Case Report

Relapse of Acute Lymphoblastic Leukaemia masquerading as Hypopyon uveitis and disc edema

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Abstract

Masquerade syndromes represent a rare but important proportion of uveitis cases. This report illustrates a case of B-cell type of acute lymphoblastic leukemia during maintenance therapy in a 11-year- girl who relapsed after 2 years in the form of with hypopyon uveitis and disc edema in left eye. Her ocular problem was managed successfully with topical and oral corticosteroid.

Keywords: Disc Edema, Hypopyon, Leukemia, Uveitis

Introduction

Ocular masquerade syndrome (OMS) is a group of diseases that occurs with ocular inflammation and is misdiagnosed as a uveitis.[1] The causes of OMS may be benign or malignant disorders.[1] Malignant diseases include: intraocular lymphomas (non-Hodgkin's lymphoma of central nervous system, systemic non-Hodgkin's lymphoma metastatic to the eye, Hodgkin's lymphoma, leukemia, carcinoma metastastic to the eye (lung, renal, breast) uveal melanoma, childhood malignancies (retinoblastoma, leukemia, meduloepithelioma, juvenile xanthogranuloma), paraneoplastic syndromes (cancer-associated retinopathy, melanoma-associated retinopathy, bilateral diffuse uveal melanocytic proliferation).[2]

The anterior segment is an uncommon site of extramedullary relapse, accounting for between 0.5% and 2.6% of all relapses in large published series of acute lymphoblastic leukaemia (ALL).[3] Anterior segment infiltration and presence of lymphoblasts in the anterior chamber without any other CNS involvement in ALL is very rare but herein we report a case where the relapse of ALL presented as hypopyon uveitis in one eye and disc edema in the other eye.

Case presentation

A-11-old-girl had presented with low grade, intermittent fever for 8 months and progressive pallor for last 2 months. On examination, child was pale with generalised lymphadenopathy and hepatosplenomegaly. Peripheral smear revealed predominant lymphoblasts; bone marrow aspiration showed 69% of lymphoblasts suggestive of ALL-L2. Cytochemistry showed Periodic Acid Schiff stain and Myeloperoxidase stain negative suggestive of B-cell type of ALL according to French-American-British Classification of ALL. Child was started on induction followed by maintenance therapy as per MRC-2003 protocol.

After 2 years of maintenance therapy, she developed pain, redness, photophobia with diminuation of vision in left eye. On examination, vision was 20/20 and 20/60. Left eye showed conjunctival and ciliary congestion with stromal edema with fresh KPs in cornea. The anterior chamber showed 4+ cells with fibrin strands and hypopyon of 2mm height but no posterior synechiae (Figure 1). The iris was brown with irregular segmental swelling. Right eye anterior segment was unremarkable. Fundus examination revealed edematous disc in right eye (Figure 2) but normal in left eye. Applanation intraocular pressure was 14 mmHg in right but raised to 37 mmHg in left eye. The goldmann visual field test of right eye showed enlarged blind spot.

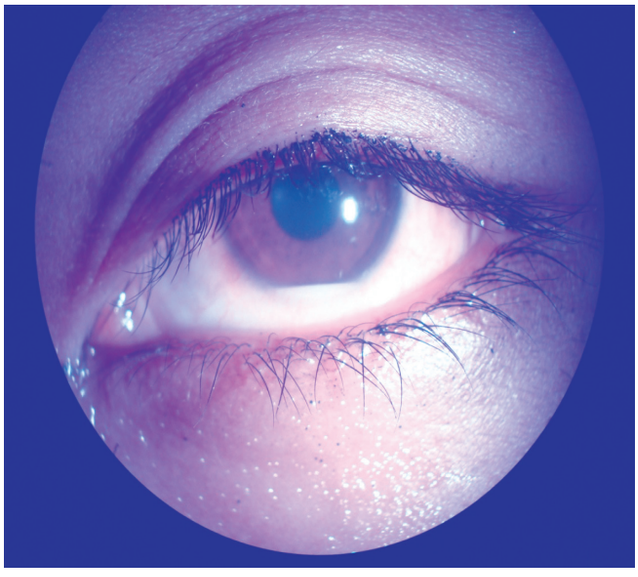

Figure 1 Clinical photograph of left eye shows hypopyon in the anterior chamber.

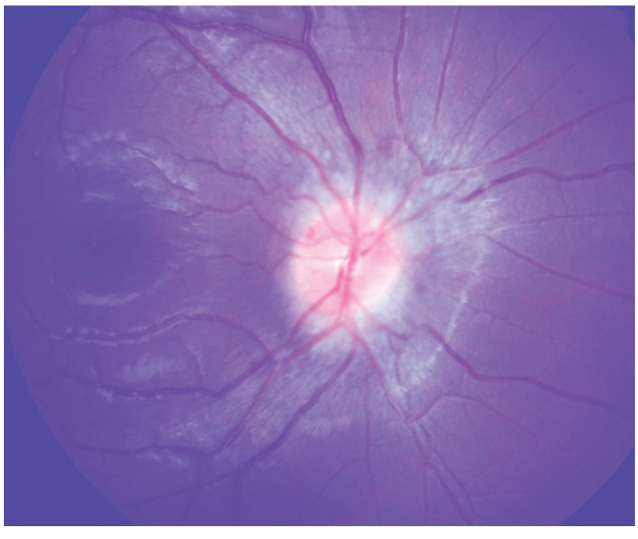

Figure 2 Fundus photography of right eye shows edematous disc at presentation

A clinical diagnosis of right eye disc edema and left eye acute intraocular inflammation with pseudohypopyon with secondary glaucoma was made. She was started on topical steroid (prednisolone acetate 1%), topical cycloplegic (atropine 1%) and topical glaucoma medication (Timolol maleate 0.5%) in left eye. On follow-up, IOP had reduced and the pain disappeared but there was no remarkable changes in the anterior chamber cells and hypopyon. Interestingly she developed cells in right eye also. She was consulted with her oncologist and low dose oral steroid (0.75mg/kg/day) was started along with her topical medications in both eyes. The vision improved to 20/20 bilaterally, the cells in both eyes reduced, pseudohypopyon of left eye disappeared. The disc edema of right eye also gradually started resolving (Figure 3) and intraocular pressure was maintained. Though her eyes improved, she expired 3 months later due to CNS relapse.

