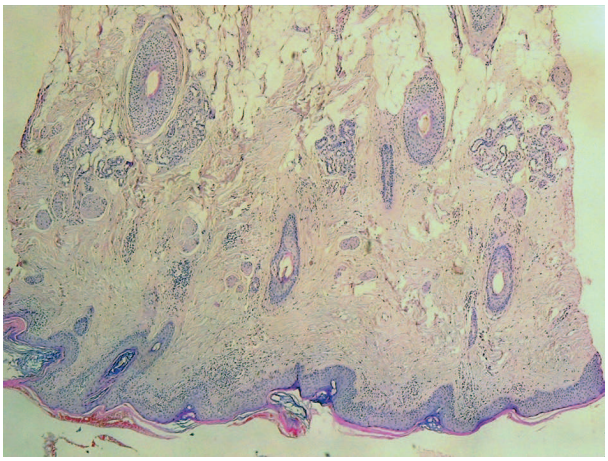

Fig. 2. Hyperkeratosis, hypergranulosis, irregular acanthosis, vacuolar degeneration of the basal keratinocytic layer at the site of follicular ostia, thick fibrosis with many fibrotic tracts, replacing the previously existing follicular units in the deep dermis.

Topical therapy with methylprednisolone cream and oral hydroxychloroquine were introduced. At the 3-month follow-up, the erythema was resorbed, no follicular papules were seen, and pinpoint atrophic areas with occasionally spared one-to-two follicles, were presented. The patient was recommended to continue the oral antimalarial regimen for one year.

DISCUSSION

Scarring alopecia represents a great number of dermatological disorders with very obscure pathophysiology and unpredictable clinical evolution. There are various attempts of classification based on history, clinical or pathological features.

The congenital group of cicatricial alopecias includes Darier's disease, sex-linked ichthyosis, dystrophic epidermolysis bullosa, incontinentia pigmenti, polyostotic fibrous dysplasia, HHHH (hypotrychia, hyperkeratosis, hydrocystomas, hypodontia) syndrome, obstetric traumas, organoid and epidermal nevi, simplex hereditary hypotrichosis, follicular keratosis, aplasia cutis congenital, porphyries, etc. (3).

Acquired scarring alopecias result from physical and chemical traumas, cutaneous infections, tumor infiltrations, and some specific dermatological disorders such as discoid lupus erythematosus, lichen planis, sarcoidosis, deep morphea, nodular cystic acne, etc. (4).

Various subjective symptoms have been reported in the clinical setting of cicatricial alopecias. For example, 60% of the patients with lichen planopilaris claimed to have itch, burning, and pain (3,4) and 30% of them have co-existing androgenic alopecia. Mucinous alopecia occurs concomitantly with dyshidrosis and dysesthesia. Permanent chemotherapy-induced alopecia is commonly triggered by busulphan and is often associated with tingling sensation. It usually presents with loss of hair follicles due to high-dose regimens or subsequent bone marrow transplantation. Dissecting folliculitis of the scalp and folliculitis decalvans are always associated with burning and tenderness. Pressure alopecia usually causes itch and exacerbates seborrheic dermatitis. Pseudopelade, traction alopecia, lipedematous alopecia, and erosive pustular dermatitis of the scalp are almost never associated with subjective symptoms (5).

Physical examination is usually insufficient to make the diagnosis alone. However, there are some changes that have been related specifically to certain clinical entities. Typically, lichen planopilaris always shows perifollicular erythema and hyperkeratosis (6). Central centrifugal cicatricial alopecia is classically described as starting from the crown and spreading centrifugally. Brocq pseudopelade is non-inflammatory progressive alopecia, characterized by large atrophic patches with irregular borders and centrally dispersed hairs, occasionally with true retractile follicular depressions. Alopecia mucinosa affects the head and neck region with the formation of prominent follicles and indurated plaques with fine scales (7). Keratosis pilaris atrophicans affects the eyebrows and spares the scalp. Atrophoderma vermiculata affects the cheeks, causing primarily follicular hyperkeratosis with resulting atrophic depressions. Discoid lupus erythematosus starts with erythematous papules and plaques that cause follicular plugging. Poikilodermic skin lesions are usually seen in 50% of patients with scalp involvement. Scarring alopecia changes can precede the cutaneous lesions with many years. Folliculitis decalvans starts with multiple pinpoint erythematous pustules that can confluent into a large abscess. Classically, round- to irregular-shaped atrophic flesh-colored or ivory-white areas of scarring alopecia are seen in advanced cases. Tufted follicles are often detected (8). Dissecting folliculitis of the scalp shows painful, bulbous, firm or fluctuant nodules. Lymphadenopathy may be present. Long-standing and progressing disease results in cribriform irregular hypo- and hypertrophic scars (9). The very rare cases of erosive pustular dermatoses of the scalp have large, well-circumscribed, crusted plaques with exudative erosions and pustules. Episodic pustular flares with progressive slow enlargement are common. Bacterial colonization is often superimposed (10).

