

Title: T γ d Hepatoesplenic non-Hodgkin Lymphoma Case Report in Childhood

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Background: Hepatoesplenic lymphoma γ d T cell is a relative rare subtype of peripheral lymphoma in childhood. Clinical features include young age, male predominance, tumor restricted to liver, spleen and bone marrow, peripheral blood cytopenias, and generally without lymphadenopathy. The neoplastic cells are of T cell lineage, with CD3+, CD2+, CD7+, CD4-, CD8- immunophenotype. Natural killer cell associated antigens such as CD16 and CD56 are often expressed. It has an aggressive course and highly unfavorable prognosis.

Objective: Report a rare subtype of non Hodgkin lymphoma in childhood

Method: From June 1988 to January 2006, 235 patients (childhood and adolescents) with non-Hodgkin lymphoma were treated in our service and only one was hepatoesplenic lymphoma gamma delta. A 9 year old girl presenting with a huge splenomegaly which was found incidentally after an abdominal contusion six months ago. Also at presentation was found peripheral cytopenia (hemoglobin 8,8mg/dl and platelets 98.000) and pyrexia. The morphologic analysis of bone marrow aspiration showed 47% of neoplastic T cells and hemophagocytosis. Immunophenotypic features were positive for CD3, CD2, CD16 and CD45RO. The T cell lineage was confirmed by the immunohistochemistry of bone marrow biopsy (positive for CD45RO, LCA and negative for CD20, MPO, CD79A, TdT and CD34) and the clonal γ d T cell receptor (TCR) was also positive. Conventional cytogenetic analysis showed 48,XX,+3, add (7)(p22), +8[4]/46,xx[16]. Chemotherapy with cyclophosphamide, doxorubicin, vincristine and prednisone was started and repeated 2 times, however it was ineffective. Another chemotherapy scheme was tried (etoposide and cytarabine) but the patient persisted with splenomegaly and pancytopenia. Splenectomy was performed after 4 months from diagnosis.

Result: After surgery, adjuvant chemotherapy was initiated and the patient is currently alive, six months after diagnosis.

Conclusion: The conventional chemotherapy (CHOPP) it was ineffective for this child with T gamma delta non Hodgkin lymphoma.

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