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Case Report

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RITUXIMAB IN STEROID RESISTANT CHILDHOOD PEMPHIGUS

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ABSTRACT

Pemphigus vulgaris is an autoimmune vesico-bullous disorder involving the skin and mucous membrane characterized by painful erosions in the mucosae as well as flaccid vesiculo bullae on the skin. Pemphigus can affect all age group. The exact prevalence of paediatric pemphigus is unknown, but probably it affects 1.4 – 3.7% of total cases of pemphigus.[1][2][3] Rituximab is increasingly used in adult patients with pemphigus vulgaris who are nonresponders to conventional line of therapy.[4] However there is limited clinical data on safety and efficacy of rituximab in the paediatric age group. Here, we report a 15-year-old boy of pemphigus vulgaris who was treated with rituximab and achieved complete remission

KEYWORDS: Pemphigus, Rituximab, Paediatric.

INTRODUCTION

Pemphigus is a group of autoimmune disease characterized by flaccid vesicles and bullae due to loss of adhesion between keratinocytes.[5] Most common type of pemphigus in both adult as well as in children is pemphigus vulgaris (PV). PV in children aged less than 12 years is known as childhood PV and in aged between 12-18 years as juvenile PV.[6][7] Rituximab, a chimeric monoclonal anti-CD20 antibody, targets an integral membrane protein involved in B-cell activation and proliferation.[8] Rituximab destroys B cells mainly through antibody-dependent cell-mediated cytotoxicity; other mechanisms include-complement-mediated lysis, direct disruption of signaling pathways and triggering of apoptosis. Use of rituximab has been indicated in pemphigus patients who fail to respond to conventional treatment or when their use is contraindicated. The course and treatment modalities of juvenile pemphigus are essentially the same as those in adults. Although corticosteroids and immunosuppressive drugs are considerd as the 1st line of therapy, Some juvenile pemphigus patients are refractory to this conventional treatments and require additional therapies as seen in our case.[7]

CASE HISTORY

A 15-year-old boy presented with 18-month-history of painful oral erosion and flaccid fluid-filled blisters predominantly on the face, upper trunk and groin [fig-1] which ruptured to form progressive erosions with crusting, Biopsy showed suprabasal cleft and acantholytic cells and was diagnosed as a case of juvenile pemphigus. Patient was started on prednisolone 1mg/kg body weight for 10 months. Initially there was a good response but later on patient again started developing active lesion in the oral cavity,chest and on thighs[fig-2]. At this time, indirect immunoflorescence for desmoglein was done and found to be 175.40 for desmoglein (dsg1)[negative -<20] and 139.50 for desmoglein(dsg3)[negative-<20] Considering a recalcitrant case[fig-3],[fig-4]. Patient was investigated for pre rituximab evaluation. In view of the relapse occurring on steroids alone patient was also strated on low dose rituximab, at a dose of 375 mg/m^2 at 15 days interval.[1][4] The patient showed good response, recalcitrant lesions healed within 3 weeks with hyperpigmentation[fig-5]. and post rituximab dsg1 was found to be 4.05 and dsg3 was 50.29. After the pulse therapy of rituximab he was on maintenance therapy with azathioprine 50 mg for 1 year. At this time his dsg levels are significantly lowered [dsg 1 <2, dsg3 7.24]. Now he is in clinical remission at 8 months of follow-up period, and all the treatment is stopped at present.

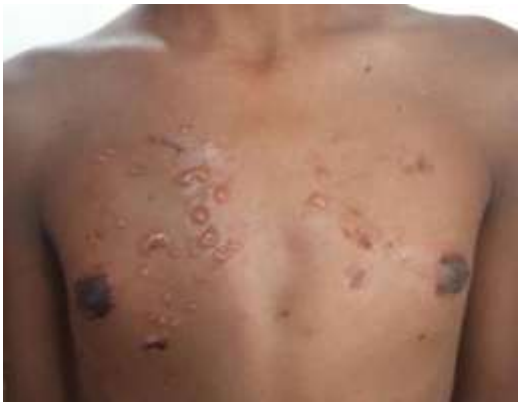

Figure-1.

