Academic Journal of
Pediatrics & Neonatology
ISSN 2474-7521

Juniper Publishers
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Case Report
Volume 13 Issue 4 - January 2024
DOI: 10.19080/AJPN.2024.13.555921

Acad J Ped Neonatol

Disseminated BCG Disease in Severe Combined Immunodeficiency Infant

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Submission: January 11, 2024; **Published:** January 23, 2024

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Abstract

We report a 7-month-old infant presented with persistent fever, weight loss, anemia and hepatosplenomegaly. He received BCG vaccine at the age of 3 months. The infant was diagnosed with SCID with disseminated BCG disease (BCGiosis).

Keywords: BCG; Tuberculosis; SCID; Vaccine; Disseminated

Abbreviations: SCID: Severe Combined Immunodeficiency; HSCT: HEMATOPOIETIC Stem Cell Transplantation; TB: Tuberculosis; BCG: Bacillus Calmette-Guerin; MTB: Mycobacterium Tuberculosis

Introduction

BCG is a live attenuated vaccine that is administered routinely in the neonatal period or early infancy in TB endemic countries. Usually, BCG is associated with local complications only but patients with severe combined immunodeficiency (SCID) are predisposed to live attenuated vaccines related lethal complications, especially BCG leading to disseminated disease (BC Giosis). SCID is the most severe and lethal inherited primary immunodeficiency with a prevalence of approximately 1 in 50,000 live births. It is called a pediatric emergency as it invariably leads to fatality in infancy without early aggressive therapy and Hematopoietic stem cell transplantation (HSCT) or other specific therapy [1] (Figure 1).

Case Presentation

A 7-month-old Kuwaiti boy born to non-consanguineous parents with positive family history of immunodeficiency in the maternal uncle. He was admitted with 23 days history of fever which did not respond to multiple courses of oral antibiotics and antipyretics. There was no history of skin rash, abnormal movements, cough, vomiting or loose motions. There was no history of urinary symptoms, raw milk ingestion, travelling abroad or contact with sick patients. On examination the baby was vitally stable, hydrated, pink in RA with no signs of respiratory distress or dysmorphism. His growth parameters were in the 10th centile on admission to the hospital. He had mild hypotonia and hepatosplenomegaly with unremarkable other systemic examination. Septic work up was normal including blood, urine and CSF cultures and sensitivity. His CBC showed lymphopenia with normocytic anemia. Blood film was negative for blast cells. Immunology works up was done as a part of pyrexia of unknown origin. Lymphocyte phenotyping showed markedly low IgG levels and lymphocyte phenotyping showed absent T & B Cells which is a picture of SCID. The boy started to receive monthly IVIG and was kept on sceptrin and fluconazole prophylaxis upon immunologist recommendation till arranging hematopoietic stem cell transplantation (HSCT). The fever did not come down and the patient developed generalized oedema including pericardial effusion confirmed by echocardiography and there was hypoalbuminemia. CT chest and abdomen was done with picture showing hepatosplenomegaly, moderate ascites, mild pericardial effusion and bilateral lung basal atelectatic changes with infective foci in lower bases. GeneXpert MTB was positive, and he started anti-TB therapy including isoniazide, rifampicin, ethambutol and Moxifloxacin with marked response in his fever pattern, weight gain and general condition. The boy was then referred for HSCT (Figure 2).

Discussion

BCG is a live attenuated vaccine that protects from miliary tuberculosis and tuberculous meningitis. Severe combined immunodeficiency (SCID) is the most severe form of primary immunodeficiency diseases characterized by a block in T lymphocyte differentiation that is variably associated with the abnormal development of other lymphocyte lineages, i.e., B and

