PAEDIATRIC DERMATOLOGY

LEUKOCYTE ADHESION DEFICIENCY TYPE 1 PRESENTING WITH PYODERMA GANGRENOSUM: A DIFFICULT TO MANAGE RARE CONDITION

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Background: Leukocyte adhesion deficiency type 1 (LAD 1) is an autosomal recessive disorder often presenting with erythematous plaques with necrotic ulcerations showing no visible pus or microscopic neutrophils. Pyoderma gangrenosum (PG) also presents with a necrotic ulcer but the base is purulent with neutrophilic infiltration. Very rarely both the disorders may present simultaneously

Observation: Our patient a 6 years old male, product of consanguineous marriage, presented with a non healing ulcer over left buttock and groin area for last two months. On examination there was a necrotic ulcer of size 10×5 centimeters with bluish margin and seropurulent discharge. Some scars of the previously healed lesions were present over the right buttock and left knee. We considered Pyoderma gangrenosum and Leukocyte adhesion defect type 1 as differential diagnoses. Skin biopsy showed collection of polymorphs in upper dermis. Stains for fungi and mycobacteria were negative. Tissue cultures were sterile. Pus culture showed growth of different organisms at different time. Total leukocyte counts were 40000/micro liter with neutrophillia. Blood sample was sent for flow-cytometric immunophenotyping for CD18+ Leukocytes on neutrophils. It showed decreased CD18 expression (control 99.95%, travel control 99.56%, patient 03.60%) which was consistent with leukocyte adhesion deficiency type 1. Patient was managed with cyclosporine and IV antibiotics according to pus culture sensitivity. He developed similar ulcerative lesions over the site of IV line also raising suspicion of pathergy.

Key Message: Proper diagnosis of a non healing necrotic ulcer in a pediatric patient is very important. PG is managed with immunosuppressants and LAD1 is managed with antimicrobials. But PG with LAD1 should be managed with a blend of immunomodulators and antimicrobials. High dose of steroids should be avoided as the patient may succumb to infections. Stem cell transplantation can improve the immune deficiency state of LAD1 patients.