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Case Report

Lichen Planopilaris, Dijagnosis and Therapy Challenge - Case Report

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Abstract: Background: Lichen planopilaris is relatively rare inflammatory disorder that results in cicatricial alopecia. It is a rare, cutaneous form of lichen planus which affects hair follicles, most commonly on the scalp area. It is caused by an autoimmune disorder that leads to follicular destruction and permanent hair loss. Case history: We are presenting the case of a patient who is 21 years old and who suffers from cicatricial alopecia and follicular hyperkeratosis of the parietal region of the scalp. Initially, she was diagnosed with seborrheic dermatitis and has been treated with local corticosteroid therapy for several months by her doctor. She was admitted to the Department of Dermatology with lessions located on the scalp. A clinical examination revealed exudative erythema with prominent circumferential hair follicle openings and the presence of sticky, yellowish crusts and scales whose removal caused bleeding. There were a several locations with significant cicatricial alopecia which look like porcelain. Dermoscopy showed perifollicular squamas. Routine laboratory analyses and immunological analysis were in normal range. Finally, after taking a biopsy and histopathological findings, we have come to a differential diagnosis which may include fibrous and suppurativ follicular disorders, as well as lichen planopilaris. The patient was treated with keratolytic lotions, combination of corticosteroid preparation and salicylic acid applied topicaly, and tacrolimus 1% cream which was used twice per day for several months. This terapy was successful, crusts did not return and progression of the disease was stopped.

Keywords: Lichen Planopilaris, Alopecia Cicatricialis, Inflamation

1. Introduction

Lichen planopilaris (LPP) is a form of severe alopecia characterized by perifollicular erythema, follicular keratosis and degeneration of follicular openings. LPP is a rare variant of cutaneous lichen planus (LP), which affects hair follicles, most often on the scalp area, but it can also affect other regions of the body covered with hair, like axillary and pubic regions [1]. A typical finding is a cicatricial alopecia on the scalp area with loss of follicular ostia. The most common symptoms associated with the above disease are itching, tingling and pain [2]. Signs of inflammation on the scalp area may precede the hair loss for several months [3]. There are two clinical entities of this disease, the first one is Graham-Little-Piccardi-Lasseur syndrome which in addition to the scalp cicatricial alopecia, is clinically characterized by nonscarring pubic and axillary alopecia, as well as grouped follicular papules on the body and extremities, while the other entity is a frontal fibrosing alopecia (FFA) [4]. Diagnosis of LPP is confirmed by clinical picture, dermoscopic and histopathological findings. Recent studies have shown that women with FFA and LPP have less chance of suffering from diabetes, hypertension, heart disease and hypothyroidism, but they have greater probability (4.37%) to suffer from systemic lupus erythematosus (SLE) [5].

2. Case Report

We present a female patient who is 21 years old with changes on the scalp for over one year. Disease initially manifested itself as a flaking of the skin accompanied by itching in the parietal region of scalp. As a result of scratching, smaller wounds appeared in the affected area, accompanied by secretion of yellowish liquid and the subsequent appearance of sticky deposits that could not be easily separated from the skin. Changes were considered to be seborrheic dermatitis, and the local corticosteroid therapy was applied for a long time. After hair loss, changes were considered to be Pseudopelade Brocq by her doctor. During 5 months of therapy, potent topical corticosteroids were used continuously, without improvement.

2.1. Dermatological Local Finding

Changes were in the form of pronounced exudative erythema with prominent circumferential hair follicle openings, the presence of sticky, yellowish crusts and scales whose removal causes bleeding (Figure 1) located in the larger part of the parietal region of the scalp, while the remaining parts of the scalp, the skin, mucosa and nails had no pathological changes. There are a several fields with significant cicatricial alopecia which look like porcelain. The hair in the parietal region as a whole is sparse compared to the rest of the scalp. Polytrichia is present in some parts. Pull test was positive.

