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10 year old boy with Idiopathic CD4 lymphocytopenia (ICL) and Cervical node MDR-TB - A rare case report

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Abstract : Idiopathic CD4 lymphocytopenia (ICL) is defined as CD4 count 300 cellsmm3 or a CD4 count that is 20 percent of the total T cell count on 2 occasions, with absence of HIV infection and absence of any defined immunodeficiency. We report a 10 year old boy who presented with complaints of multiple swellings with discharging sinus in the neck with history of ATT intake since the age of two and half years due to non resolution of swelling with sinus. Culture and sensitivity test for MTB was done and was diagnosed as Extra pulmonary (cervical node) MDR-TB. In this child the CD4 counts at 6 months interval met CDC criteria for ICL (155 Cellsmm3 in Dec, 2010 and 147 cellsmm3 in May, 2011). Reversal of CD4CD8 ratio was present. Patient has no other opportunistic infection except for cervical node MDR-TB. To our best knowledge this is one of those rare cases in paediatric age group which warrants a mention, not only because of CD4 depletion and but also presence of extra pulmonary (cervical node) MDR TB in a 10 year old boy. The child is now on Interferon and DOTS PLUS therapy and doing well.

Keyword :ICL, MDR-TB, ATT, CD4 count.

Background:
Idiopathic CD4 lymphocytopenia (ICL) is a condition characterized by depletion of CD4 T-cells without evidence of human immunodeficiency virus (HIV) infection. ICL is defined as CD4 count <300 cells/mm^3 or a CD4 count that is <20% of the total T cell count on 2 occasions, with absence of HIV infection and absence of any defined immunodeficiency. To our best knowledge this is one of those rare cases in paediatric age group which warrants a mention, not only because of CD4 depletion and but also presence of extra pulmonary (cervical node) MDR TB in a 10 year old boy

Case History:
10 year old boy presented with complaints of multiple swelling with discharging sinus in the neck. Patient was on ATT since age of two and half years due to non resolution of swelling with sinus.Culture and sensitivity test for MTB done and was diagnosed as Extrapulmonary (cervical node) MDR-TB (resistant to INH, RIFAMPICIN, STREPTOMYCIN, ETHAMBUTOL, OFLOXACIN and sensitive to KANAMYCIN, ETHIONAMIDE).We analyzed the cause for Extrapulmonary MDR-TB in this child who is HIV negative. His CD4 found to be decreased. So he was further evaluated.

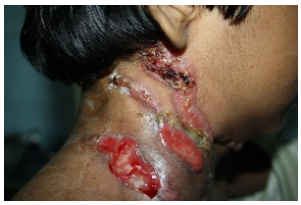

Fig 1 - Active discharging sinus, right lateral view

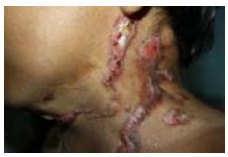

Fig 2 - Multiple scars with sinuses, left side of neck

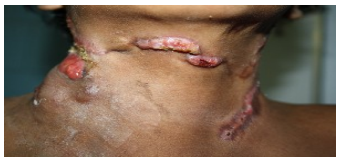

Fig 3 - Active swellings with sinus, frontal view

Methods:
We evaluated the patient clinically. All basic investigations and also CD4, CD8 flow cytometry, three rapid serological tests for HIV 1 and 2 were done.

Immunoglobulin profile, lymphocyte phenotype tests for CD3/ CD127/PD1/Regulatory T cells /Ki67/CD19/CD56(NK Cells), Nitro Blue Tetrazolium test were done to find the cause of CD4 depletion. Every attempt was made to rule out presence of any other infection by clinical examination and laboratory studies.

But the only drawback is HIV for ELISA has not been done since the three rapid serological tests for HIV 1 & 2 has been the practice in our hospital. Ideally ELISA for HIV should have been done.

Results:
In the patient there was no evidence of either HIV infection. CD4 counts done at 6 months met the CDC criteria of <300 cells/mm^3 or <20% on both the occasions. Reversal of CD4/CD8 ratio was present. The patient had normal levels of immunoglobulin. NBT ruled our Chronic Granulomatous disease.
Patient has no other opportunistic infection except for MDR-TB.

Discussion:
Idiopathic $CD4^+$ lymphocytopenia (ICL) is a rare non–HIV-related syndrome with unclear natural history and prognosis. It is a heterogeneous condition diagnosed typically in middle age, usually after an opportunistic infection, although it can also be an incidental laboratory finding.
The spectrum of opportunistic infections in ICL seems to overlap with that found in HIV-positive patients with similar CD4 T-cell counts. Cryptococcosis and nontuberculous Mycobacterial infections were the most frequent in ICL. In addition to opportunistic infections, autoimmune diseases were common.
In short, ICL is a heterogeneous yet distinctive condition that is quite different clinically and immunologically from infection with HIV.
In our case report, ICL was diagnosed in a 10 year old boy who presented with non-healing, multiple, bilateral discharging sinus in the neck. To our best knowledge, this is a rare presentation of ICL with Extrapulmonary MDR-TB and that too in a paediatric group.
The child after complete evaluation has been started on interferon therapy for ICL and DOTS PLUS therapy for MDR-TB and is doing well now

.**Conclusion**:
The important differentials that should be taken into consideration in case of CD4 lymphocytopenia are infections like HIV, EBV, CMV, Cryptococcus, Sarcoidosis, malignancy conditions like MALT lymphoma and Burkitt's lymphoma, myelodysplastic syndrome and autoimmune diseases like primary Sjogren's syndrome and systemic lupus Erythematosus and even chemotherapeutic drugs.
Idiopathic CD4 T-lymphocytopenia should be considered in all immunocompetent patients when there is no plausible explanation for recurrent infections in a patient and patients should be thoroughly evaluated and other causes of CD4 depletion should be ruled out.

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