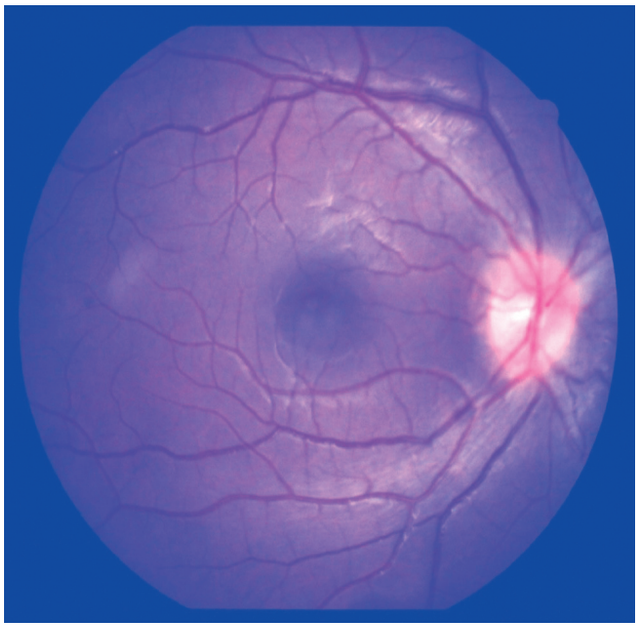

Figure 3 Fundus photography of right eye shows resolving disc edema after treatment.

Discussion

Masquerade syndromes are classically defined as those conditions that include, as part of their clinical findings, the presence of intraocular cells but are not due to immune-mediated uveitis entities.[4] Ocular masquerade syndrome is found in 5% of all of the uveitis cases.[5] The occurrence of uveitis in a diagnosed case of leukemia is very likely to be a sign of CNS relapse [6,7] however anterior segment may remain a sanctuary site for leukaemic cells during treatment accounting for between 0.5% and 2.6% of the ALL relapses.[8] Ocular involvement may occur by direct infiltration of neoplastic cells, hemorrhage, or by ischemic changes and sometimes can be the presenting feature of the disease de novo or an initial feature of a relapse.[9]

The absence of posterior synechia and increased intraocular pressure differentiates anterior segment leukemic infiltrates from other causes of anterior uveitis. The secondary glaucoma was possibly due to a combination of tumor infiltration affecting the right eye and systemic steroid therapy affecting the eyes.[9] In patients with a malignant cause of masquerade

syndrome, cytologic analysis of intraocular fluids has been reported to be an essential diagnostic procedure with positive yield in 64% of cases.[10] In our case, anterior chamber tap, being an invasive procedure, was not attempted on as the uveitis responded rapidly to topical and oral steroid with cycloplegic therapy.

Conclusion

Clinician must be vigilant about uveitic masquarade syndrome and atypical hypopyon uveitis can be an indicator of the relapse in cases of acute lymphoblastic leukemia.

Conflict of Interest: None declare

Reference

1. Nussenblatt RB, Whitcup SM. Uveitis: fundamentals and clinical practice, 3rd ed. St. Louis, MO: CV Mosby; 2004. p. 409-19
2. Amaro MH, Muccioli C, Abreu MT. Ocular masquerade syndrome due to intraocular lymphoma--two forms of retinal pigment epithelium involvement: case reports. Arq Bras Oftalmol. 2007 May-Jun; 70(3):521-5. PubMed PMID: 17768564.
3. Bunin N, Rivera G, Goode F, Hustu HO. Ocular relapse in the anterior chamber in childhood acute lymphoblastic leukaemia. J Clin Oncol 1987; 5:299-303.
4. Tasneem A F, Shwetha B A, Amanpreet Kaur, Merlin Benzy. Ocular Masquerade syndrome. MedPulse–International Medical Journal September 2014; 1(9): 534-536.
5. Gass JD, Sever RJ, Grizzard WS, Clarkson JG, Blumenkranz M, Wind CA, et al. Multifocal pigment epithelial detachments by reticulum cell sarcoma. A characteristic funduscopic picture. Retina. 1984; 4(3):135-43.
6. Patel SV, Herman DC, Anderson PM, Al-Zein NJ, Buettner H. Iris and anterior chamber involvement in acute lymphoblastic leukemia. J Pediatr Hematol Oncol. 2003; 25:653-656.
7. Wadhwa N, Vohra R, Shrey D, Iyer VK, Garg S. Unilateral hypopyon in a child as a first and sole presentation in relapsing acute lymphoblastic leukemia. Indian J Ophthalmol. 2007; 55:223-224.
8. Bunin N, Rivera G, Goode F, Hustu HO. Ocular relapse in the anterior chamber in childhood acute lymphoblastic leukaemia. J Clin Oncol 1987; 5:299-303.
9. Hegde SP, Ursekar AT, Chitale AA. Relapsing acute myeloid leukemia presenting as hypopyon uveitis. Indian J Ophthalmol 2011; 59:391-3.
10. Rothova A, Ooijman F, Kerkhoff F, Van Der Lelij A, Lokhorst HM. Uveitis masquerade syndromes. Ophthalmology 2001; 108:386-99.