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First case of Leukocyte Adhesion Defect (LAD1) confirmed by Flow Cytometry in Sudan

Suhad Abdelrahman^{1,2,3}, Erwa N HH^{4,5,6}

1 Paediatrics Unit, Soba University Hospital

2 Pediatric and child health council, Sudan Medical Specialization Board

3 Membership of Royal College of Paediatric and child health

4 Immunology and Allergy Unit, Soba University Hospital

5 Faculty of Medicine, Department of Medical Microbiology, University of Khartoum

6 Clinical Immunology Specialty Council, Sudan Medical Specialization Board, Khartoum, Sudan⁴

Abstract

Background: There are 3 types of Leukocyte Adhesion Defects (LADs) which are rare primary immunodeficiency disorders with compromised neutrophil adhesion and transmigration to infection or inflammation sites resulting in recurrent infections. Here we present a child with severe LAD1 which is reported to occur in 1 in 10 million births. Only about 300 cases have been reported worldwide. With limited facilities in Sudan no cases were confirmed or reported to the literature before.

Objective: To report the first case of LAD 1 in Sudan diagnosed by flow cytometric measurement of surface CD11b/CD18.

Methods: evaluating a Sudanese boy who presented at the immunology clinic aged 53 days following surgical treatment for complicated non healing umbilical granuloma with omphalitis, and fistulised anal abscess, with perianal fasciitis, for which he had a colostomy. He was born to first degree consanguineous parents. He had three siblings lost to similar presentations.

Results: CBC showed marked leucocytosis of both neutrophils and lymphocytes. Wound and blood culture revealed Pseudomonas and Staph aureus. Serum IgG and IgM were significantly raised. Lymphocyte subsets confirmed the lymphocytosis. CD18 expression on neutrophils and monocytes was significantly reduced (3.20%) with marked reduction/loss of CD 11c (1.10%). CD15, CD11a and CD11b expression was normal.

Conclusion: We report the first LAD 1 case confirmed by flow cytometry in Sudan. The patient was planned for stem cell transplantation but died at age of 74 days highlighting the need for a National PID care centre.