Bilateral Hypopyon Uveitis in Chronic Myeloid Leukemia

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Established Facts

- Atypical or recalcitrant hypopyon uveitis can have an underlying etiology of masquerade syndrome.
- Leukemic hypopyon can be an indicator of a blast crisis, a central nervous system involvement, or a sign of relapse.

Novel Insights

- Bloodstained hypopyon can be an indicator of underlying leukemia like chronic myeloid leukemia.
- Anterior chamber paracentesis and peripheral blood smear can aid in the diagnosis of leukemia.

Keywords

Hypopyon · Chronic myeloid leukemia · Masquerade syndromes

Abstract

Background: A leukemic hypopyon is considered an early sign of central nervous system involvement or systemic relapse. A differential diagnosis of masquerade syndromes should be considered in cases of hypopyon uveitis that are atypical or unresponsive to treatment. We report a case of a 45-year-old man who presented with bilateral hypopyon uveitis and was subsequently diagnosed as having chronic myeloid leukemia. **Method:** Retrospective case review. **Results:** A 45-year-old diabetic male presented with dimin-

ished vision in both eyes for 10 days. Ophthalmic evaluation revealed rubeosis iridis, hypopyon, and signs of proliferative diabetic retinopathy with panretinal laser photocoagulation scars. He subsequently presented 1 week later with a blood-stained hypopyon in his right eye and a persistent hypopyon in his left eye. A peripheral blood smear and subsequent bone marrow trephine biopsy confirmed the diagnosis of chronic myeloid leukemia in blast crisis and he was referred to an oncologist for further management. **Conclusion:** A recalcitrant or atypical hypopyon uveitis can be an indicator of a blast crisis or a central nervous system involvement or sign of a relapse in cases of leukemia. The presence of unusual bloodstained hypopyon helped in identifying the presence of chronic myeloid leukemia and aided in a prompt oncology consultation.

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Introduction

Hypopyon is a sediment forming in the dependent part of the anterior chamber of the eye. It occurs in association with intraocular inflammation, infection, and rarely in malignancies. It is a dramatic but short-lived finding secondary to intraocular inflammation. Ocular manifestations of leukemia are not rare and may involve

60–90% of patients at some time during the course of their disease [1]. The frequency of involvement of sites within the eye varies with the type of leukemia. The occurrence of leukemic hypopyon, however, is rare, is more common in acute versus chronic leukemia, and in general is considered an early sign of central nervous system involvement or systemic relapse. It is important to be aware of the differential diagnosis of masquerade syn-

Fig. 1. Slit-lamp photograph of the patient at the time of presentation showing a hypopyon in the right (a) and left eye (b). He presented 1 week later with a sudden decrease in vision in his right eye, which had developed a bloodstained hypopyon (c). The left eye had a persistent hypopyon (d). Peripheral blood smear (Leishman stain) showing an increased shift to the left of leukocytes (e, 40× magnification), marked leukocytosis (f, 10× magnification), and presence of promyelocytes, metamyelocytes, myeloblasts, and band cells (g, h).

dromes in cases of hypopyon uveitis that are atypical or unresponsive to treatment. Metastatic secondaries in the eye should always be considered a differential diagnosis in cases of recalcitrant or nonresponsive uveitis, as ocular and leptomeningeal metastases are known to occur in leukemias and lymphomas.

We report a case of a 45-year-old man who presented to us with bilateral hypopyon uveitis and was subsequently diagnosed as having chronic myeloid leukemia (CML).

