A Case of Pemphigus Foliaceus in a 40-Year-Old Female Successfully Treated with Doxycycline and Prednisone Combination Therapy*

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Here, we present a 40-year-old female with multiple pruritic occasionally painful vesicles, papules, and plaques in a circinate pattern on seborrheic areas, progressing to erosions and scales. Clinical findings led to the diagnosis of pemphigus foliaceus (PF). Initial treatment with prednisone and clobetasol ointment, however, did not fully suppress blister formation and healing of erosions. Skin punch biopsy revealed a subcorneal split and intracorneal neutrophilic infiltrates, while enzyme-linked immunoassay (ELISA) revealed elevated anti-desmoglein 1 (Dsg1), consistent with PF. Doxycycline was then added to the previous regimen, resulting in remission. We discuss the role of doxycycline as a cost-effective adjunctive treatment in patients with refractory PF.

INTRODUCTION

Pemphigus foliaceus (PF) is an autoimmune blistering disease (AIBD) mediated by autoantibodies targeting Dsg1 responsible for cell-to-cell adhesion of keratinocytes in the upper epidermis [1,2].

PF is characterized by flaccid vesicles on seborrheic areas. However, examination more often reveals scaly or crusty erythematous patches, plaques, and erosions with cornflake scales. The lack of blisters and mucosal involvement lead to diagnostic delay as this may be misdiagnosed as eczema, seborrheic dermatitis, or psoriasis [2]. Histologic examination may not be enough and DIF, ELISA, and/or indirect immunofluorescence (IIF) may be used to aid diagnosis.

No optimal therapeutic approach has been established for pemphigus. Glucocorticoids remain as cornerstone in treatment, and may be used in combination with other immunomodulatory therapies such as rituximab, azathioprine, or mycophenolate mofetil. However, present literature supports that combined therapies have not been shown to be more effective than glucocorticoids alone. Unfortunately, as therapy is prolonged, numerous drug-induced side effects can occur, many of which are serious. Moreover, most steroid-sparing agents available are expensive and not widely available especially in third world countries like the Philippines [1,3,5].

Contrary to present evidence-based data, this case initially did not have complete remission to monotherapy with prednisone but responded well with the addition of doxycycline.

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CASE REPORT

A 40-year-old Filipino female presented with a 2-year history of multiple flaccid vesicles, erythematous papules, plaques, patches, erosions as well as whitish scales on the trunk, face and extremities. This was perceived by the patient to be exacerbated by stress and not triggered by sun exposure and medications. Past medical, family, and social history were all unremarkable. She consulted a dermatologist and was clinically diagnosed as pemphigus. She was initially given prednisone 60 mg per day tapered weekly to 5mg per day, clobetasol 0.05% ointment applied twice a day on the body, and hydrocortisone 1% cream applied twice a day on the face, that all afforded temporary minimal decrease in thickness but did not result in complete resolution despite compliance to treatment. A re-appearance of new lesions on the trunk after 3 months, prompted consultation at our institution. Routine histology showed subcorneal splitting and intracorneal neutrophilic infiltrates, DIF revealed intercellular IgG deposition, and ELISA showed elevated anti-desmoglein 1 levels, all of which were consistent with the initial clinical diagnosis of PF.

The lack of response to prednisone, as well as the limited availability of other steroid-sparing medications posed a challenge in the management of the case. Doxycycline 100 mg twice a day was then added to the treatment plan and resulted in gradual decrease in the thickness, number and erythema of the lesions. With clinical improvement, prednisone was tapered gradually for 2 months until zero with no flaring of lesions. Our patient did not experience any side effects with intake of doxycycline.

CASE DISCUSSION

Pemphigus foliaceus (PF) is a rare autoimmune disorder that typically occurs in patients aged 40 to 60 years and affects males and females equally. Clinically, it is characterized by an often thick keratotic scale on an erythematous base in a "seborrheic" distribution. PF patients are rarely ill. Further diagnostic tests reveal subcorneal acantholysis mediated by IgG anti-desmoglein-1 (DSG1) antibodies [1,2,5,8]

A number of studies confirm the efficacy of doxycycline in conjunction with topical steroids as a first treatment strategy in bullous pemphigoid. The Bullous Pemphigoid Steroids and Tetracyclines (BLISTER) study revealed favorable results in patients given 0.5mg/kg body weight prednisolone or 200mg doxycycline daily and 30g steroid cream applied as needed for 3 weeks [3]. The British Association of Dermatologists 2012 guideline recommends use of systemic and topical steroids along with doxycycline may be used in BP. However, this treatment regimen and its use with pemphigus foliaceus is less documented [6,7].

