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CASE REPORT

Extramedullary Precursor B-Cell Lymphoblastic Leukemia: A Rare Presentation & Opportunity?

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ABSTRACT

Renal presentation of the B-cell acute lymphoblastic leukemia is extremely rare. We are presenting such a case.

B lymphoblastic leukemia/lymphoma (B-ALL/LBL) accounts for 2% of lymphoid neoplasms diagnosed in the United States. B-cell acute lymphoproliferative disease manifest as pure leukemia (B-ALL) in 80% of cases, isolated extramedullary disease (B-LBL) in 10% and mixed B-ALL/B-LBL in 10% of cases.

Case Report

B lymphoblastic leukemia/lymphoma (B-ALL/LBL) accounts for 2% of lymphoid neoplasms diagnosed in the United States. B-cell acute lymphoproliferative disease manifest as pure leukemia (B-ALL) in 80% of cases, isolated extramedullary disease (B-LBL) in 10% and mixed B-ALL/B-LBL in 10% of cases. Extramedullary presentations that do not have marrow involvement at diagnosis may present diagnostic challenges [1]. To the date, case reports have described extramedullary disease with pancreas and GI involvement only,in the setting of relapse in pediatric populations or post-allogenic stem cell transplantation in adults [2]. Herein we present a unique case of Philadelphia chromosome positive, B-cell ALL presenting with bilateral kidney, gastric and duodenal involvement.

A 49-year-old Caucasian woman was admitted with a one month history of headaches,ecchymosis andpancytopenia. Initial bone marrow biopsy was normo-cellular without any abnormal findings. Computed tomography of theabdomen revealed bilateralheterogeneously enhancing kidneys. She was conservatively treated withGranulocyte-colony stimulating factor(G-CSF) and erythropoietin, her counts normalized in 4-5 days with significant associated clinical improvement. She was discharged after core needle biopsy of renal mass was done. Results were pending at that time.

The patient was re-admitted after final pathologycame out to be consistent with “aggressive precursor B-cell lymphoblastic leukemia, ALL, L1.”Pathology was performed with Immunohistochemistry (IHC) and Fluorescence in situ hybridization (FISH). BCR-ABL fusion gene overexpressed on polymerase chain reaction (PCR). Markers were checked that included CD 10 – 19 -22 -38, HL-DR 1. Blastswere positive for Terminal deoxynucleotidyl transferase(TdT).

The patient also underwent Esophagogastroduodenoscopy which showed involvement of duodenal and gastric antral mucosa by B-ALL. Repeat bone marrow biopsy was consistent with B-ALL, L1 and Philadelphia chromosome (t9; 22) was positive. BCR-ABL fusion gene transcript was observed (p190) with a sensitivity of <0.001 %. The time period between the first and repeated bone marrow sample was 21 days. Repeated bone marrow exam showed 84% blasts. Lumbar puncture followed the bone marrow biopsyand was remarkable for 5% blasts presence. There were 17 white blood cells present in cerebrospinal fluid and 90% of them were lymphocytes. Protein was 20 mg/dl and Glucose within the normal range.

Therefore, ALL-negative bone marrow with pancytopenia was present and we doubt it was a sampling error. Unilateral bone marrow was done and patchy disease might have been missed by puncture. We double checked to make sure no samples were mixed up and that the samples were in fact from same patient. Blood was also evaluated with morphology and flow cytometry. In peripheral blood there were 21% blasts pr esent.

The patient subsequently underwent induction chemotherapy with hyper-CVAD (Cyclophosphamide, Vincristine, Adriamycin and Dexamethasone)along with intrathecal methotrexate and Ara-C. Given her Ph-chromosome positive status, she was also started onlmatinib. The patient completed four cycles of the above regimen. After the second cycle she attained minimal residual disease on repeat bone marrow biopsyand BCR/ABL1 levels below detectable level on PCR.She is currently being evaluated for bone marrow transplantation.

Extramedullary ALL is rare and has classically been described in the setting of relapsed disease in pediatric population. Kebaili et al described a 13 years old female with left solid renal mass discovered on routine ultrasound imaging 32 months after complete remission [3]. Similarly, De Alivia and Menell described a 16 years old male presenting with painless hematuria and solitary left exophytic renal mass 36 months after completing chemotherapy for ALL [4]. In both cases, renal biopsy confirmed lymphoblastic infiltration while bone marrow biopsy/aspirate were normal. While we are only beginning to understand the complex nature of tumor microenvironment, it is believed the chemokine stromal-cell-derived factor-1(SDF-1) plays an important role in leukocyte trafficking and is constituently expressed in both bone marrow and stromal cells. Overexpression of SDF-1's receptor, CXCR4, on malignant lymphoblasts is associated with extramedullary infiltration in patients with acute leukemia, irrespective of peripheral blast counts [5]. We can postulate that high definition genomic analysis of this patient's DNA may reveal overexpression of CXCR4. Further understanding of the complexities of the tumor microenvironment may yield exciting venues of targeted therapy in the future.

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