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Case Report

Oil Red O positive vacuolated blasts in a case of CD45 negative, CD19 negative B-lymphoblastic leukemia☆

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Abstract B-lymphoblastic leukemia is a clonal hematopoietic disorder of precursors B-lymphoblasts being most frequently encountered in children. Expression of CD19, a pan B-cell marker is noted in the majority of the cases with lack of CD19 expression being extremely rarely reported in the medical literature. We report the very rare case of a B-lymphoblastic leukemia with a triple negative immunophenotype: CD19 negative, CD45 negative and CD10 negative with vacuolated lymphoblasts and an aleukemic presentation in a 20-year-old man.

1. Introduction

A clonal hematopoietic stem cell disorder acute lymphoblastic leukemia affects overall 1.7 per 100,000 people annually and the percent of survival reaches 67.5% at 5 years. The large majority of acute lymphoblastic leukemias (ALLs) cases develop in children less than 5 years of age with a peak incidence of 4 to 5 cases per 100,000 people. [1,2] ALL represents about 15% of all malignancies in 1–15 year olds, 5% in 15–19 year olds, and <10% of malignancy in >20 year olds [3]. The large majority of these ALLs are of B-cell lineage and their correct diagnosis via immunophenotyping is essential for their treatment. Most of the B-lymphoblastic leukemia cases are CD19 and CD45 positive, co-express cytoplasmic CD79a, terminal deoxynucleotidyl transferase (TdT) and HLA-DR and are CD10 positive. To our knowledge, there has been only one other case of CD45 negative CD19 negative B lymphoblastic leukemia reported in the medical literature [4]; however, it lacks the morphological features of the current case.

2. Case report

A 20-year-old, Caucasian man with a 20-lb unintentional weight loss over the course of 2 months and recent heavy night sweats, presented with intermittent spasm–back pain with radiation to his chest and ongoing cough. The patient denied fever, fatigue, shortness of breath, or dyspnea.

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Complete blood cell counts on admission were remarkable for mild leukopenia (white blood cells 2.5 × 10^9/L), mild anemia (hemoglobin 12.7 g/dL), and mild thrombocytopenia (platelets 144 × 10^9/L). Rare blasts were identified on scanning of the peripheral blood smear, and they were of medium to large size and contained many intracytoplasmic fine vacuoles. An anterior mediastinal mass and a small right pleural effusion were noted on a chest x-ray and a computed tomography (CT) scan examination of the chest, abdomen and pelvis was remarkable for an 11.4 × 9.1 cm anterior mediastinal mass with mass effect (Fig. 1) on the superior vena cava and left mainstem bronchus. There was also subcarrinal, left hilar, right axillary, celiac, peripancreatic, periaortic and periureteral lymphadenopathy and ill-defined small rounded cortical lesions were noted in both kidneys. The patient underwent bilateral iliac crest bone marrow biopsies and aspirate examination with immunophenotypic analysis by both flow cytometry and immunohistochemistry, with associated molecular and cytogenetic/FISH analysis. Based on the findings a diagnosis of B lymphoblastic leukemia, with an unusual immunophenotype (CD10 negative, CD19 negative, CD45 negative) was rendered. The patient started induction chemotherapy on the DFCI 05-001 protocol. Multiple complications developed during his induction and oral chemotherapy treatment including *Staphylococcus* bacteremia, recurrent pancreatitis, and recurrent pneumonia and bronchiectasis. His oral chemotherapy maintenance course was also extended by nine months due to a period of non-compliance during his initial treatment. Two and a half years after the initial diagnosis of ALL, the patient remains in remission being currently on maintenance chemotherapy. His most recent CBC indices are within reference ranges.

3. Materials and methods

Immunohistochemistry, flow cytometry immunophenotypic analysis, karyotype analysis, in situ hybridization studies (FISH), and PCR studies were performed as for the clinical workup of this case. Immunohistochemical stains were performed on bone marrow biopsy cores using: CD3 (monoclonal PS1, Novocastra), PAX-5 (BV6, Diagnostic Biosystems), CD79a (monoclonal JCB117, Cell Marque), TdT (monoclonal SEN28, Vector), CD34 (monoclonal QBEnd/10, BioGenex), CD43 (monoclonal DF-Ti and MT1, DAKO and BioGenex coctail), myeloperoxidase (polyclonal, DAKO), CD68 (monoclonal KPI, Cell Marque), lysozyme (polyclonal, DAKO), and CD117 (Diagnostic Biosystems) using an automated immunostainer (DakoCytomation, Inc, Carpinteria, California). The 5-color flow cytometry studies were performed on bone marrow aspirate samples anticoagulated with EDTA, using Beckman Coulter IOTest monoclonal antibodies (Hialeah, Florida) analyzed on a Beckman Coulter FC500 flow cytometer using CXP software. The blast populations were Boolean gated based on specific antigen expression patterns, with a marker being considered positive when expressed in more than 20% of the blast forms above the isotypic control. The karyotype analysis, FISH and molecular studies were performed based on the classic recognized protocols.

4. Results

Patient's bone marrow examination was remarkable for a markedly hypercellular marrow (approx. 90%–100% on right

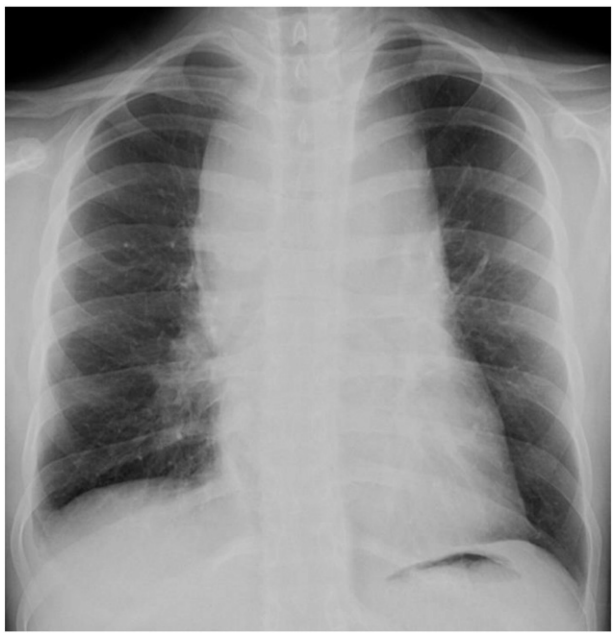

Fig. 1 CT image of the anterior mediastinal mass.