Case

A 45-year-old diabetic male presented with diminution of vision in both eyes for 10 days. His vision at presentation was 20/100 in the right eye and counting fingers close to the face in the left eye. Anterior segment evaluation revealed rubeosis iridis and a hypopyon in both eyes (Fig. 1a, b) and retinal evaluation revealed the signs of bilateral proliferative diabetic retinopathy with panretinal laser photocoagulation scars. He was advised to undergo routine uveitis investigations and review with reports. He subsequently presented 1 week later with a sudden decrease in vision in his right eye. Anterior segment examination revealed the presence of a bloodstained hypopyon in his right eye out of proportion to the rubeosis (Fig. 1c) and a persistent hypopyon in his left eye (Fig. 1d). The patient's glycemic status was well under control. All investigations to rule out systemic causes of bilateral uveitis were within normal limits. He, however, had a high total blood leukocyte count of 190,000/mL. An anterior chamber paracentesis was done, which revealed predominantly neutrophils and eosinophils and was suggestive predominantly of inflammation. Microbiology of the anterior chamber paracentesis revealed no organisms on routine smears and cultures.

However, his peripheral blood smear revealed a total leukocyte count of 116,000/mL, an increased shift to the left of leukocytes (Fig. 1e), marked leukocytosis (Fig. 1f), and the presence of band forms, promyelocytes, metamyelocytes, and myeloblasts (Fig. 1g, h). His bone marrow trephine biopsy confirmed the diagnosis of CML in blast crisis. The patient was referred to an oncologist and chemotherapy was started for the patient. The patient was subsequently lost to follow-up. However, the presence of unusual bloodstained hypopyon helped in identifying the presence of CML and aided in a prompt oncology consultation.

Discussion

Hypopyon can occur during the lymphoid blast crisis in known cases of CML. One of the hypotheses for the occurrence of hypopyon in masquerade syndromes is the sequestration of cells in the long posterior ciliary vessels and their passage into the anterior chamber through the iris vessels [2]. Hypopyon in masquerade syndrome is mostly bloodstained and is associated with elevated intraocular pressure [3, 4]. However, unlike uveitic entities,

masquerade syndromes usually do not have significant cells in vitreous.

Ayliffe et al. [4] reported a patient with acute myeloid leukemia whose only sign of relapse was bilateral anterior uveitis with pseudohypopyon that was refractory to topical and systemic steroids and an anterior chamber paracentesis revealed myeloblastic leukemic cells with morphologic characteristics similar to those present in the original bone marrow biopsy obtained 14 months earlier. In our case, though the paracentesis did not reveal any blast cells, we still believed that the bloodstained hypopyon was unusual and warranted further systemic evaluation. The absence of blast cells in paracentesis can probably be explained by an inadequate sample.

Santoni et al. [5] reported a case of bilateral hypopyon in a 40-year-old patient with CML just after an episode of leukemic meningiosis. Lipton et al. [6] reported a case of leukemic hypopyon as a solitary site of relapse in a patient with CML in lymphoid blast crisis. Sudharshan et al. [7] reported a similar case of bilateral hypopyon uveitis where blood smear revealed immature cells (myelocytes, band forms, and promyelocytes) consistent with a picture of CML.

Bilateral hypopyon uveitis is an uncommon initial presenting sign of CML and it is imperative for ophthalmologists and oncologists to be aware of the differential diagnosis of masquerade syndromes in cases of hypopyon uveitis. The occurrence of leukemic hypopyon can be an indicator of a blast crisis, a central nervous system involvement, or a sign of relapse.

We believe that it is important to consider masquerade syndromes as a differential diagnosis in cases of hypopyon uveitis that are atypical or unresponsive to treatment. The presence of unusual bloodstained hypopyon helped in identifying the presence of CML in our case and aided in a prompt oncology consultation.

Acknowledgements

Support was provided by Hyderabad Eye Research Foundation (SK), Hyderabad, India. The funders had no role in the preparation, review, or approval of the manuscript.

Statement of Ethics

This submission complies with the guidelines for human studies and animal welfare regulations. This paper does not involve any experimental drugs that do not have FDA approval. The treatment of all subjects described in this paper was carried out after obtaining prior informed consent. No animal experiments were carried out with regards to this paper.

Disclosure Statement

All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bu-

reaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or nonfinancial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter or materials discussed in this paper.

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