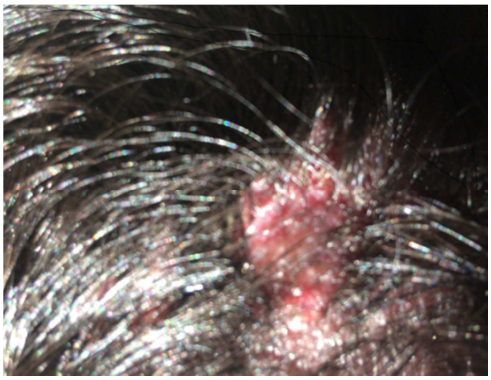

Figure 1. *Inflammation of the parietal scalp region with adhesive crusts and scales.*

2.2. Clinical Laboratory Findings

Routine laboratory analyses were *within reference range*. ANA and anti - ds DNA antibodies were negative, the IgE, IgA and IgG antibodies and immune complexes were *within reference range*. Thyroid gland hormones and anti TPO antibodies were *within normal limits*. Polycystic ovaries and other abnormalities have not been determined by gynecological examination. Hormonal status (serum levels of estrogen, progesterone and testosterone) were *within reference range*.

2.3. Dermoscopic Finding

Perifollicular squamas are present.

2.4. Histopathological Findings

The epidermis is a slightly hyperkeratotic with rare micro abscess in the coral layer, with follicular hyperkeratosis, and with partially formed keratin plugs. Dermis is a little enlarged, fibrotically altered, abundantly infiltrated by a diffuse mixed inflammatory cellular infiltrate made up of neutrophils, rare eosinophils, with prevalence of monoclonal cells of the lymphocyte type, as well as macrophages and plasma cells. The inflammatory infiltrate with mononuclear structure is present perifollicularly. Some hair follicles show the perifollicular lamellar fibrosis, while the vascular channels of the dermis have the irregular lumen, some with dilated - “teleangiectatic-like” appearance, with perivascular cell infiltrate. (Figure 2).

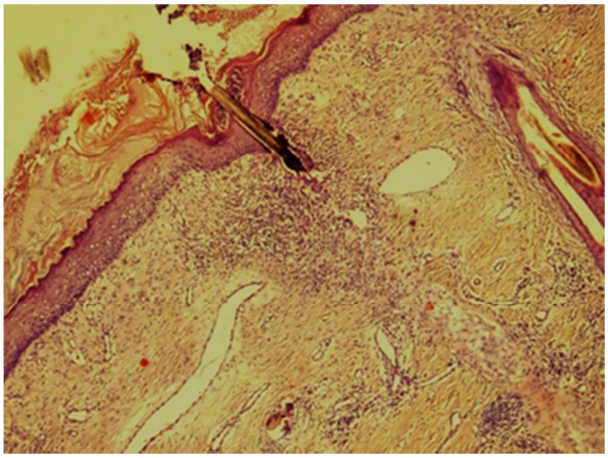

Figure 2. *Necrotizing (peri) folliculitis, follicular plug, dermal fibrosis, perifollicular lamellar fibrosis, superficial inflammatory infiltrate and blood vessels of telangiectatic-like appearance (H&E, x50).*

In affected parts of the dermis, hair follicles are significantly reduced, while remaining ones have almost entirely torn follicular epithelium with the remaining central hairs surrounded by multinucleated foreign-body giant cells, lymphocytes and plasma cells. (Figure 3).

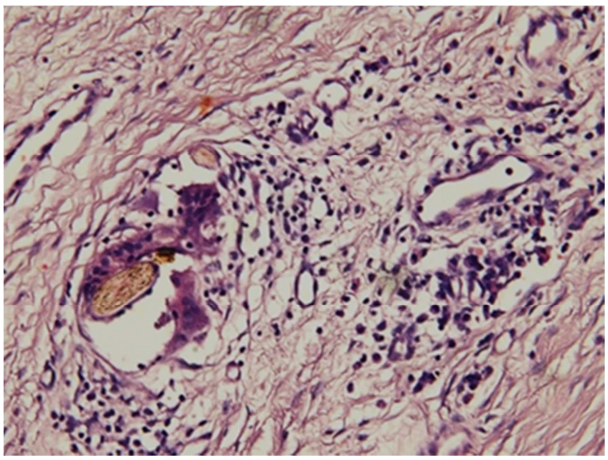

Figure 3. *Dermal fibrosis , multinucleated giant cells and lymphoplasmacytic infiltrate infiltrate around the hair residues (H&E, x200).*

The hair follicles are predominantly in the

dermohipodermal joint. In addition, there is a significant reduction of the sebaceous glands and the signs of the polytrichia.

The above described micromorphological finding may indicate an alopecia cicatricialis, with hair follicle damage by the type of necrotizing folliculitis and the foreign body granuloma type reaction. Differential diagnosis may include fibrous and suppurativ follicular disorders, as well as lichen planopilaris.

