IgA Pemphigus, Intraepidermal Neutrophillic Type in an 8-year Old Filipino Female: A Case Report*

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ABSTRACT

Introduction: IgA pemphigus is a rare, chronic, relapsing, benign group of autoimmune intraepidermal blistering dermatosis with an unknown etiology. It is characterized by significantly pruritic, vesiculopustular lesions that occur mainly on the trunk and proximal extremities. Histopathologic and immunofluorescence studies show intraepidermal blisters and deposition of immunoglobulin A in the intercellular spaces of the epidermis, respectively.

Case Report: To our knowledge, we present the first reported pediatric case of IgA pemphigus, intraepidermal neutrophilic type, in an 8-year old Filipino female with a 2-year history of generalized papules and flaccid pustules, some forming an annular pattern. Diagnosis was confirmed by histopathology and direct immunofluorescence. Enzyme-linked immunosorbent assay for Desmoglein 1 was negative. Complete clearance of lesions was achieved with dapsone, colchicine and prednisone.

Keywords: IgA Pemphigus, Intraepidermal neutrophilic dermatosis

INTRODUCTION

IgA pemphigus is a rare bullous dermatosis with only 70 cases reported worldwide.[1] It is characterized by pruritic, vesiculopustular eruption caused by epidermal deposition of IgA autoantibodies.[3] IgA pemphigus is currently divided into two subtypes: intraepidermal neutrophilic dermatosis-type (IEN-type) and subcorneal pustular dermatosis-type (SPD-type).[2] To our knowledge, this is the first reported case of IgA pemphigus, intraepidermal neutrophilic type in the Philippines.

CASE REPORT

An 8-year-old female presented with a 2-year history of pruritic vesicles and pustules, some coalescing to form annular patterns. Lesions initially appeared on both lower extremities, eventually becoming generalized. Patient was seen and unsuccessfully treated by several physicians until she was referred to Pediatrics as a case of Systemic Lupus Erythematosus. However, clinical and laboratory evaluations were unable to support said diagnosis. The patient was subsequently referred to Dermatology department for evaluation.

Physical examination revealed generalized involvement, with sparing of the palms and soles. Lesions consist of multiple, well-defined, erythematous to hyperpigmented papules, pustules and annular plaques that coalesced becoming polycyclic, with central clearing and erythematous,

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scaly, indurated borders (FIGURE 1). Oral and genital mucosa were uninvolved. Potassium hydroxide mount revealed long septated hyphae. Initial skin biopsy demonstrated psoriasiform dermatitis with intraepidermal pustules (FIGURE 2). Initial impression was generalized tinea corporis. Patient was started on oral and topical antifungals and antibiotics, with partial improvement.

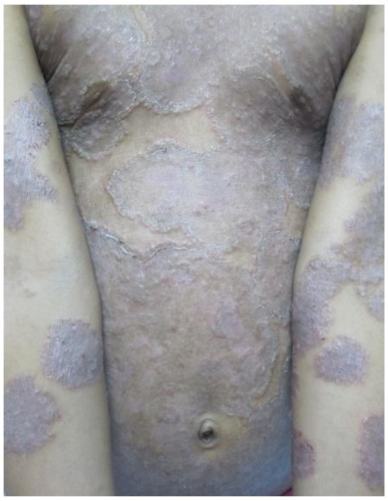

Figure 1. Multiple, well-defined, erythematous to hyperpigmented papules, pustules and annular plaques that coalesced becoming polycyclic, with central clearing and erythematous, scaly, indurated borders.

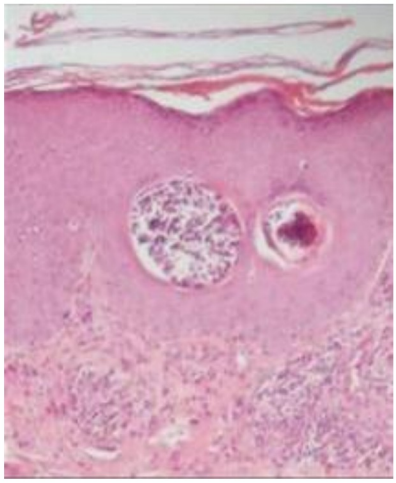

Figure 2. Intraepidermal vesiculopustular d ermatitis with predominantly neutrophilic infiltrates.

On subsequent visits, multiple flaccid pustules, some coalescing to form annular patterns, were noted (FIGURE 3). Healed lesions presented with hyperpigmented patches. At this time, an autoimmune blistering disease at was considered. Previous medications were discontinued and patient was started on oral prednisone at 1.5mg/kg/day. Repeat skin biopsy showed intraepidermal vesiculopustular dermatitis with predominant neutrophilic infiltrates (FIGURE 4). DIF revealed abnormal granular intercellular deposits of IgA (+2) in the epidermis (FIGURE 5). No abnormal deposits of IgG, IgM, C3 and Fibrinogen were seen in the epidermis, basement membrane zone or dermal vasculature. Enzyme-linked immunosorbent assay for Desmoglein 1 was negative. Final diagnosis was IgA pemphigus, intraepidermal neutrophilic (IEN) type.

