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Widespread BCGosis in a child with Severe Combined Immunodeficiency is complicated by Strong Immune Reconstitution upon Transplant

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Abstract

Individuals with primary immunodeficiency disease (PID) are more susceptible to adverse reactions after receiving the BCG vaccine, ranging from localised disease (BCGitis) to disseminated disease (BCGosis).

Keywords: BCGosis; BCGitis; Severe combined immunodeficiency; Bone marrow transplant

Introduction

The live attenuated BCG vaccine prevents tuberculosis (TB). It has been around for about 90 years. In nations where TB is endemic, it is given to all newborns and infants as a part of the national childhood immunisation programme. Rare but terrifying side effects, including local/regional illness (BCGitis) and distant/disseminated infection (BCGosis), have sporadically been documented, especially with immunocompromised people. Compared to other newborns, patients with severe combined immunodeficiency (SCID) have a higher prevalence of BCG illness after immunisation **[1-2]**. After hematopoietic stem cell transplantation, disease flares frequently happen (HSCT). The probability of having BCG-related problems after HSCT was reported to be increased in individuals with lower natural killer (NK) cells and NK negative SCID mutations **[3].**

Methods

Although uncommon, BCGitis and BCGosis pose a serious threat to children with primary immunodeficiencies (PID). We describe a 7-month-old baby with SCID (JAK-3 exon-15 mutation). She had a history of sibling deaths from acute pneumonia infancy, a low absolute lymphocyte count (ALC 1000/cubic mm), and severe, chronic pneumonia when she was first seen. When there were no 10/10 matched family members or unrelated donors available, she had a haplo-identical bone marrow transplant (BMT) with ex-vivo T-cell depletion with her father as the donor after her clinical condition from pneumonia had stabilised. For conditioning, the drugs thiotepa, fludarabine, treosulphan, and anti-thymocyte globulin were utilised **[4-6]**. From D+5 to engraftment, the child received tacrolimus and mycophenolate mofetil. Ex-vivo T-cell depleted allogenic stem cells (with 0.2% TCR-alpha CD3 cells) were given to her at a rate of 10106 CD34 cells/kg.

After myeloablative treatment on day two following stem cell infusion, she experienced an erythematous painful swelling in the left deltoid area (the site of the BCG vaccination). Fever, axillary lymphadenopathy, hepatosplenomegaly, or other body swellings were absent. BCGitis was taken into consideration because of the involvement's typical place. Nonetheless, she was treated with broad-spectrum antibiotics, antifungals, in addition to the antituberculous therapy because she had severe neutropenia following conditioning chemotherapy (ATT: isoniazid, rifampicin, ethambutol and pyrazinamide) **[7-10]**. A left shoulder ultrasonogram showed a hypoechoic lesion that eventually developed into a thick-walled collection with a size of 91412 mm and 0.8 ml in the deep subcutaneous plane. On D+2, the USG chest and abdominal radiographs were both normal.

The wound was treated with a daily dressing after the pus was aspirated. On Zeihl Neilson staining and MGIT culture media, Pus displayed many acid-fast bacilli; Gene Xpert demonstrated positive with rifamipicin sensitivity. After 3 weeks of ATT, there was a notable clinical improvement, with considerable wound healing and a normalisation of the chest x-ray. ATT was kept going for a total of 12 months (HRZE for 2 months and HRE for 10 months). At the one-year post-HSCT follow-up, she is doing well and has gained enough weight.

Three primary immunodeficiency patients with immune reconstitution inflammatory syndrome (IRIS) to Mycobacterium Avium Complex (MAC), on D+9 to D+42 post-HSCT, were described by Manion et al. All three patients had elevated levels of C-reactive protein, IFN-, tumour necrosis factor (TNF-), IL-6, and IL-18, which is consistent with IRIS flares **[8].**

Three primary immunodeficiency patients with immune reconstitution inflammatory syndrome (IRIS) to Mycobacterium Avium Complex (MAC), on D+9 to D+42 post-HSCT, were described by Manion et al. All three patients had elevated levels of C-reactive protein, IFN-, tumour necrosis factor (TNF-), IL-6, and IL-18, which is consistent with IRIS flares **[8]**.

Different immunosuppressive regimens utilised for the prevention or treatment of GVHD are hypothesised to contribute to lower occurrences and less severe courses as compared to HIV-AIDS **[9,10].**

It can be difficult to distinguish between an infection that is getting worse and an exaggerated inflammatory reaction to an infection since the two conditions require quite different treatments. Tubeculostatic medication must be intensified in order to prevent BCG illness flares, which usually happen after HSCT; adjuvant immunosuppressive therapy may also be helpful.

Discussion

In underdeveloped nations, the World Health Organization (WHO) advises BCG vaccination from birth. The BCG vaccine is

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thought to be safe for healthy immune systems, although less than one in a thousand recipients experience substantial local reactions, and less than one in a million experience serious disseminated disease. According to previously published statistics, 50–76% of BCG-infected patients exhibit immunodeficiency. In a study by Ong et al., 10 patients with disseminated BCGosis were described. The median age at symptom onset was 3.8 months (0.8-7.4), and all of the patients had underlying PID, including four patients with SCID, three with mendelian susceptibility to mycobacterial diseases (MSMD), one with anhidrotic ectodermal dysplasia with PID (EDA-ID), one with combined immunodeficiency.

Conclusions

Infants with SCID should not receive the BCG vaccine. However, because the vaccine is given right away after birth, many patients are only identified as having SCID when BCG problems arise. In order to avoid adverse effects, including life-threatening disseminated BCGosis, screening for underlying immunodeficiency prior to live vaccination becomes essential in patients with a positive family history.

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