2.5. Therapeutic Approach

Therapy was focused on variety of issues. The consequences of steroid dermatitis with thinned epidermis and bleeding on touch should have been cured first. It was necessary to remove the thick sticky crusts and scales, which were especially pronounced on the perimeter of cicatricial fields. Mild gels for removing cradle cap in babies and keratolytic lotions have helped in removing plaque. The use of topical corticosteroids was discontinued for 3 months. Antibiotic ointment and keratolytic lotions were applied instead, and medicated shampoo for dandruff was used to wash hair. This resulted in initial severe inflammation to be cured (Figure 4). The biggest therapeutic problem was the remaining sticky crusts and scales that were formed as soon as the use of keratolytics was stopped.

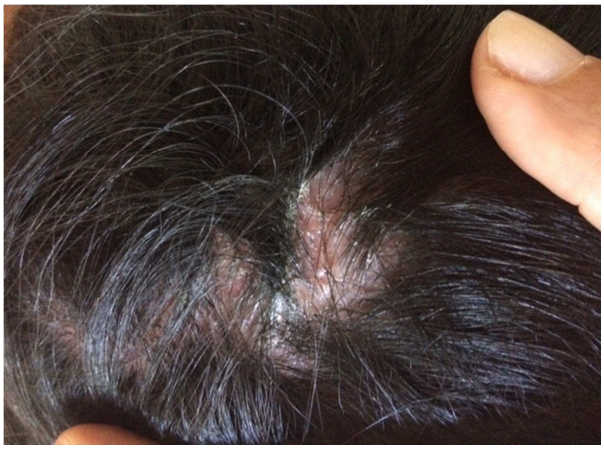

Figure 4. *3 months from the beginning of the treatment, multifocal porcelain atrophy of the skin with peripheral follicular hyperkeratosis without signs of inflammation.*

Taking into account that the patient is young, and bearing in mind potential unwanted effects of systemic administration of medications, we decided to apply topical therapy which fortunately resulted in regression of changes. Three months later a combination of corticosteroid preparation and salicylic acid was applied topically for 14 days, which gave a satisfactory result with removal of a large part of scales and inflammation. In order to remove the remaining deposits the maintenance therapy included tacrolimus 1% cream which was used twice per day for several months. The maintenance therapy was successful as crusts did not return and progression of the disease was stopped (Figure 5).

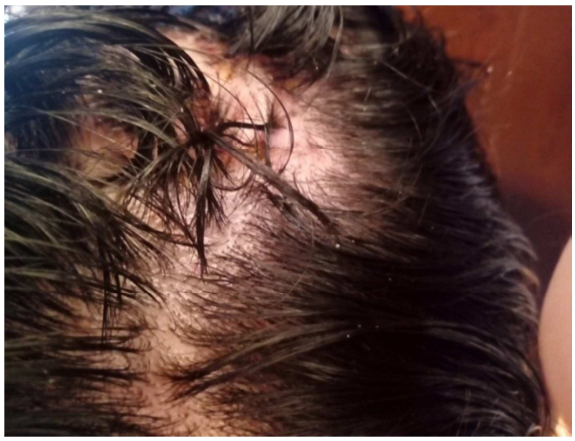

Figure 5. *Six months of topical application of tacrolimus 1% cream resulted in the removal of crusts without recurrence up to now.*

3. Discussion

Cicatricial alopecia may appear as a result of numerous diseases such as Lupus erythematosus, Alopecia mucinosa, Lichen planopilaris, alopecia as a side effect of chemotherapy, Pseudopalade Broq, Erosive pustular dermatosis, Traction alopecia, etc.

LPP is a rare disorder characterized by chronic lymphocytic infiltration that leads to the selective destruction of hair follicles which eventually results in cicatricial alopecia [4]. Only 30% of patients have lesions on the skin and mucosa [6], and the remaining 70% have only a change in the area of hair-covered skin. The mechanism of the disease occurrence has not been studied enough. Numerous authors regard it as being a specific autoimmune disorder where T lymphocytes affect follicle antigens with consequent damage to the follicle stem cell [7]. Possible mediators include b-FGF and TGF-β that may be responsible for the activation of fibroblasts. Recent studies emphasize the possible role of PPAR-Y in the destruction of pilosebaceous unit typical of LPP [8].

To determine the diagnosis, one should exclude discoid lupus, folliculitis decalvans as well as other forms of cicatricial alopecia. The diagnosis ought to be based on the clinical picture, the possible dermoscopic examination [9], and histopathological findings. Dermoscopy can detect perifollicular scales at LPP. In FFA dermoscopy shows minor perifollicular scaling, lonely hair/predominance of follicular openings with only one hair at the hair-bearing margin [9].

