

DISSEMINATED BCG INFECTION RESEMBLING LANGERHANS CELL HISTIOCYTOSIS IN AN INFANT WITH SEVERE COMBINED IMMUNODEFICIENCY: A CASE REPORT

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Purpose: The purpose of this report is to present a case of rare congenital immunologic disease, severe combined immunodeficiency disease (SCID) T-/B+/NK- in which routine BCG vaccination led to life threatening dissemination. **Patient and methods:** A 6-month-old-boy presented with prolonged mucocutaneous candidiasis, severe anemia, skin lesions resembling the infiltrative eczema of Langerhans cell histiocytosis (LCH), rash and subcutaneous nodules with histiocytic infiltration. Pathologic specimens obtained from surgical excision of a subcutaneous nodule were examined. Numerous S-100 negative and CD1a negative histiocytes were seen containing BCG intracellular growth. Histopathology and immunohistochemistry confirmed the diagnosis of BCG dissemination. Laboratory findings show profound absence of humoral and cell-mediated immunity. Immunoglobulin levels and T cell numbers were consistently low, while the percentage of B-lymphocytes was increased. The patient died at eight months of age from BCG dissemination with multiple organ damage. **Conclusion:** Disseminated cutaneous BCG infection may resemble to the infiltrative eczema of LCH that may mislead the clinician. Diagnostic biopsy is necessary, especially in countries where BCG is routinely administered in infancy. Furthermore, efforts to improve early diagnosis of SCID and bone marrow transplantation are essential to prevent the occurrence of end organ damage secondary to infective complications and BCG dissemination.

