Idiopathic Follicular Mucinosis in a 32-year old Filipino Man Successfully Treated with Narrow-band UVB: A Case Report*

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ABSTRACT

Introduction: Follicular mucinosis is a rare skin condition, which can present as an idiopathic benign condition or as a secondary symptom of a malignant disorder, most commonly mycosis fungoides. Since its first description in 1957, only 90 cases have been reported worldwide. This condition is characterized by erythematous infiltrated plaques with scaling, sometimes with follicular prominence on the head and neck. Definitive diagnosis is made histologically.

Case: A 32-year old male presented with a 3-year history of recurrent scaling with eruption of pruritic erythematous papules and plaques over the face, neck and chest. Different local treatments (topical corticosteroids, antifungal cream and moisturizers) were ineffective. Overall, he was healthy. Histopathological examination of the lesion showed typical histological picture of follicular mucinosis. Alcian blue stain was positive. The patient was successfully treated with narrow-band UVB (NB-UVB) therapy with no recurrence of lesions after one- month follow-up.

Conclusion: Idiopathic follicular mucinosis is a benign condition with localized involvement and excellent prognosis. However, long-term follow-up and monitoring is recommended because of its association with malignancy.

Follicular mucinosis is a rare dermatosis, characterized histologically by the presence of mucin deposits within the hair follicle and sebaceous glands.[1] There are three clinical types determined. The first and the most common type occurs as an idiopathic benign disorder, typically seen in young patients. The second type affects people 40-70 years of age and has a more chronic course.

The third type usually occurs in the elderly and presents with widespread lesions. It can arise as a secondary symptom of a malignant disorder, most commonly mycosis fungoides.[2]

CASE REPORT

A 32-year-old Filipino male presented with a 3-year history of recurrent scaling over the glabella, nasolabial folds, preauricular areas, pinnae of the ears and maxillary area, which would resolve spontaneously or whenever the patient applies a facial moisturizer. One week prior to consult, there was persistence of the scaling with the eruption of pruritic erythematous and some skin-colored papules and plaques over the face, neck and chest. The patient applied virgin coconut oil three times a day which provided a slight decrease in scaling. Some papules eventually coalesced to form plaques while some formed erythematous nodules over the face. The patient discontinued application of virgin coconut oil and self-medicated with cetirizine 10mg once daily which gave temporary relief of the pruritus.

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Review of systems was unremarkable.

Past medical, family and personal histories were noncontributory.

Upon consult, the patient had stable vital signs. Examination of the skin revealed multiple erythematous follicular scaly papules, nodules and plaques over the forehead, cheeks, nasolabial folds, chin, ears, posterior auricular area, neck, and chest (Figure 1 A-D), with no loss of hair and no palpable lymph nodes. The rest of the physical examination was normal.

The initial impression at that time was seborrheic dermatitis. The patient was prescribed hydrocortisone + miconazole (Daktakort) cream, applied twice daily over the affected areas and cetirizine 10 mg daily, which resulted to an increase in erythema, scaling, number and size of papules and plaques over the affected areas. Follow-up consult was done in which the patient was reassessed with allergic contact dermatitis secondary to hydrocortisone + miconazole (Daktakort) cream. The patient was then given prednisone 10 mg daily for 5 days. Cetirizine was continued while hydrocortisone + miconazole (Daktakort) cream was shifted to mometasone furoate cream. The patient noted a decrease in the number and size of the lesions with resolution of some lesions. However, recurrence of lesions was noted 5 days after discontinuation of medications. Intralesional triamcinolone injection 10mg/ml was injected once over the nodules, which provided slight improvement. However, the scaly papules and plaques over the face and chest persisted. A 3-mm skin punch biopsy was obtained from an indurated plaque over the left temporal area. Histopathology showed irregular acanthosis, moderate to dense superficial and deep perivascular and periadnexal mixed cell infiltrates consisting of lymphocytes, histiocytes and numerous eosinophils admixed with extravasated red blood cells (Figure 2A). Mucin is deposited within the follicles and throughout the dermis, confirmed by alcian blue stain (Figure 2B).

With these findings, an impression of follicular mucinosis was given.

