Priapism in Childhood B-Cell Acute Lymphoblastic Leukemia: A Rare Presentation

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Received: 25 August 2019 / Accepted: 5 September 2019 / Published online: 25 September 2019
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Dear Editor,

Priapism refers to prolonged and persistent erection of penis without any sexual stimulation. Priapism is rare in children, with sickle cell disease (SCD) being the most common cause in this age group [1]. It can also be seen in association with hyperleukocytosis and has been reported in 1–5% of adult patients with leukemia. Priapism is however unusual in childhood leukemias [2]. We report a case of 12-years old child who presented with priapism as the chief complaint and was eventually diagnosed as B-Acute lymphoblastic leukemia (B-ALL).

A 12-years male patient presented with progressive pallor for 15–20 days, headache, fever and dizziness for 10–15 days and priapism for two days. There was no history of bone pains, bleeding episode or blood transfusion in the past. On general physical examination, the patient was conscious, oriented and pale. He had mild hepatomegaly and no splenomegaly. Systemic examination was unremarkable. Penis was swollen and tender to touch. Based on the clinical history of fever, priapism and mild hepatomegaly, an infective cause or SCD were considered as differentials.

Hemogram findings revealed bicytopenia with marked leucocytosis (haemoglobin of 6.2 g/dL, TLC of $501 \times 10^9/L$, platelet count of $76 \times 10^9/L$ and ESR of 90 mm/h). On

peripheral smear examination there were 95% blasts which were 1.5–2 times the size of a small lymphocyte, with scant cytoplasm and inconspicuous nucleoli, few cells showed convolutions and nuclear indentations. Bone marrow aspirate smears were hypercellular for age with diffuse replacement by blasts (Fig. 1a). Cytochemistry stains (MPO, SBB & PAS) were negative. Immunophenotyping on peripheral blood showed CD19, CD20, HLA-DR, CD34 and CD79a positivity on CD45-side-scatter (Fig. 1b, c). CD10, TdT, CD38, CD56, CD1a, CD117, CD3, CD5, CD7, cMPO, cCD3, CD13, CD33 and CD64 were negative. Immunophenotype findings were consistent with B-ALL.

Priapism is uncommon in pediatric population. Most common cause of priapism in pediatric cohort is SCD, others being trauma, leukemias, idiopathic, drug-induced and miscellaneous causes [1]. It is seldom seen in childhood leukemias. Amongst these too, B-ALL presenting as priapism is very rare [2]. The literature search based on mesh terms ‘priapism’, ‘B-cell leukemia’ and ‘hyperleukocytosis’ revealed only three case reports, immunophenotyping was however not done in these cases to establish the B or T-lineage [3]. Chronic myeloid leukemia is the most common hyperleucocytic leukemia presenting with priapism and most of the cases are adults [4].

Hyperleukocytosis (TLC $> 100 \times 10^9/L$) is an uncommon presentation in acute leukemias, seen in 10–18% cases of pediatric ALL [5]. Even in ALLs, majority of hyperleucocytic ALL are T-ALL [3]. Factors commonly associated with hyperleukocytosis include age < 1 year, male gender, T-cell type, massive hepatosplenomegaly, mediastinal mass, CNS involvement and cytogenetic abnormalities like hypodiploidy, 11q23 rearrangements, Philadelphia positive ALL and loss of p16 expression [5]. The index case was a male but did not have any other associations. Hyperleukocytosis is associated with a poor

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Fig. 1 a–c Bone marrow aspirate smear showing complete replacement by blasts (1000 $\times$). Flow cytometry plots showing (b) 85.6% blasts in the CD45-side scatter and c CD19 and CD20 positivity in the gated population

overall survival and because of early death of the patient, molecular tests could not be performed.

Our case is unique as ALL presenting with priapism and hyperleukocytosis is very rare, more so in B-ALL. It is thus important to ask for hemogram and examine the peripheral smear in all cases of priapism. This is of importance even in a young patient in order to rule out B-ALL which has a different treatment protocol from T-ALLs.

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Informed Consent For this type of study, formal consent is not required.

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