Disseminated tuberculosis complicating Bacillus Calmette–Guérin (BCG) vaccine as only presentation of Severe combined immunodeficiency (SCID). Report of three cases

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Background

Severe combined immunodeficiency disease (SCID) is a rare primary immunodeficiency disease. It is commonly manifest as an early onset of respiratory and intestinal infections like recurrent diarrhea, oral candidiasis, failure to thrive and/or interstitial pneumonitis(1)

Case description

We report a unique presentation of SCID in three male patients, outcome of consanguineous marriage and all received the BCG vaccine at birth. All three cases presented with regional lymphadenopathy at age of 3 months that progressed to generalize lymphadenopathy treated with anti-tuberculous. The first and second case were twins. The first case had uneventful history until the age of 33 month when he developed multiple Suppurative Tuberculous lymphadenitis confirmed by biopsy.

Case description

The second and the third cases were diagnosed with Disseminated Tuberculosis at age of 24 months as they developed fever, anemia, weight loss, tuberculous peritonitis and lymphadenopathy confirmed by biopsy. After excluding other immunodeficiency disease and based on the immunoflocytometry and immunoglobulin analysis, the first case was diagnosed as CD4,CD16 lymphopenic SCID, the second case as CD4, CD8, CD19, CD16 lymphopenic SCID with hypogammaglobulinemia and the third case as CD3, CD4, CD8 lymphopenic SCID with hypogammaglobulinemia. They received supportive management, anti-Tuberculous (Isoniazid, Rifampicin, Ethambutol, Ciprofloxacin , Clatirthromycin), prophylactic Trimethoprim/ Sulfamethoxazole and Immunoglobulin infusion. The patients at the time of writing this report are all alive and thriving normally and they did not have any other bacterial, viral or fungal infection, the twins are 3 years old and the third case is 30 month old.

Discussion

- The three reported cases despite being diagnosed with SCID they presented only with complicatiom of the BCG vaccine.
- BCG vaccine is the only vaccine available against tuberculosis. The vaccine is known to increases the mortality and morbidity in patient with SCID (2).
- The patients were diagnosed with possible BCG infection duo to the unavialbility of PCR primer of mycobacterium bovin in Sudan for a difinitive diganosis.
- The definitve treatment for SCID is haemopiotic Stem Cell Transplantation HSCT although none of our patients had recieved it, they are still alife and thriving very well.

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Conclusion

SCID may not exhibit the classical manifestation of recurrent infections, and it may present only as a complication of BCG vaccine alarming to maintain high susceptibility in such patients especially in Sudan were BCG vaccine usually given at birth.

References

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