NK lymphocytes [2]. Though BCG vaccine is generally safe, an important complication is a life-threatening disseminated BCG infection especially in children with primary immunodeficiency [3]. The estimated incidence of disseminated disease was 2 cases per million vaccinated children, and the mortality rate was 80% [4]. Definitive systemic symptoms, such as fever or sub-febrile states, weight loss, or stunted growth, and at least two areas of involvement beyond the site of a BCG vaccination, such as lymph nodes, skin, soft tissues, lungs, spleen, liver or bones [5]. Our patient fulfilled the criteria as he had fever, weight loss, hepatosplenomegaly plus positive GeneXpert MTB which was recommended by WHO in 2010 for the diagnosis of pulmonary tuberculosis and subsequently in 2013 for the diagnosis of extra-pulmonary tuberculosis [6]. Those patients should receive anti-tuberculous treatment including four or more anti-TB drugs, until full recovery then, a prophylactic program with two drugs should be continued, until complete immunological reconstitution after HSCT is achieved [7] (Figure 3).

Figure 1

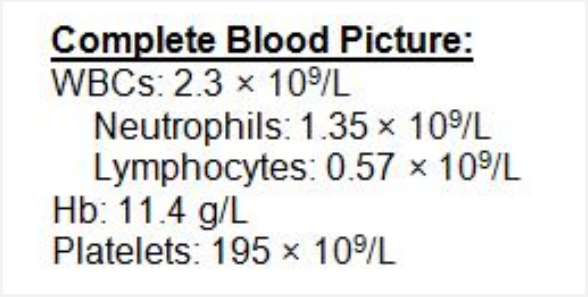

Figure 2

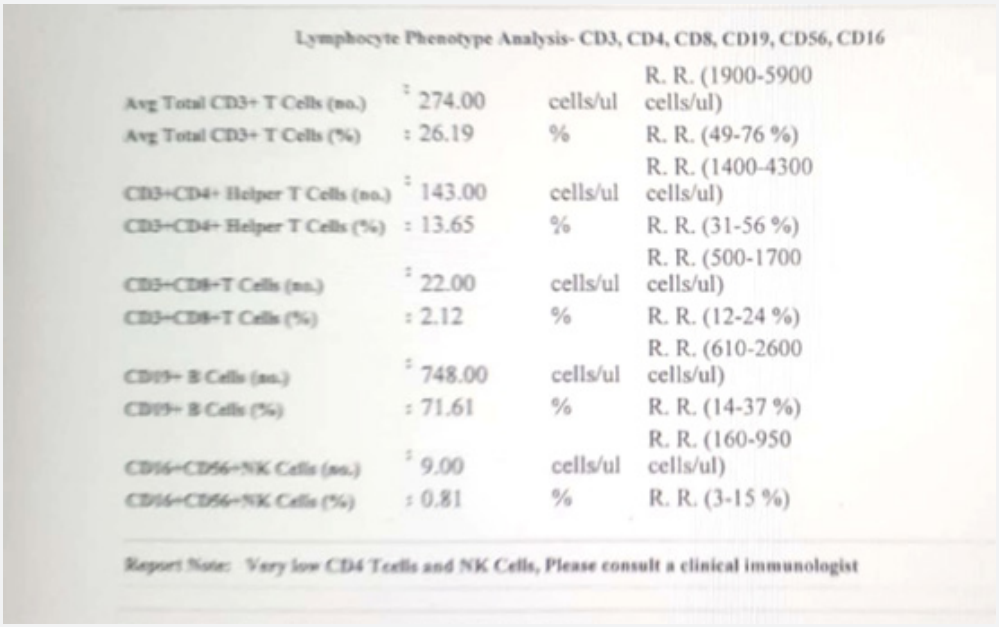

Figure 3

How to cite this article: Nashaat Elsayed F, Taha Mohamed D, Hatem Ibrahim A. Disseminated BCG Disease in Severe Combined Immunodeficiency Infant. Acad J Ped Neonatol 2024; 13(4): 555921. DOI: 10.19080/AJPN.2024.13.555921

Conclusion

BCG disease is an important presentation of primary immunodeficiency, especially SCID after BCG vaccine. Neonatal screening for lymphopenia should be considered to avoid late diagnosis of SCID and organ damage. We recommend delaying BCG vaccine to be after 6 months of age especially for those with family history of immunodeficiency or recurrent infections.

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