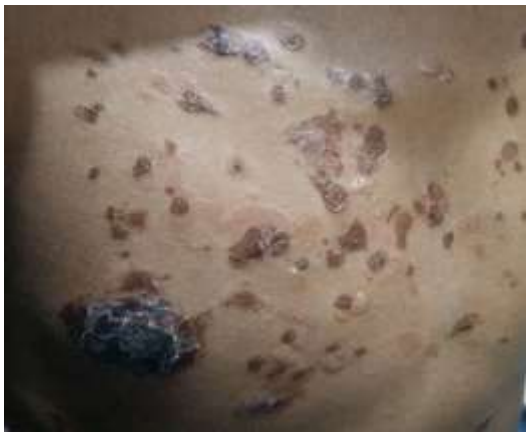

Figure-2.

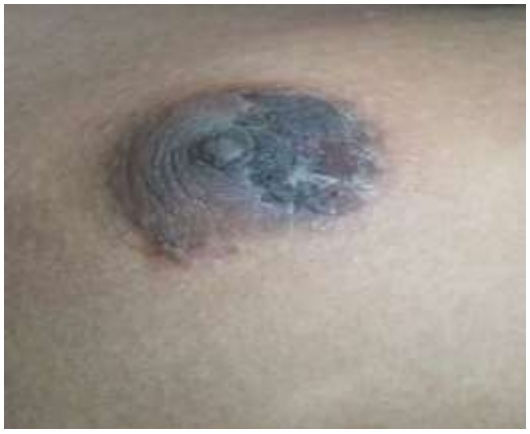

Figure-3.

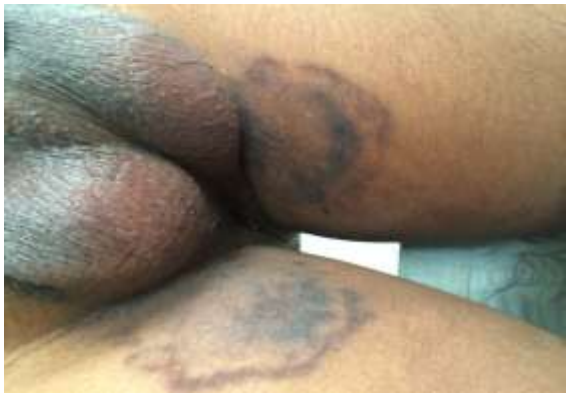

Figure-4.

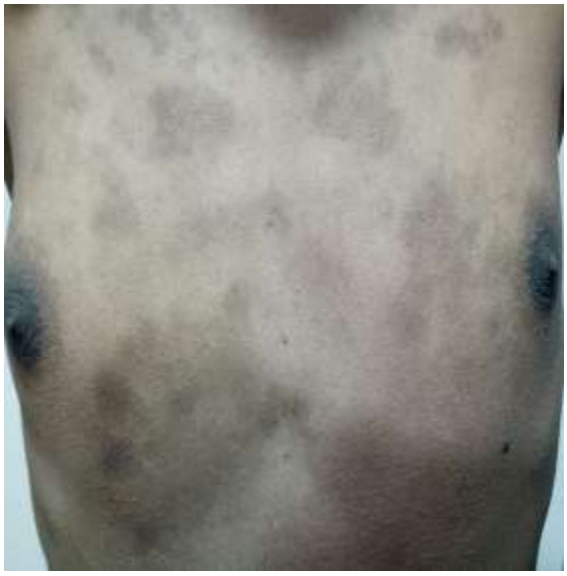

Figure-5.

DISCUSSION

Treatment of pemphigus vulgaris is challenging. The first line of therapy is systemic corticosteroids, however use of corticosteroids in paediatric age group is associated with serious risk. Osteoporosis, adrenal suppression, hyperglycemia, dyslipidemia, cardiovascular disease, cushing syndrome, psychiatric disturbance and immunosuppresion are amongst the most serious side effects specially when used for long term in high doses.[9][10] Immunosuppresive drugs can be used to control the disease but these first line of therapy can give rise to increased morbidity and poor control of disease. Recent studies have shown long term safety and efficacy of rituximab in pemphigus patients. Paediatric use of rituximab is still limited and there are very few case reports of the same. Our patient had persistent painful lesion over groin which did not improve with the conventional treatment modalities. Therefore, balancing the potential benefit and risk, we consider to start him on with low dose of 2 cycles of rituximab 375 mg/m^2 at an interval of 15 days.[4] Our case achieved clinical remission. Duration of therapy with rituximab is shorter in juvenile PV and it also associated with decreased mortality(2.9%) when compared with adults(10-15%).[9] So we believe low dose rituximab also can be effective and safe in treating childhood pemphigus patient in Indian scenario.

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