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Features of Immune Dysregulation in Primary Immunodeficiency Patients at Soba University Hospital, Khartoum – Sudan

Goda, R.[1]*, Mohamed, A. S I.[1], Salim A[1], Masoud S O[1], Erwa N HH[1, 2, 3]
*rayangoda@gmail.com**
*Sudan**

[1] *Immunology and Allergy Unit, Soba University Hospital, Khartoum – Sudan*
[2] *Faculty of Medicine, Department of Medical Microbiology, University of Khartoum, Khartoum Sudan*
[3] *Clinical Immunology Specialty Council, Sudan Medical Specialization Board, Khartoum, Sudan*

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Abstract

Background: We reviewed all patients with primary Immunodeficiency diseases (PID) that presented to immunology clinic at our hospital for features of immune dysregulation.

Purpose: To describe the clinical manifestations and basic immunological findings in Sudanese patients with features of immune dysregulation. Additionally, we looked into how these findings affected their management and outcome.

Methods: Retrospective analysis of all patients referred with PID looking for features of immune dysregulation: autoimmunity, allergy, lymphoproliferation and cytopenias.

Results: Total number of patients with suspected PID was 120 patients. Sixteen patients had features suggestive of immune dysregulation in the form of autoimmunity evident as autoimmune cytopenia, endocrinopathies, lymphoproliferation, inflammatory bowel disease, enteropathy, arthropathy and allergy. Most reported allergic features included eczema and food allergy. Half of the patients had altered immune phenotypes with low immunoglobulin levels; either IgG or IgA (5 patients), increased IgE was noted in 4 patients. Four patients (25%) had low CD4 helper T cells, increased double-negative T (DNT) cells were seen in 4 patients and one patient had low CD4+CD25+FOXP3+ T cells. Management proved difficult due to recurrent infections, medication side effects or refractory disease. Almost half of the patients were lost to follow up, with 3 deceased patients.

Conclusion: Immune dysregulation in patients with PIDs is poorly identified and hence under reported in Sudan and Africa. Inter-clinic collaboration is recommended to identify more patients, with the provision of better diagnostic and therapeutic modalities including targeted therapy and specific genetic diagnosis.

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