In this case, we present doxycycline as a treatment for PF that can be used in conjunction with prednisone and clobetasol ointment. Doxycycline is a tetracycline antibiotic which acts via the inhibition of bacterial ribosomes. It also has useful anti-inflammatory properties which make it useful in blistering diseases. Unlike other non-steroidal adjuncts, doxycycline is widely available and affordable, which may prove to be a promising management in patients with PF.

CONCLUSION

This study provides evidence that the use of doxycycline, in conjunction with oral and topical steroids may be effective in the management of PF and adds to the list of medications that may be used as a steroid-sparing agent in PF.

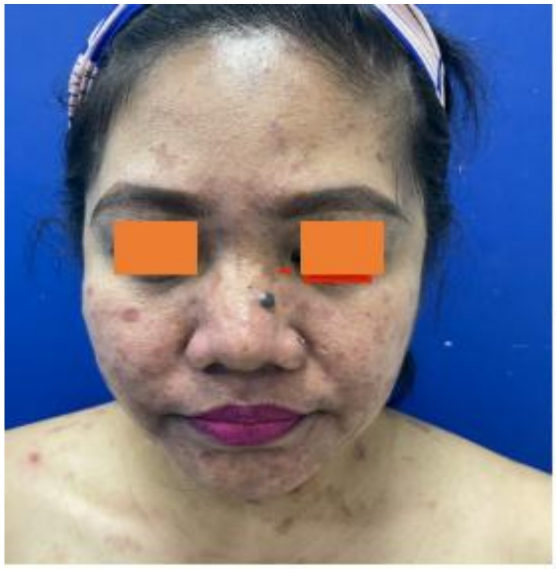

Figure 1-A

Figure 1-B

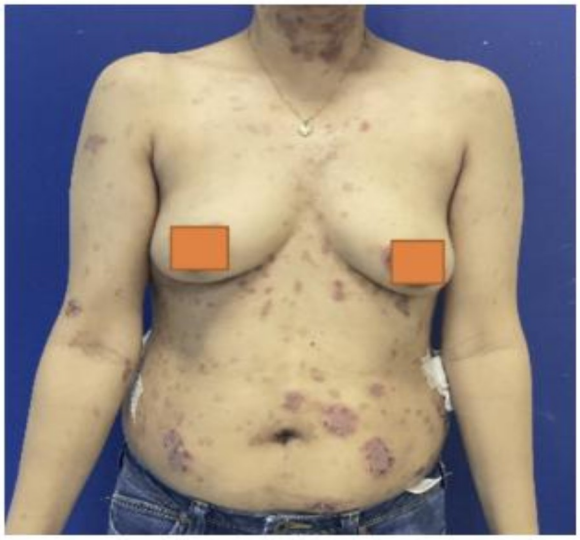

Figure 1-C

Figure 1-D

Figure 2-A

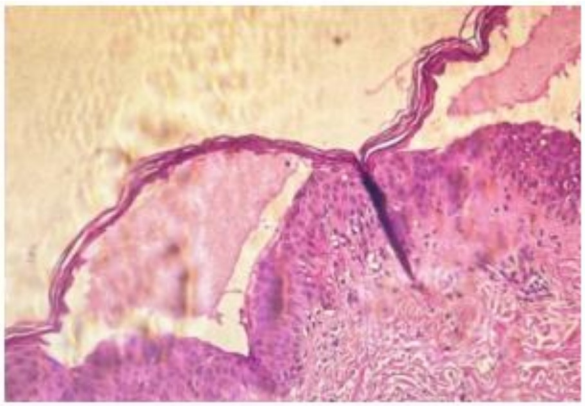

Figure 2-B

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