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Unilateral Subhyaloid Hemorrhage as a Presenting Sign of Chronic Myeloid Leukemia

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
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Patient: Female, 19-year-old
Final Diagnosis: Chronic myeloid leukemia
Symptoms: Blurring of vision
Medication: —
Clinical Procedure: —
Specialty: Hematology • Ophthalmology

Objective: Unusual clinical course

Background: Chronic myelogenous leukemia (CML) is a malignant myeloproliferative neoplasm of pluripotent stem cell origin. Ophthalmic manifestation as an initial presentation in cases of CML is extremely rare. Frequently, ocular lesions in CML are asymptomatic. However, vitreous or foveal involvement can result in a symptomatic visual loss and earlier presentation. Here, we report a rare case of monocular vision loss due to subhyaloid hemorrhage in a case of CML.

Case Report: A 19-year-old healthy woman presented to the Emergency Department with sudden painless decrease in vision in her left eye for 1 day. Fundus examination revealed multiple intraretinal hemorrhages with some white-centered hemorrhages in 4 quadrants in both eyes, and subhyaloid hemorrhage involving the fovea in the left eye. Complete blood count and peripheral blood smear were consistent with the diagnosis of chronic myeloid leukemia. After referral to the hematology service, the diagnosis was confirmed based on bone marrow aspiration and chromosomal analysis. The patient then received the appropriate management and continued to follow up with the hematology service.

Conclusions: This case report highlights the rarity of ocular involvement as an initial manifestation of chronic myeloid leukemia, and the importance of systemic work-up for the diagnosis of this entity. A multidisciplinary team approach involving ophthalmologists, hematologists, and oncologists is paramount for the diagnosis and management of CML.

Keywords: Leukemia, Myelogenous, Chronic, BCR-ABL Positive • Retinal Hemorrhage • Vision, Monocular

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/936266

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Background

Chronic myeloid leukemia (CML) is a myeloproliferative disorder that is associated with the translocation between the long arms of chromosome 9 and 22, leading to the fusion of BCR and ABL1 and formation of the BCR-ABL1 oncogene [1]. Ocular manifestation as the initial presentation of CML has been estimated to occur in about 5-10% of cases [2]. There are 2 possible mechanisms for retinal findings in CML. The first is by direct invasion of neoplastic cells to the orbit and other tissues (iris, choroid, optic nerve, and retina). The second is leukemic retinopathy associated with hematologic abnormalities like anemia and hyperviscosity, which result in tortuous and dilated veins, and perivascular sheathing due to the collection of leukemic cells [2]. To date, there is only 1 such case reported in the literature, of a young male patient presenting with unilateral subhyaloid hemorrhage as a first sign of CML [3].

Case Report

A 19-year-old healthy woman presented to the Emergency Department (ED) with sudden painless decrease in vision in her left eye for 1 day. Past medical and family histories were negative for blood dyscrasias. She denied any history of trauma, history of using medications, or history of constitutional symptoms (eg, fever, weight loss, fatigue). On physical examination, blood pressure on her visit to the ED was 120/70 mmHg and body temperature was 37.2°C. Visual acuity was 20/20 in the right eye and 20/100 in the left eye. Intraocular pressure was 17 mmHg in the right eye and 16 mmHg in the left eye. An anterior segment examination was unremarkable in both eyes. A fundus examination revealed multiple intraretinal hemorrhages with some white-centered hemorrhages (Roth's spots) in 4 quadrants in both eyes (**Figure 1A, 1B**) (Fundus Camera Topcon TRC-50EX). In addition, the left eye showed a subhyaloid hemorrhage involving the fovea (**Figure 1B**). Fundus fluorescein angiography (FFA) (Fluorescein Angiogram Topcon TRC-50EX)