The patient was treated with ten sessions of narrow-band ultraviolet B (NB-UVB) therapy twice a week, starting at 200 mj, with the dose increased by 20% with each succeeding session. The therapeutic dose was increased by only 10% on the 9th session and was maintained on the 10th session. Desonide cream was also applied over affected areas twice daily for a week. Total resolution of lesions was noted after 10 sessions of UV8 and no recurrence was noted after one month of follow-up.

DISCUSSION

In 1957, Pinkus[3] first coined the term alopecia mucinosa to 6 cases of follicular papules or indurated plaques with alopecia. This was characterized histologically by mucinous change in the pilosebaceous follicle.[4] Since alopecia is not conspicuous outside the scalp or eyebrows, this disorder is also known as follicular mucinosis. The accumulation of mucin creates an inflammatory condition and subsequent degenerative process. When this condition affects hair-bearing areas, a non-scarring alopecia, also known as alopecia mucinosa can result.[2]

The exact pathogenesis is unknown, although the role of cell-mediated immunity and circulating immune complexes have been considered.[2] It is considered a reaction of the follicular epithelium to diverse stimuli[5] and can occur in several inflammatory, infectious and neoplastic skin conditions.[6] Follicular mucinosis affects all races and ages with equal sex predilection.

Follicular mucinosis typically presents as erythematous infiltrated plaques with scaling and sometimes follicular prominence on the head and neck.[2] These plaques are covered by open porous pilosebaceous, small follicular papules and alopecic areas. However, atypical presentation includes nodular forms that may mimic folliculitis, chronic eczema, seborrheic dermatitis[4], acneiform lesions?, and urticaria. In this case, the patient was initially managed as a case of seborrheic dermatitis, however, no clinical response was observed after the patient was given a combination of corticosteroid and antifungal medication. The loss of hair in plaques with follicular papules but minimal inflammatory changes should suggest the diagnosis but a definitive diagnosis of follicular mucinosis is made histologically. Therefore, multiple biopsies and serial sectioning of tissue are necessary to confirm the diagnosis.[4]

Histologically, follicular mucinosis is characterized by mucinous degeneration in the external sheath of the hair follicle and of the sebaceous gland. This could be followed by formation of cystic spaces, which are partially covered by mucin. Inflammatory infiltrates in the primary form of follicular mucinosis are perivascular and perifollicular, comprised of lymphocytes and eosinophils.[5] Differentiation between primary and lymphoma-associated follicular mucinosis based on histological basis can be very difficult. Nevertheless, generally, the presence of a high number of eosinophils in the inflammatory infiltrate and marked mucinous alterations in the follicular epithelium favor a benign form, while the presence of epidermotropism and dense perifollicular infiltrate of atypical cells point to the lymphoma-associated form of follicular mucinosis.[4]

The patient's biopsy showed moderate to dense mixed cell infiltrates in the superficial and deep perivascular and periadnexal areas. The infiltrates mainly consisted of lymphocytes, histiocytes and numerous eosinophils, with no findings of epidermotropism of the hair follicular epithelium or dense infiltrate with atypical cells. Mucin deposition was also noted within the follicles and throughout the dermis. With the above findings, classification under the benign type of follicular mucinosis can be supported. Other factors that could point to the benign nature of the condition include the young age of onset, confinement of lesions to the face and neck and short recovery period.

There is no standard therapeutic approach for follicular mucinosis. Therapeutic options include topical and systemic psoralen and ultraviolet A (PUVA), NB-UVB, topical and intralesional steroids,[8] minocycline and in more severe cases, topical nitrogen mustard, dapsone, indomethacin,[9] retinoids, electron beam therapy, radiation, chemotherapy and interferon alpha.[9] The patient noted complete resolution of lesions after undergoing ten sessions of NB-UVB therapy (Figure 3 A-C) with no report of reappearance of new lesions.

