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Hyper-Immunoglobulin E (Hyper-Ige) Syndrome In Skin Of Color: A Retrospective Single-Centre Observational Study

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Abstract : Introduction: Hyper-IgE syndrome is a rare primary immunodeficiency syndrome characterised by triad of severe atopic dermatitis, recurrent pulmonary infections, and recurrent staphylococcal skin infections. The diagnosis requires a high degree of suspicion, typical clinical features, and not mere rise in serum-IgE levels, which may be seen in multiple conditions. Genetic studies are not always possible in a resource poor setting. This study highlights various presentations of Hyper-IgE syndrome in skin of color children. Case-series: Our study had six children of Hyper-IgE syndrome aged twomonths to tenyears. All had onset in first ten months of life except one with a late-onset at two years. All had recurrent eczematoid rash, which responded poorly to conventional treatment, secondary infection, multiple episodes of hospitalisation for pulmonary infection, and raised serum IgE levels. One case had occasional vesicles, bullae, and crusted plaques over both the extremities. Genetic study was possible in only one of them who was found to have pathogenic homozygous deletions of exon-15 to 18 in DOCK8 gene following which he underwent bone marrow transplant (BMT), however, succumbed to lower respiratory tract infection two months after BMT and rest of them received multiple courses of antibiotics, oral/ topical steroids, and cyclosporine intermittently with variable response. Discussion: Our study highlights various characteristics, presentation, and management of this rare syndrome in children. Knowledge of these manifestations in skin of color will facilitate early identification and contribute to optimal care of the patients as representative data on the same is limited in literature.

Keywords : absolute eosinophil count, atopic dermatitis, eczematous rash, hyper-immunoglobulin E syndrome, pulmonary infection, serum IgE, skin of color

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