

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

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Background

There are 3 types of Leukocyte Adhesion Defects (LADs) which are rare primary immunodeficiency disorders with compromised neutrophil adhesion and migration to infection or inflammation sites. This leads to recurrent infections. 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 previously.

Purpose

To report the first case of LAD 1 diagnosed in Sudan using the flow cytometer to assess the expression of surface CD11b/CD18.

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Methods

Evaluating a 53 days old Sudanese boy who presented at the immunology clinic following surgical treatment for complicated non healing umbilical granuloma with omphalitis, and fistulised anal abscess, with perianal fasciitis, for which he had a colostomy (Figure 1). He was born to first degree consanguineous parents. He had three siblings lost to similar presentations.

Figure 1:
Infected Colostomy



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%) (figure 2), with marked reduction/loss of CD 11c (1.10%) (Figure 3). CD15, CD11a and CD11b expression was normal.

Figure 2:
Lack of D18 Expression

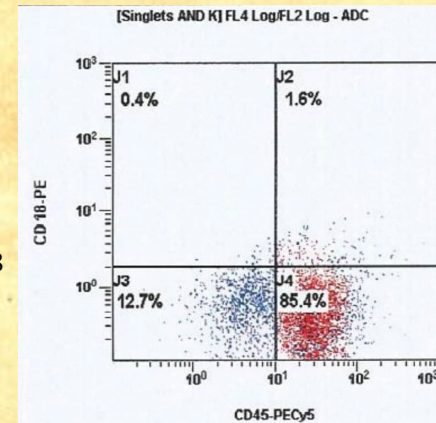
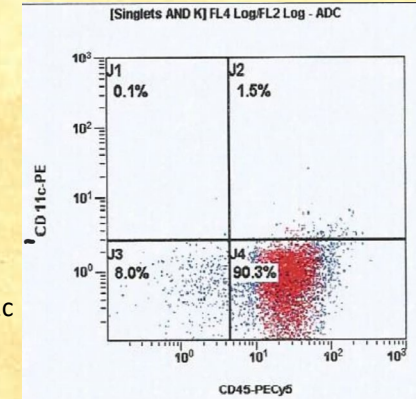


Figure 3:
Lack of CD 11c Expression



Conclusions

Flowcytometric diagnosis of LAD defects is confirmatory of a typical LAD phenotype and should be sought early in the disease course.

We report the first LAD 1 case confirmed by flow cytometry in Sudan. His plan included bone marrow transplantation abroad but died aged 74 days highlighting the need early diagnosis and a National PID Centre.



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