In a report by Taniguchi[10], a 56-year old Japanese female, who presented with indurated erythematous plaques on the trunk and lower extremities was diagnosed with folliculotropic mycosis fungoides, and was successfully treated with NB-UVB. The minimal erythema dose (MED) was determined to be at 1.2 J/cm^2. The surface of the body affected was irradiated with 70% MED (0.8J/cm2) on the first session, increased by 10% on the second session. Papules and nodules on the trunk and lower extremities decreased after 8 exposures, after which she continued irradiation at the same dose once a week. After 24 exposures, all of the lesions had completely disappeared. On follow-up after 3 years, no new skin lesions were observed. Ultraviolet B (UVB) has been shown to decrease the antigen presenting capacity of Langerhans cells, increase interleukin-2 and interleukin-6 production by human keratinocytes, as well as increase the level of TNF alpha. UVB is thought to be therapeutic by suppressing the function of the neoplastic population of clonal T cells in the skin by serving as an upregulator of the immune system.[10]

CONCLUSION

Follicular mucinosis is a rare dermatosis that typically presents as erythematous infiltrated plaques with scaling and sometimes, follicular prominence on the head and neck. Based on the age of onset of the condition, confinement of the lesions to localized areas, fast recovery period after treatment and with the histopathologic findings, the patient most likely has the idiopathic type of follicular mucinosis. Although his clinical presentation is atypical in that it mimicked seborrheic dermatitis, the patient was managed accordingly through histopathologic findings. However, even if with excellent prognosis, the patient must be monitored and followed-up long-term because of the condition's association with malignancy.

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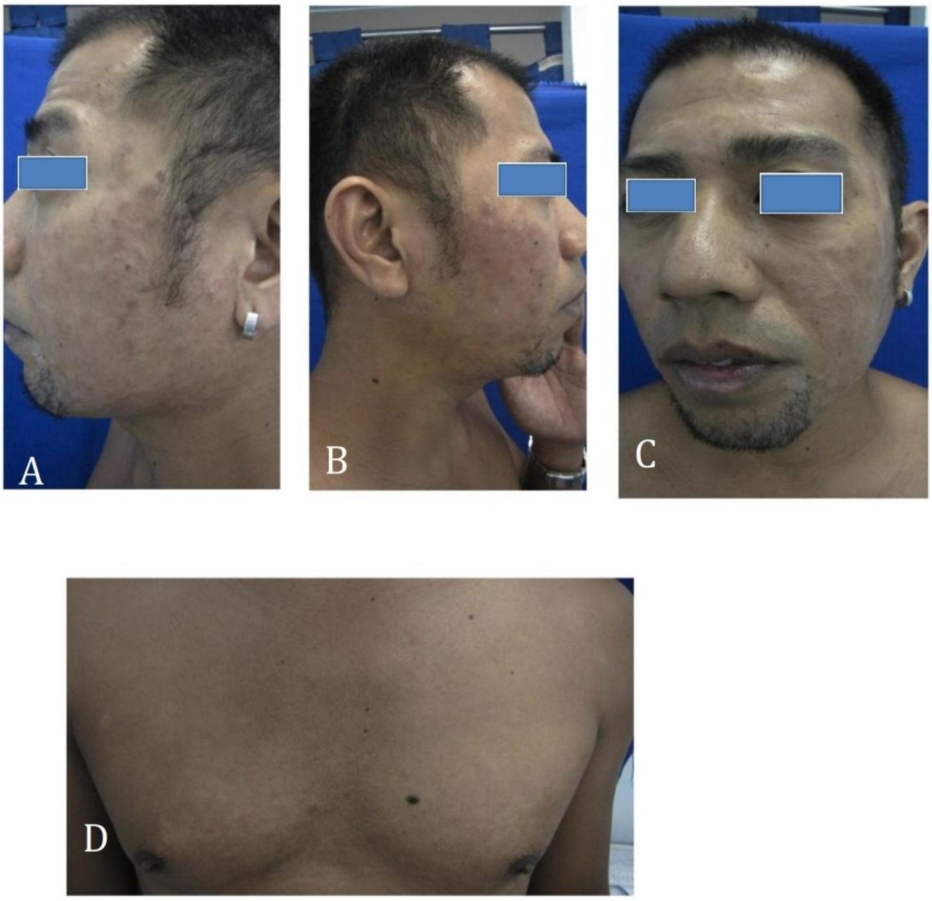