There are certain histopathological criteria which are based on more comparative inspections of the preparations used by people with LPP [10]. In developed form infundibular hyperplasia and follicular plugging are accompanied by a dense lichenoid infiltrate of lymphocytes, pigment incontinence and necrotic keratocytes/apoptosis (“cytoid or Civatte bodies”) [10]. Fully developed form shows loss of sebaceous glands and stem cells, initial fibrosis. In late form one can see thinning of follicular epithelium, peri-infundibular, superficial scar embracing the infundibulum accompanied by perifollicular mucin and wedge-shaped loss of elastic tissue and scant lymphocytic inflammation that “backs away” from

the follicle, [11], infundibular tufts/follicular fusion (compound follicles) due to mild scarring around hair follicles and their infundibula in particular. That is in contrast to tufted folliculitis or Folliculitis decalvans where a much more destructive and granulomatous scaring reaction is seen. Sometimes a moderate and superficial stromal foreign-body reaction to released hair shaft material may exist [10].

DIF test shows nonspecific IgM deposits and, less often IgA and IgG and C3 deposits in the cell bodies around the follicles [12].

Treatment of LPP is a great therapeutic challenge. The main goal is to alleviate the symptoms and stop progression of the disease in order to preserve hair. Treatment depends on the severity of symptoms. Hydroxychloroquine is effective in reducing the symptoms of the disease if it is applied for 6-12 months continuously [13].

Methotrexate, according to some studies, was more efficient than hydroxychloroquin in a cumulative weekly dose of 15 mg after 6 months of use [14]. A possible mechanism is based on the inhibition of purine and pyrimidine synthesis, suppression of transmethylation reactions with accumulation of polyamines, reduction of antigen-dependent T-cell proliferation [15, 16]

Taking into consideration that LPP shows significantly decreased levels of PPAR-γ, a transcriptional factor that regulates lipid metabolism and peroxisome biogenesis [17] studies were conducted where it was applied systemic with success, PPAR-γ agonist, pioglitazone hydrochloride at a dose of 15 mg / per day [18] for 8 months.

Tetracyclines have variable effects on the course of the disease, administered as monotherapy at a dose of 200 mg daily for 3-6 months [19].

Studies have been made showing that mycophenolate mofetil, which is used in the treatment of autoimmune and inflammatory skin diseases, was applied, because it has a better safety profile than azathioprine; however, it requires additional testing before standard use of this drug can be approved [20].

Retinoids can be administered in the treatment of the LPP, Acitretin in a dosage of 25-50 mg daily. They are used just in a small number of therapeutically resistant cases due to adverse effects (skin and mucosal dryness, hypercholesterolemia, headaches, increased serum transaminase...) [21].

A for the topical therapy, corticosteroids are the first therapeutic choice [7]. Potent corticosteroids such as clobetasol propionate can be applied. In frontal fibrosing alopecia, considered as an LPP entity, an intralesional application of triamcinolone acetonide in the loss of hair on the eyebrows was attempted with a great success. The drug was administered at a concentration of 10 mg / ml, 0.125 ml per eyebrow [22].

Tacrolimus gives good results in patients resistant to corticosteroid therapy, when it has been administered for 9 months or more [23].

Low-frequency laser therapy which has been applied for 6 months, wavelength of 630 nm, energy 4 J/cm², applied for 3 minutes per day in the examined patients caused a reduction in the symptoms of the disease and an increase in the thickness of the terminal hair [24].

4. Conclusion

Lichen planopilaris is a relatively rare disease, whose progression leads to a cicatricial alopecia. The disease is often resistant to therapy. Early diagnosis is extremely important, because timely treatment prevents the occurrence of Cicatricial alopecia. Therapy in early diagnosed disorders should be initiated with topical corticosteroids and tacrolimus, and if necessary, it should be combined with systemic therapy in some cases. In the case of our patient, although histopathologic findings showed that she was in late stage of the disease, with developed cataractic alopecia, topical therapy gave a satisfactory result without recurrence. The patient will be regularly monitored to prevent relapse of the disease. Treatment of this dermatitis requires time and patience, with a goal to reduce the symptoms and prevent progression of the disease by using the therapy with the least side-effects.

Abbreviations

LPP- Lichen planopilaris
FFA- Frontal fibrosing alopecia

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