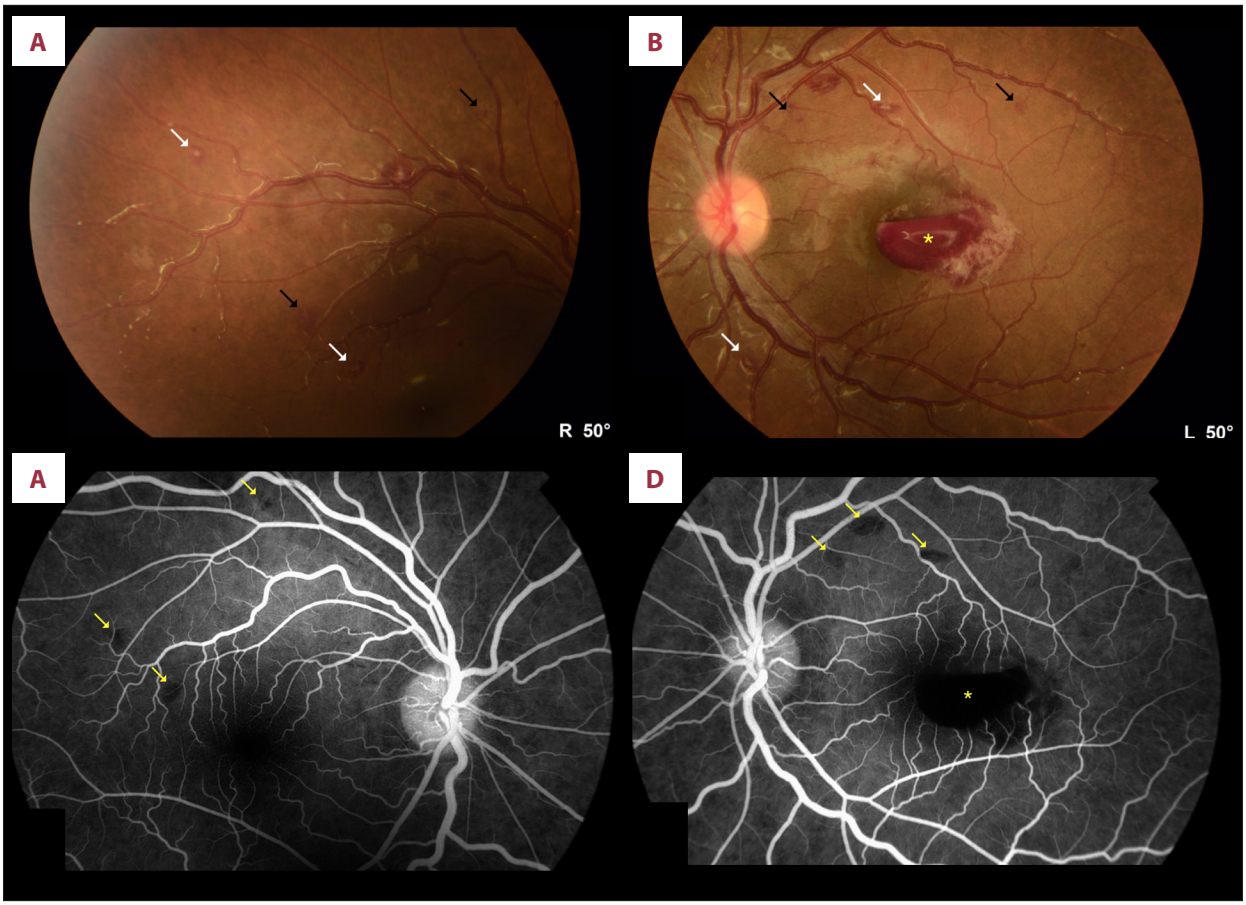

Figure 1. Color fundus photo of the right eye (**A**) and left eye (**B**) showing multiple intraretinal hemorrhages (black arrows) with few white-centered hemorrhages (Roth's spots) (white arrows) involving the macula and posterior pole. Subhyaloid hemorrhage over the fovea in the left eye (yellow asterisk) (**B**). Fundus fluorescein angiography (FFA) of the right eye (**C**) and left eye (**D**) demonstrating hypofluorescent areas due to a blockage effect from intraretinal (yellow arrows) and subhyaloid hemorrhages (yellow asterisk).

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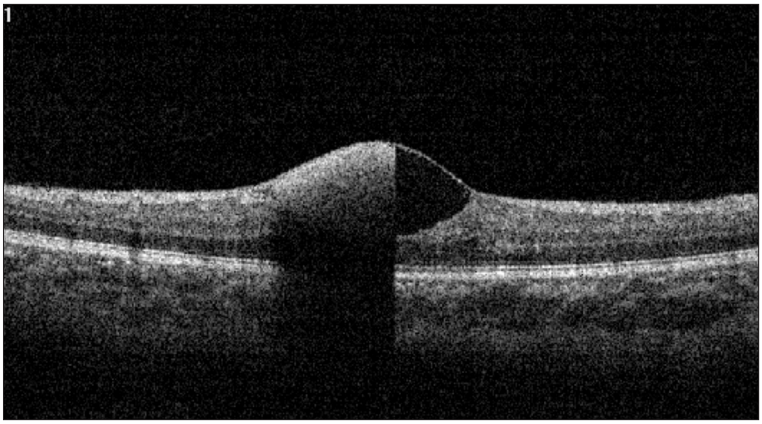

Figure 2. Spectral domain optical coherence tomography depicting dense hyperreflective materials (hemorrhage) under the internal limiting membrane (ILM), consistent with premacular subhyaloid hemorrhage.

demonstrated hypofluorescent areas due to a blockage effect from intraretinal and subhyaloid hemorrhages, with no other abnormalities detected (**Figure 1C, 1D**). A spectral domain optical coherence tomography (SD-OCT) (Optovue, Inc, Freemont CA, USA) depicted dense hyperreflective materials (hemorrhage) under the internal limiting membrane (ILM), consistent with premacular subhyaloid hemorrhage (**Figure 2**).

