NP 07

ACUTE LYMPHOBLASTIC LEUKEMIA IN A PEDIATRIC PATIENT WITH HEMOPHILIA B: A RARE CLINICAL CHALLENGE

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New Products

Abstract

Background: There are no reported cases of acute lymphoblastic leukemia(ALL) in patients with hemophilia B. There is one case report of a young adult with hemophilia B and acute myeloid leukemia(1). Currently, there is no best practice recommendation for the management of patients with hemophilia B and ALL. Objectives: To report our experience in managing a pediatric patient with congenital hemophilia B and ALL, which presents a rare and unique clinical challenge. Design/Method: Retrospective chart review Results: This is a 2y/o male with hemophilia B diagnosed at birth from a cord blood sample showing a factor IX level <1%. Mother is a known carrier and maternal grandfather has severe hemophilia B. Patient started prophylaxis with a standard half-life product through central venous access around 8months of age, following a spontaneous wrist bleed. The schedule was 35 units/kg twice a week. He had no spontaneous joint or soft tissue bleed on this regimen. 3mo ago he presented with pain and swelling of the right wrist. He fell on an outstretched hand the day before and received 100% factor infusion. X-ray showed metaphyseal lucencies with overlying soft tissue swelling. No evidence of fracture. Due to this finding, additional labs were done. WBC 4.8K, hemoglobin 7.1 g/dL, platelets 18K and 31% blasts. Flow cytometry confirmed the diagnosis of preB-ALL. On exam, he had pallor, scattered petechiae and cervical lymphadenopathy. Based on recovery studies and thrombocytopenia, the prophylaxis was changed to 50 units/kg every third day. The platelets are kept above 30K at baseline. For lumbar punctures, he has been corrected to 100% factor level and platelets kept above 50K. However, due to the risk of port infection with frequent accessing, he was switched to long-acting albumin fusion factor IX product on day 22 of induction. The current prophylaxis regimen is 75units/kg weekly and the schedule is adjusted to coincide with lumbar puncture days whenever needed. He has tolerated all his procedures well without increased bleeding, including end of induction bone marrow aspiration, biopsy and lumbar puncture with intrathecal chemotherapy. He is currently in remission and is in interim maintenance phase of treatment per COG protocol AALL0932. Conclusion: Long-acting factor IX products could potentially decrease the number of infusions and need for frequent central venous access in immunocompromised patients with hemophilia B. In addition, a higher trough level with a weekly schedule could provide better bleed control in patients with severe thrombocytopenia due to underlying malignancy. A baseline platelet count of at least 30K is recommended during treatment. More treatment guidelines need to be established.(1) Clark C et al, Pediatric Blood and Cancer Jan 2011