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Clinical Image
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Hepatosplenic γδ T- Cell Lymphoma Presenting with Hemophagocytic Lymphohistiocytosis

*Hua Pan, Feng Hou, Yong-An Ni, Ke-Lei, Hui-Juan Xu, Gao-Yan Wang and Li-Rong Sun**

Department of Pediatric Therapy Center, Affiliated Hospital of Qingdao University, China

Clinical Image

A previously healthy 13-year-old girl presented with a two-week history of fever and a three-day history of peripheral blood cytopenias. She was found to be pale, hepatosplenomegaly in the absence of lymphadenopathy. A blood count showed a microcytic anemia (Hb 61 g/l, MCV 75 fl), thrombocytopenia (platelet count 57×10^9/l), and a white cell count of 11.1×10^9/l. Other laboratory tests showed fibrinogen 3.97 g/l, ferritin 3351 ng/ml, albumin 16 g/l, C-reactive protein 160 mg/dl. In bone marrow, 31% of cells represented an infiltrate consisting of medium-size lymphocytes with pale cytoplasm and irregularly shaped nuclei without prominent nucleoli (Figure 1A); Marrow flow cytometry showed an abnormal clonal population of CD4- and CD8-negative T cells which showed homogeneous coexpression of CD2, CD3, CD7 and TCR γδ, and partial expression of CD56, with neither cytoplasmic nor surface membrane expression of CD5, CD4, CD8, CD6, CD57 and HSTL. Fusion gene was negative. The peripheral blood karyotype was normal female, 46XX. The diagnosis is acute lymphoblastic leukemia (T cell type, intermediate risk). After one course of chemotherapy with CCCG-ALL2015 regimen, the bone marrow reached complete remission, and the liver and spleen could not be touched under the ribs.

Four months later, the patient developed recurrent fever, jaundice, hepatosplenomegaly and peripheral blood cytopenias. The ferritin was 23760 ng/ml. ALT 345U/L, AST217U/L, fib 0.8 g/L and glycerin 3.7 mmol/L. Bone marrow examination showed suppression of erythropoiesis and 27% atypical lymphoid cells and hemophagocytosis (Figure 1B). The patient was clinically diagnosed with HLH upon fulfillment of at least 6 of 8 diagnostic criteria. Spleen biopsy showed a diffusely infiltration of small to medium-sized atypical lymphocytes, the atypical lymphocytes are present within the cords and the sinuses of the red pulp. Histological examination showed CD3+, CD4-, CD8-, T Cell Receptor (TCR) αβ- and TCR γδ + abnormal lymphoid infiltrates (Figure 2).

To confirm the clonality and the configuration of TCR genes, southern blot and polymerase chain reaction studies have demonstrated monoclonal TCR γ, TCR δ and TCR β chain gene rearrangements using genomic DNA extracted from marrow of the patient. Together these findings were diagnostic of Hepatosplenic γδ T Cell Lymphoma (HTCL) and Hemophagocytic Lymphohistiocytosis (HLH). Combination chemotherapy consisting of cyclophosphamide, vincristine, bleomycin, etoposide, doxorubicin, and prednisolone (CyclOBEAP) was initiated. Considering the combination of hemophagocytic syndrome, the patient was treated with dexamethasone and etoposide. After receiving this CyclOBEAP therapy for 6 wk, improvement of hepatomegaly and complete remission were obtained. The body temperature was normal and the spleen retracted. After one month, fever recurred, the three lines decreased, liver and spleen progressively increased, and bone marrow

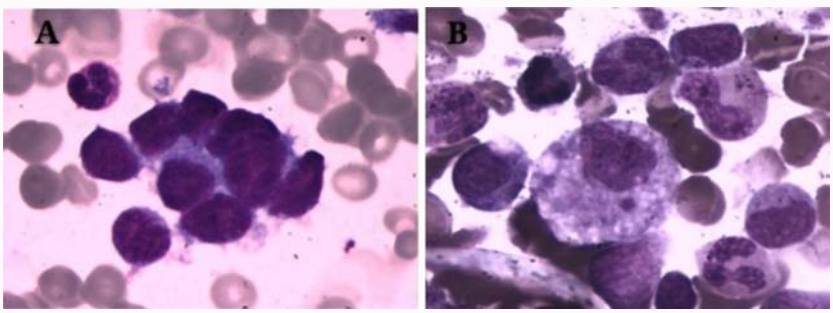

Figure 1: (A) Representative bone marrow smear at the diagnosis of acute lymphoblastic leukemia. (B) Bone marrow smear showed hemophagocytosis at the diagnosis of HLH.

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Correspondence:
Li-Rong Sun, Department of Pediatric Therapy Center, Affiliated Hospital of Qingdao University, 16 Jiangsu Road, Shinan 266003 Qing dao, China, E-mail: sunlr@vip.sina.com

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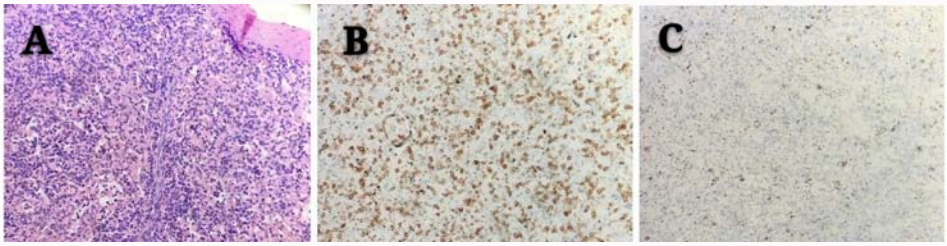

Figure 2: (A) Biopsy showing many medium-sized, atypical lymphoid cells within the dermis. These cells possess indistinct cytoplasm, round nuclei and a condensed chromatin pattern (hematoxylin and eosin). (B) Neoplastic lymphoid cells positive for CD3 (immunoperoxidase). (C) Neoplastic lymphoid cells negative for CD20 (immunoperoxidase).

recurred. The child died of infection.

HTCL is a rare, aggressive, extranodal T-cell Non-Hodgkin Lymphoma (NHL) that accounts for 3% of all T cell lymphoma subtypes in the United States, 2.3% in Europe, and 0.2% in Asia [1]. HTCL arises from a small subset of γδ TCR expressing lymphocytes. The lymphoma exhibits a sinusoidal pattern of infiltration in the spleen, liver, and bone marrow. It is characterized by an aggressive clinical course and poor response to conventional chemotherapy [2]. HLH is a life-threatening disorder that can occur in a familial primary form or be a response to infections, rheumatological disorders or neoplasms, particularly T-lineage lymphomas. It has firstly been reported in association with γδ T cell lymphoma in children. Early detection of HLH possibly helps to reduce morbidity by initiating prompt treatment of allogeneic bone marrow transplantation [3].

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