Clinical presentation should be additionally confirmed with scalp biopsy. Histology examination is considered mandatory in all cases with cicatricial alopecia (11). Two biopsies – one vertical and one horizontal – are usually performed. The widely accepted North America Hair Society classification recognizes four categories of scarring alopecias: lymphocytic (discoid lupus erythematosus, lichen planopilaris, Brocq pseudopelade, central centrifugal cicatricial alopecia); neutrophilic group (folliculitis decalvans and dissecting folliculitis of the scalp); mixed group (folliculitis keloidalis), and nonspecific cicatricial alopecias with inconclusive clinical and histopathological findings (scarring cases of traction alopecia, trichotillomania, etc.) (12).

The histological verification of lymphocytic and neutrophilic scarring alopecias validates the proper therapeutic approach. Neutrophilic alopecias are usually treated with antibiotic and anti-inflammatory drugs (13). Severe cases of dissecting folliculitis may be introduced on isotretinoin therapy. Lymphocytic alopecias provoke greater scientific interest. The unclear pathophysiology and the obscure inflammatory targets do not allow specific pathogenetic therapy. A therapeutic ladder has been suggested to oppose alopecia's recalcitrant nature (14). Mid- to high-potency topical corticosteroids are generally well-accepted to cope the initial inflammation. Of the patients, 70% to 83% had good improvement after 90 days of therapy. However, the relapse rate upon cessation was more than 80%. Intralesional corticosteroids can further decrease the inflammation and reduce the number of recurrences. Since there are no convincing data for the topical use of calcineurin inhibitors, topical corticosteroids are considered first-line therapy with very safe side effect profile. Recommended second-line therapy includes systemic corticosteroids, immune-suppressors and anti-metabolites. Corticosteroids should be used as bridge therapy or for a short-term therapy of the acute inflammatory phase. Long-term use is limited by the association of adverse effects. Cyclosporine and hydroxychloroquine are the best corticosteroid-sparing agents. Cyclosporine is reported to have 77% treatment success rate. Optimal dose is 4 mg/kg/day for a maximum of 6 months. No consistent data exist for the therapeutic benefit of hydroxychloroquine (6). Some authors reported significant improvement after 1 year of 400 mg/day, whereas other did not see good results. Antimetabolite mycophenolate mofetil has been shown to be rather effective. Of the patients who previously failed other therapies, 83% had a significant improvement with 0.5 mg twice daily for a period of 4 weeks, which was increased to 1 mg daily for additional 20 weeks. Very low risk of end-organ damage and cancer has been reported with mycophenolate mofetil (15). It is considered safe and gen-

erally well-tolerated. Oral retinoids, tetracyclines, methotrexate and 5-alpha reductase inhibitors are considered third-line therapy (6). Adjunctive therapy includes pioglitazone hydrochloride (15 mg/day), oral peroxisome proliferator-activated receptor gamma agonist, as nowadays the inflammation in lymphocytic scarring alopecia is suggested to be a result of abnormal peroxisome proliferator-activated receptor (PPAR) gamma activity (16).

CONCLUSION

The variable clinical and pathological presentation turns the accurate diagnosis of cicatricial alopecias into a challenging task. An attentive clinicopathological correlation is always advisable. The recalcitrant nature, unclear pathophysiology and lack of consistent clinical trials greatly affect the utilization of definitive therapeutic approach and further challenge the dermatologists in their every-day routine. Greater scientific interest with more research efforts are needed to further highlight this extremely interesting dermatological problem.

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