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DISSEMINATED BCG INFECTION

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BCG vaccines are known to cause severe diseases in immunocompromised children. The impaired immunity specifically alters host defences against mycobacteria. Parental consanguinity and familial forms are frequent. The syndrome is often described as Mendelian susceptibility to mycobacterial infection. In most cases it is autosomal recessive inheritance.
Disseminated forms of BCG infection are heterogenous. Clinical forms correlate with the type of histopathological lesions present. Children with lepromatous like granulomas generally die of overwhelming infection, children with tuberculoid granulomas have favourable prognosis.
The disorders whose common pathogenic mechanism is impaired interferon gamma immunity, generally manifest in childhood. The severity of the clinical fenotype depends on the genotype.
The authors present cases of two girls with Disseminated BCG infection. Genetic analysis revealed that both girls have a defect in IL12 receptor, completely impairing its function, resulting in decrease of IL12 and subsequently IFNgamma. Further evaluation of the immunological status of the two patients revealed different reactivity of the cell mediated immune response, one child showing proliferative response to several mycobacterial antigens, the other child did not showing any recognition of mycobacterial antigens, suggesting a complete anergy of cells.
The cases of two girls stress the importance of being aware of any underlying immune defect that might be the cause for clinical symptoms of non healing inoculation sites and enlarged axillary lymph nodes. It also stresses the importance of effective treatment immediately after diagnosing a disseminated uncontrolled infection by BCG vaccine, consisting of a combination therapy of at least three anti TB drugs.