Masquerade uveitis with hypopyon as a solitary feature of relapsed leukaemia in a child

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Accepted 6 May 2021

DESCRIPTION

A 5-year-old boy presented with redness and watering in his left eye for 3 days. The child was on treatment for pre B-cell acute lymphoblastic leukaemia (ALL). At the time of diagnosis of ALL, he had presented to the hospital with fever for 2 weeks' duration and found to have hepatosplenomegaly with thrombocytopenia. Bone marrow studies and Cerebrospinal fluid (CSF) analysis confirmed the diagnosis of ALL with central nervous system (CNS) involvement and he received two consecutive weekly intrathecal methotrexate doses. He was started on treatment as per regimen A (three drug induction) of UKALL 2003 protocol.¹ At the end of induction, bone marrow analysis was consistent with disease remission with minimal residual disease <0.01% suggestive of standard risk. At the time of presentation to us, the child had just completed delayed intensification phase and started on first maintenance cycle of treatment. On ophthalmologic evaluation, the best-corrected visual acuity was 20/20 in the right eye and 20/200 in the left eye. Anterior segment examination revealed circum-corneal congestion in the left eye, anterior chamber cells 4+ with white hypopyon measuring around 2 mm in height (figure 1).² The pupil was meiotic, pupillary reaction was sluggish. Lenticular opacity was present in the posterior subcapsular region. Intraocular pressure was raised to 28 mmHg. Ultrasound B-scan showed choroidal thickening and exudative retinal detachment. CSF analysis and bone marrow aspiration were negative for leukaemic cells. Anterior chamber paracentesis revealed atypical lymphoid cells suggestive of leukaemic infiltration (figure 2). Anterior uveitis with hypopyon in the left eye was the sole clinical feature suggestive of relapse of ALL in this child. Reinduction treatment as per ALL R3 protocol was started following which, ocular and cranial radiotherapy, as well as subsequent bone marrow transplant, was planned. Unfortunately, he succumbed to his illness during the reinduction phase within a couple of months after relapse was diagnosed.

ALL is a haematologic malignancy originating from haematopoietic cells of B-cell or T-cell lineage. Although it can affect children and adults, it predominantly affects children between the age of 1 and 4 years of age.³ Advances in modern diagnostic and treatment protocols have achieved nearly 80%–85% cure rate in children with ALL.⁴ Unfortunately, relapse occurs in 15%–20% of the cases.⁵ The most common sites for relapse are bone marrow, CNS and testis.⁴ Ocular relapse is

Figure 1 A clinical photograph (A) and slit-lamp image (B) of the left eye showing white hypopyon (black arrow) measuring around 2 mm in height, the lateral border of which extends up to upper two-thirds of the temporal limbus.

rare, accounting for 0.5%–2.5% of the cases of leukaemic relapse.^{6–8} Ocular relapse can occur in isolation or along with relapse in bone marrow or CNS.⁴ Ophthalmic manifestations in patients with ALL can be a presenting sign of the malignancy or may indicate relapse of the disease.⁷ Iris and anterior chamber involvement is more often seen in ALL than chronic lymphoid leukaemia or myeloid leukaemia.⁹

Ophthalmic manifestations of leukaemia can occur due to three reasons. (1) Direct infiltration of ocular tissues such as iris, choroidal and optic nerve. (2) Haematological abnormalities causing leukaemic and anaemic retinopathy. (3) Neuro-ophthalmic signs such as papilloedema due to CNS

Figure 2 Anterior chamber aspirate cytology—high power view showing monomorphic population of lymphoblasts with high nuclear: cytoplasmic ratio, condensed coarse chromatin and scant cytoplasm (×400, Pap stain).

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To cite: Mendonca TM, Lashkari HP, Kini J, et al. *BMJ Case Rep* 2021;**14**:e240485. doi:10.1136/bcr-2020-240485

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involvement and raised intracranial tension.^{10 11} The presenting symptoms of anterior segment involvement could be redness, ocular pain, photophobia, watering and diminished vision.⁸ Occurrence of ocular inflammation in a patient with a history of leukaemia should raise suspicion of intraocular infiltration or relapse of leukaemia. Clinical signs of intraocular relapse or infiltration include, signs of anterior uveitis/iritis with or without hypopyon, iris nodules or thickening, pupillary abnormalities, secondary glaucoma, retinal/choroidal infiltrates and serous retinal detachments.⁸ These patients may be wrongly diagnosed to have 'atypical conjunctivitis' or 'uveitis' and more often treated with antibiotic and steroids. High index of clinical suspicion is necessary in such cases as the prognosis of ocular relapse in leukaemia is guarded and it requires aggressive local and systemic therapy.

The role of eye care professional is very important in diagnosis of masquerade uveitis in acute leukaemia. Ocular manifestations in leukaemia could be the first sign of the diagnosis or relapse of the disease following treatment. Since the management is challenging, and prognosis is poor after an ocular relapse, high index of suspicion and timely diagnosis is the key for a better outcome.

Learning points

- Iris and anterior segment involvement is more common with acute lymphoblastic leukaemia (ALL) than myeloid leukaemias.
- Ophthalmic manifestations in patients with ALL can be presenting sign of the malignancy or may indicate relapse of the disease.

Contributors TMM: Manuscript writing, concepts. HPL: Clinical management, manuscript editing. JK: Additional resources (pathology). TV: Manuscript editing.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Disclaimer Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

Competing interests None declared.

Patient consent for publication Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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