AUTOIMMUNE BULLOUS DISEASES

SUCCESSFUL USE OF RITUXIMAB IN A CASE OF JUVENILE PEMPHIGUS VULGARIS ASSOCIATED WITH HETEROTAXY SYNDROME AND HEPATITIS B

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Background: Pemphigus vulgaris is a chronic autoimmune blistering disease characterized by presence of antibodies against desmosomal adhesion proteins. Juvenile pemphigus vulgaris(JPV) are usually rare and severe in presentation. The exact mechanism of autoimmunity has not been defined but role of infectious triggers like hepatitis B and C in genetically susceptible individuals has been defined. Usually juvenile pemphigus are severe and resistant to conventional treatments. Here by we present a case of JPV with hepatitis B positive serology refractory to conventional therapy and successfully treated with rituximab.

Observation: A 17 year old girl presented with history of multiple painful mucosal erosions and flaccid blisters over the body since 1 year. A diagnosis of pemphigus vulgaris was made and confirmed with both skin biopsy and direct immunofluorescence findings. After 6 cycles of Dexamethasone pulse, patient developed steroid induced psychosis which forced to start of steroid sparing immunosuppressants with no satisfactory response. Patient was planned for rituximab, was found to be HbsAg positive, anti HCV antibody negative and with normal HBV DNA levels. Further on ultrasonography and CECT Abdomen patient had malrotated gut, annular pancreas, polysplenia and diagnosed as Heterotaxy syndrome. Patient was started on tenofovir 300 mg once daily along with oral corticosteroids at 0.5mg/kg/day which slowly tapered and disease activity well controlled. After 9 months of tenofovir therapy and tapering doses of oral corticosteroids , patient started developing multiple new lesions. After complete routine workup, low dose rituximab was given each 500mg, two weeks apart. Patient was completely lesion free after 6 months and no adverse effects seen.

Key message: Juvenile pemphigus are severe and refractory to conventional therapies. Complete routine laboratory investigations along with hepatitis serology should be done in all the patients. Newer biological therapies like rituximab is beneficial with minimal side effects especially in juvenile pemphigus.