The patient underwent a complete systemic work-up. Complete blood count (CBC) revealed profound leukocytosis (WBC: 226×10^9/L), low hemoglobin count (Hg: 4.6×10^9/L), and normal platelet count (294 000/uL). A peripheral blood smear (PBS) showed leukocytosis with marked left-shift granulocyte, 6% basophilia, 7% eosinophilia, and 3% blast cells. The patient was referred to the hematology team and subsequently underwent bone marrow aspiration, which revealed hypercellular bone marrow. Chromosomal analysis was done, and BCR-ABL was detected. Eventually, the patient was diagnosed with chronic myeloid leukemia in chronic phase (CML-CP). She received 1 unit of packed red blood cells (PRBCs) and was started on 1.5 g hydroxyurea twice a day along with 300 mg allopurinol twice a day. The patient continued to follow up with the hematology service. Ten months following presentation, the patient achieved 20/20 vision in both eyes, with complete resolution of the subhyaloid hemorrhage in the left eye.

Discussion

Leukemic ophthalmopathy is rarely reported in cases of CML. In fact, the estimated prevalence of leukemic retinopathy in CML has been reported to be 13%, with 68%, 42%, and 33% in cases of acute myeloid leukemia, acute lymphoid leukemia, and chronic lymphoid leukemia, respectively [5]. Vision-threatening complications such as macular subhyaloid hemorrhage and vitreous hemorrhage have exclusively been reported in acute leukemias. As a matter of fact, only 1 reported case of subhyaloid hemorrhage in patients with CML was found in the literature [3], which makes our case the second reported case in the English literature.

Sanjay and Huang [3] reported a young male who presented with visual disturbance due to preretinal hemorrhage over the fovea. Similarly, our patient presented with monocular vision loss secondary to subhyaloid hemorrhage with no systemic symptoms of CML. Soman et al [5] showed that by increasing hemoglobin level from 5 to 7 g/L and platelet count from 10 000 to 50 000 cells/mm^3, the possibility of having subhyaloid hemorrhage is significantly reduced by >50%. Our patient had low hemoglobin, for which she required blood transfusion, but no platelet transfusion was needed as the platelet counts were normal.

The numerous ocular manifestations in leukemia depend on a number of factors, such as age, type of leukemia, staging, and response to systemic chemotherapy or bone marrow transplantation [5]. Retinal lesions are the most common ocular features in leukemia [4]. Ophthalmic manifestations of CML can also include vitreous hemorrhage, multi-layer hemorrhage (preretinal, subretinal, and intraretinal), Roth's spots, and optic nerve infiltration [3,5,6]. Interestingly, Seraly et al [6] reported an unusual combination of multi-layer hemorrhage, retinal tear, and retinal detachment causing monocular vision loss in a patient with CML.

Several hematological disorders can mimic leukemic retinopathy and have similar fundus findings, including anemic retinopathy, sickle-cell retinopathy, and lymphoma-associated retinopathy [3]. Other differential diagnoses for subhyaloid hemorrhage are trauma, Valsalva retinopathy, proliferative diabetic retinopathy (PDR), choroidal neovascular membrane (CNVM), retinal arterial microaneurysm (RAM), and polypoidal choroidal vasculopathy (PCV), which was ruled out by history and clinical exam in the present case [7-9].

Overall, it is essential to perform ophthalmic screening in all cases of CML as it has a major impact in the prognosis of the disease. For instance, it was reported that the 5-year survival of patients with ophthalmic manifestation was 21.4% compared to 45.7% of patients without ophthalmic manifestations [10].

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Ohkoshi and Tsiaras [10] reported that patients with neuro-ophthalmic signs of central nervous system leukemia and patients with retinal or vitreous hemorrhages at the time of relapse died within a very short period.

Conclusions

In summary, this case report highlights a unique presentation of a monocular vision loss that was attributed to subhyaloid hemorrhage in a patient with CML. It is important to recognize early ophthalmic signs of CML and to perform systemic work-up to ensure prompt diagnosis and management.

Declaration of Figures; Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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