Acute Unilateral Hypopyon Uveitis in A Child with Post-BMT local Relapse of Acute Lymphoblastic Leukemia-Case Report

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ABSTRACT

Relapse into the anterior ocular segment post bone marrow transplantation (BMT) for acute lymphoblastic leukemia (ALL) in a child is rare. We report this uncommon presentation as hypopyon uveitis in a patient in remission with complete chimerism after chemotherapy for ALL. The patient presented with red eye and slit lamp examination revealed hypopyon uveitis. The diagnosis of relapsed ALL was established by anterior segment aspiration followed by documentation of relapse with immunohistochemistry and flow cytometry of the fluid sample. Thus, a high index of clinical suspicion in uveitis cases post BMT, led to the early diagnosis of ocular relapse that prevented vision loss in the patient. Atypical unilateral hypopyon can therefore be an indication of relapsing ALL.

Keywords: Acute Lymphoblastic Leukemia, ALL, Ocular Relapse, Hypopyon Uveitis

Case Report

Anterior segment infiltration in acute lymphoblastic leukemia (ALL) presenting as hypopyon uveitis is rare. We present the case of a 5-year-old female, who underwent bone marrow transplantation for ALL in March 2017. ALL was diagnosed by immunohistochemistry as well as flowcytometry (CD19+, CD10+, CD34+, TdT+ but negative for CD3, CD13, CD33, CD117 and cytoMPO). Karyotyping showed normal chromosomes. Molecular studies revealed BCR/ABL 190 transcript positive. The child received intensive chemotherapy (Berlin-Frankfurt-Munster regimen) with remission induction protocol (Chang et al., 2008) and given the poor prognostic features she received an allogeneic haemopoietic cell transplantation from an HLA-identical donor after achieving remission following a myeloablative regimen.
with Busulphan and Cyclophosphamide (BuCy) (Tutschka et al., 1987). The donor was her sister and her follow up in August 2017 revealed complete chimerism, with gut GvHD being treated with tacrolimus and tapering steroids. In December 2017, she presented with a 4-day history of red eye and ophthalmic assessment revealed right eye hypopyon with anterior uveitis (Fig. 1). There was no history of trauma. The complete blood counts and the peripheral blood film were unremarkable. Ocular examination along with an anterior chamber tap revealed pleomorphic lymphoblast suggestive of ocular relapse of ALL (Fig. 2). Flowcytometry of the anterior chamber fluid aspirate revealed that 90% of total nucleated cells were positive for CD19, CD10, CD34 but were negative for CD3 (Fig. 3). Cerebrospinal fluid and bone marrow samples were negative for blast cells and imaging studies did not show any other site of relapse. She was initially started with local antibiotic and intraconal steroids drops with good recovery of the uveitis. She was subsequently treated with hyperCVAD regimen along with the addition of imatinib mesylate (Biondi et al., 2012; Jabbour et al., 2018). The patient has been in remission at the last follow-up few months ago. This report thus is special for us as we have seen this for the first time since it exhibited isolated local ocular relapse without involvement of any other site and first time flow cytometry analysis was performed in such specimen.

Figure 1: Slit-lamp photograph showing white-colored hypopyon.

Figure 2: Blast cells are small to medium in size, high nuclear to cytoplasmic ratio, round nucleus, some had irregular nucleus outline, open chromatin and 1-4 nucleoli.
Ocular involvement is a well-documented complication of leukemia (Yassin et al., 2022). Extramedullary infiltration is common at the initial diagnosis or recurrence of childhood acute lymphoblastic leukemia, but it is rare for ocular findings to be the initial manifestation of a new or relapsed disease (Schachat et al., 1989). Furthermore, although ALL is the commonest leukemia in children, anterior segment infiltration is uncommon, and accounts for about 0.5-2.5% of relapse cases (Yassin et al., 2022). Nevertheless, ALL in children with eye involvement as hypopyon has been sparingly described with only a handful case reports (Lakhtakia et al., 2008; Wadhwa et al., 2007; Mendonca et al., 2021). However, extramedullary infiltration found less than 18 months after bone marrow transplantation, indicates a high risk. This was mitigated with the addition of oral tyrosine kinase inhibitors (Imatinib mesylate) in our patients and the patient is still currently disease free and relapse free at the last follow-up a few months ago.
Ocular involvement has been reported to occur either by direct infiltration of neoplastic cells, or hemorrhage, or ischemic changes (Ayliffe et al., 1995). Further, direct infiltration through posterior ciliary vessels could also be a probable cause (Decke and Burnstine, 1993). Importantly, relapses generally occur in certain sanctuary areas where the indolent blast cells are isolated from systemic chemotherapy or radiotherapy such as the testis in boys, or the brain and the spinal cord in the central nervous system. However, in our literature survey the anterior chamber of the eye has not been recognized sanctuary site.

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References


