Sudan Black B Positive Acute Lymphocytic Leukemia

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Introduction

Acute lymphocytic leukemia (ALL) is the most common malignant disease affecting children, accounting for 30% of childhood cancers [1]. The correct diagnosis by morphology, cytochemistry, biochemistry, immunology and cytogenetics is important as approximately 70-80% of children with ALL survive for more than 5 years without disease recurrence [2,3].

The criteria by the French-American-British group for the classification of acute leukaemia is based on bone marrow morphology and cytochemistry [4]. Greater than 3% myeloperoxidase (MPO) or Sudan black B (SBB) positive blasts have been the cytochemical criteria for the diagnosis of acute myelogenous leukaemia (AML) [4]. Though myeloperoxidase is a highly specific cytochemical marker for myeloblasts [5], SBB may be more sensitive in some leukaemic blasts that are myeloperoxidase negative and may show weak SBB staining. SBB is very useful in distinguishing AML from ALL, though rare cases of SBB positive ALL have been reported [6,7].

We present a rare case report of ALL, which had blast positive for SBB and negative for myeloid associated chemistries including myeloperoxidase.

Case Report

An 11 year old patient presented with history of high grade and intermittent fever, along with lymphadenopathy for the past one month. He also had mild epistaxis. History of bleeding from any other site was absent. He had a history of receiving two units of blood transfusion.

On examination patient was febrile, had pallor and multiple bilateral cervical (both anterior and posterior), axillary and inguinal lymphadenopathy. He had bilateral parotid gland enlargement. Liver was 3.5 cm and spleen was 6 cm palpable, firm and non-tender. FNAC of lymph nodes revealed replacement with lymphoid cells. Peripheral smear showed normal leukocyte count ($10 \times 10^3/\mu\text{l}$) with blasts of 75%, which morphologically were lymphoblasts. Platelet count was $50 \times 10^3/\mu\text{l}$. Bone marrow was cellular and showed complete

replacement by blasts. Cytochemistry revealed blasts to be negative for MPO, non-specific esterase, acid phosphatase but positive for SBB. Immunophenotyping showed blasts were positive for CD19, CD22, CD10, CD34, CD45, Cy Ig M and negative for CD3, CD7, CD14 & CD33 by flow cytometry. A diagnosis of pre-B cell ALL (CALLA positive) was made. The patient was put on ALL chemotherapy to which the patient responded.

Discussion

A positive SBB has been considered to be diagnostic of acute non lymphoblastic leukaemia when present in greater than 3% blasts [4]. However, SBB positivity has been reported in 1.6% of cases of patients with otherwise typical ALL [7]. The diagnosis of ALL in our patient was supported by morphology, the absence of myeloperoxidase, the presence of immunological markers of ALL, and the absence of reactivity with myeloid monoclonal antibodies.

This case raises some important considerations. First, it would appear that SBB can no longer be considered entirely specific for acute non-lymphocytic leukaemia, and even MPO positivity has been described in few cases of ALL [8]. It is therefore advisable to perform MPO in conjunction with SBB, since either one of them can be positive in ALL and not both. The reason for SBB positivity in blasts of a small percentage of patients with ALL is not clear. One possible explanation is that SBB stains a variety of lipids (neutral fat, phospholipids, sterol) in addition to primary granules and the positivity may be the result of altered lipid metabolism in leukaemic cells [7].

Second, immunological markers were useful in supporting the diagnosis of ALL. It showed common ALL antigen positive and were negative for myeloid antibodies. It is advisable to support the diagnosis of ALL in all cases with immunological markers.

Thirdly, we considered the possibility that this case represented acute leukaemia with mixed lymphoid and myeloid phenotype, and as such represents an infidelity of lineage expression. This possibility appeared unlikely

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since SBB was the only positive marker typically seen in AML and all other markers including immunological markers were those usually associated with ALL. However, additional studies at the ultrastructural level for myeloperoxidase and at the genetic level for immunoglobulin gene activity are required to rule out the possibility of a 'masked' mixed leukaemia.

In conclusion, it is stated that SBB reactivity may be seen in the blasts of some patients with otherwise typical ALL. This observation is important since these cases should not be diagnosed as acute non-lymphocytic leukaemia and may have a favourable response to therapy for acute lymphocytic leukaemia.

References

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