Figure 1 A-D. Multiple erythematous scaly follicular papules, plaques and nodules on the forehead, cheeks and pre-auricular area (without loss of hair), chest before treatment

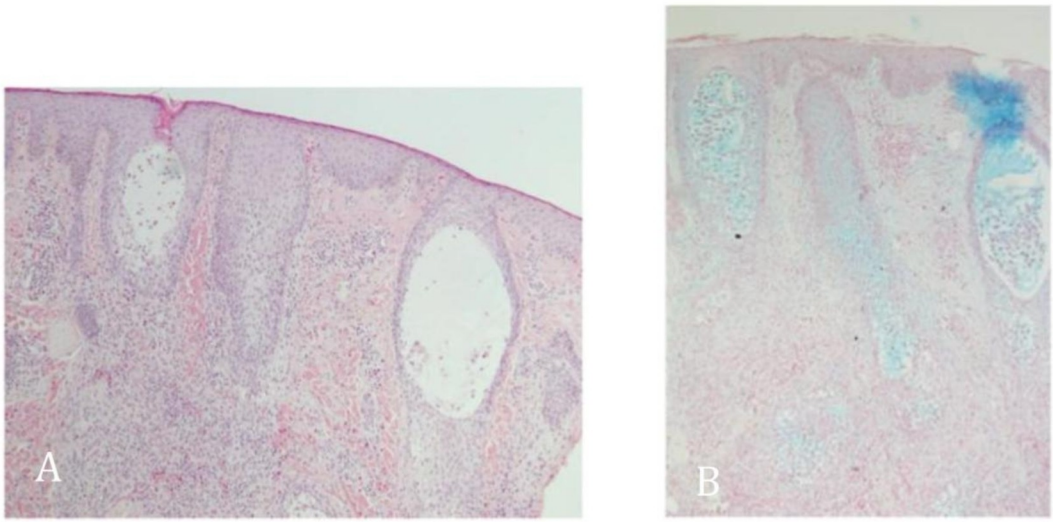

Figure 2. (A) Follicles surrounded by mixed cell infiltrates; (B) Mucin is deposited within the follicles and throughout the dermis confirmed by alcian blue stain (10x magnification)

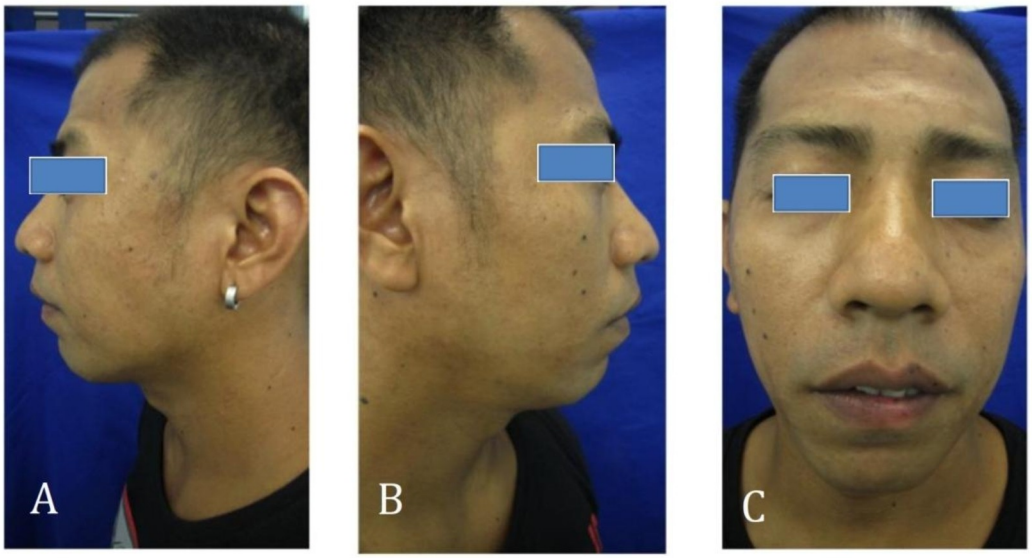

Figure 3 A-C. Resolution of skin lesions after ten sessions of NB-UVB