Acute Bilateral Hypopyon in Acute Lymphocytic Leukaemia

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Introduction

Acute lymphocytic leukaemia (ALL) is a malignant clonal disorder of the bone marrow lymphopoietic precursor cells, characterized by accumulation of lymphoblasts that lack the potential for differentiation and maturation [1]. These abnormal cells spill into the circulation and infiltrate other organs such as the liver, spleen, central nervous system (CNS) and the eye. Presence of intraocular leukaemic infiltrates correlates with CNS involvement and with decreased survival [2,3]. Ocular manifestation may be the only sign of leukemic relapse or may present several months prior to systemic relapse. Hence, prompt anterior chamber aspiration is required for the correct diagnosis and treatment.

Case Report

A six month old male child, detected to have infantile ALL, was managed with induction chemotherapy and had achieved remission. At one year of age, the mother noticed that the child was not fixing its gaze on familiar faces or objects. On ocular examination, poor pursuit of source of light was noticed. Bilateral hypopyon with a horizontal fluid level was observed, measuring 2-3 mm from inferior angles of anterior chambers in the upright position, shifting its position readily with passive changes of the head (Fig.1). Minimal flare could be seen in both eyes and few cells could be seen in the aqueous above the hypopyon. Both pupils were miotic and responded poorly to bright light. The child was initially treated with topical steroids. The hypopyon regressed but recurred again. This time an anterior chamber tap from the inferior angles was carried out under general anaesthesia.

Microscopic examination of the aspirated aqueous revealed immature lymphoid cells with scant cytoplasm and large nuclei with 1-2 inconspicuous nucleoli (similar in morphology to the blasts on initial bone marrow evaluation) (Fig. 2). A microscopic examination of CSF at this time also

revealed similar cells. A diagnosis of leukemic spread to anterior chamber of the eye and CNS was made. The child was managed with local External Beam Radiotherapy to eyes and chemotherapy with FLAG and Triple IT. The condition responded by resolution of anterior chamber hypopyon, return of good visual acuity (smiling on looking at familiar faces and reaching out to toys etc.) and clearing of malignant cells from CSF. The child has been in remission for one year since diagnosis of disease.

Discussion

ALL is the most common malignancy in children, especially affecting those aged 2-10 years [1]. Ocular involvement may be present at the initial diagnosis but only 10% patients may be symptomatic [3]. This underscores the importance of ocular examination as a part of initial evaluation.

Involvement of the anterior segment is rare and is found in 0.5-2.5% of all leukemic relapses. Symptoms can include redness, photophobia, changes in pupil shape or color, pain, and visual loss. Clinically, anterior segment leukemic infiltrates can be distinguished from anterior uveitis, in that leukemic involvement presents more commonly with low intraocular pressures, normal or miotic pupils, segmental or total ciliary injection, segmental or diffuse iris swelling and posterior synechiae. Hypopyon and hyphema, are less common [2].

The ocular manifestations may be secondary to direct infiltration of the leukemic cells, as a result of the abnormal systemic haematological parameters or opportunistic infections. The presence of intraocular leukemic infiltrates correlates with CNS involvement and with decreased survival [4, 5]. Autopsy series show the highest frequency of ocular involvement [2].

Anterior segment manifestations in leukemia are

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Fig. 1 : Bilateral hypopyon with a horizontal fluid level, measuring 2-3 mm from inferior angles of anterior chambers in the upright position.

significant because they herald an extramedullary relapse [6]. It occurs more often in ALL than other types of leukemia. Leukemic hypopyon refers to the presence of leukemic cells layered in the anterior chamber of the eye [7]. The differential diagnoses of leukemic hypopyon include infectious and inflammatory causes. Cytologic examination is the only way to make a definitive diagnosis. The leukemic blasts have the same characteristics as those seen in the bone marrow and other fluids like CSF. Conventional morphology is routinely used for diagnosis in these fluids, but can be supplemented by immunophenotypic characterization if required.

Topical therapy alone is not effective and treatment options include chemotherapy (systemic and intrathecal) and irradiation of the anterior chamber. The prognosis of this complication depends on whether it occurs during therapy or after therapy has been discontinued. Aggressive retreatment appears warranted in cases of anterior chamber relapse of ALL, as some of these children may attain prolonged new remissions [8]. Our patient was treated promptly and has shown satisfactory progress to date.

Conflicts of Interest

None identified

Fig. 2 : Smear of the aspirated hypopyon showing high cellularity due to presence of leukaemic blasts (MGG x40). Inset: Lymphoblast with scant cytoplasm, large nucleus and inconspicuous nucleoli.

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