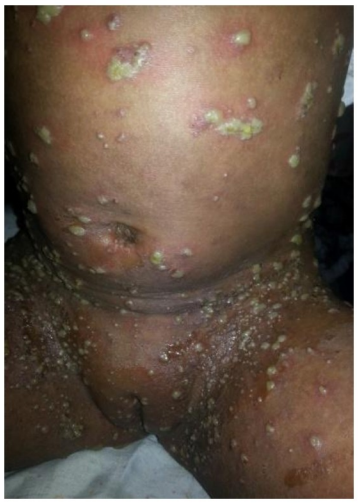

Figure 3. Multiple flaccid pustules, some coalescing to form annular patterns

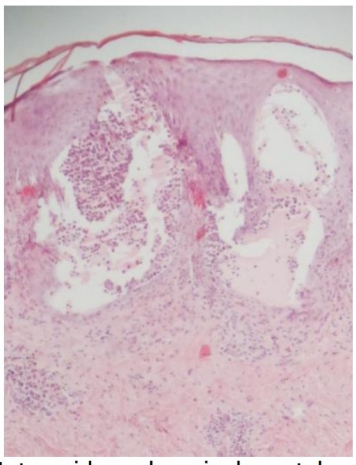

Figure 4. Intraepidermal vesiculopustular dermatitis with predominantly neutrophilic infiltrates.

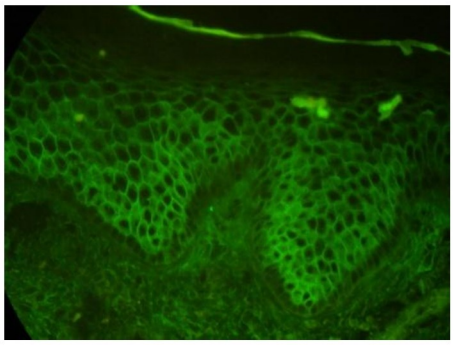

Figure 5. Abnormal granular intercellular deposits of IgA (+2) in the epidermis.

After determining that glucose-6-phosphate dehydrogenase level was normal, oral Dapsone at 1.25mg/kg/day was added after tapering the dose of prednisone to 0.5mg/kg/day, resulting in clearance of lesions within 3 weeks with few new blisters. Colchicine at 0.5mg/day was also added, which resulted in complete resolution of symptoms after 10 weeks of treatment (Figure 6a&b).

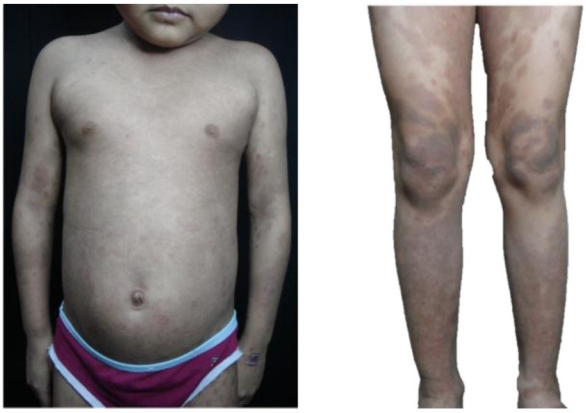

Figure 6A&B. Complete resolution of symptoms presenting as multiple, well defined, hyperpigmented patches.

DISCUSSION

IgA pemphigus is a rare, chronic, recurrent, benign autoimmune blistering dermatosis, commonly occurring in the middle age group, rarely in children.[2,4,6] The initiating mechanism is unknown.[8] Patients present with significantly pruritic flaccid pustules or vesicles that coalesce into annular, circinate or bizarre serpinginous configuration on an erythematous or normal skin.[8,9,10] The eruption tends to occur symmetrically with no atrophy or scarring, but occasionally leaves hyperpigmentation.[4] Sites of predilection are the trunk and proximal extremities, however, the scalp, postauricular, and intertriginous areas may be affected. Mucosal involvement is rare.[2,8]

IgA pemphigus is subdivided into: subcorneal pustular dermatosis (SPD) type, which targets desmocollin 1 (Dsc1), and the intraepidermal neutrophilic (IEN) type, the target antigen of which is still unknown (probably a nondesmosomal cell surface protein).[2] Although, it was reported that desmoglein 1 was a target antigen in an adult case of IEN type, this tested negative in our patient. [5,6] Subcorneal pustules with intercellular IgA deposits restricted to the upper epidermis characterize the SPD variant while the IEN variant shows deep epidermal pustules and intercellular IgA deposits in the lower or entire epidermis, as seen in our case.[7] Both demonstrate sparse, superficial acantholysis and a significant neutrophilic infiltrate.[2]

IgA pemphigus can rarely be associated with other medical disorders, including IgA monoclonal gammopathy, pyoderma gangrenosum, rheumatoid arthritis, Sjogren syndrome, Crohn's disease, gluten-sensitive enteropathy and HIV infection.[9,11,12]

In terms of treatment, dapsone is the drug of choice, in contrast to other forms pemphigus (i.e. IgG pemphigus) that respond mainly to steroids. The initial dosage of dapsone is 50-150 mg daily, with complete remission most often obtained.[1,8] In cases of dapsone intolerance, sulfapyridines and retinoids (acitretin) are alternatives.[8,11,13] Systemic corticosteroids are less effective, although it can suppress generalized flares when given at high doses.8,16 Responses using colchicine, azithromycin, cyclosporine, PUVA, and infliximab have also been reported in some cases.[8, 14, 15]

CONCLUSION

To our knowledge, this is the first reported pediatric case of histopathology and DIF-confirmed IgA pemphigus in the Philippines, with excellent response to dapsone and colchicine. This entity should be considered in patients presenting with pruritic vesicles to flaccid pustules forming circinate crusted plaques, with sites of predilection on the trunk, intertriginous areas and scalp.

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