SIGHT-THREATENING OCULAR COMPLICATIONS IN CHRONIC MYELOID LEUKEMIA: A CASE OF VITREOUS HEMORRHAGE AND RETINAL DETACHMENT AT PRESENTATION

Sanjog Kandel,[1] Guru Sharan Sah,[1] Jayaram Adhikari,[1] Bibek Kandel,[1] Bhawana Giri[1]

ABSTRACT

INTRODUCTION

Chronic myeloid leukemia (CML) is a myeloproliferative neoplasm that typically presents with constitutional symptoms, splenomegaly, and hematologic abnormalities. Ocular involvement is rare, and vitreous hemorrhage with retinal detachment as presenting features is exceedingly uncommon.

We report a 29-year-old female with sudden painless blurring of vision in the right eye for 5 days. The patient was a diagnosed case of chronic myeloid leukemia in chronic phase whose hematological workup showed marked leukocytosis with myeloid precursors. Bone marrow aspirate confirmed CML in chronic phase, and cytogenetic analysis demonstrated t(9;22) with BCR–ABL1 positivity. The patient was initiated on imatinib 400 mg daily for the last 2 weeks before ophthalmic presentation.Ophthalmic examination revealed dense vitreous hemorrhage and retinal detachment.

The patient showed partial resolution of vitreous hemorrhage and improved visual acuity over 2 weeks and hematological improvement with continued imatinib therapy.

KEYWORDS

Chronic myeloid leukemia, Vitreous hemorrhage, Retinal detachment

1. Department of Medical Oncology, B.P. Koirala Memorial Cancer Hospital, Chitwan, Nepal

https://doi.org/10.3126/jucms.v13i03.88836

For Correspondence
Dr. Sanjog Kandel
Department of Medical Oncology
B.P. Koirala Memorial Cancer Hospital
Chitwan, Nepal
Email: drsanjograj@gmail.com

INTRODUCTION

Chronic myeloid leukemia (CML) is a clonal myeloproliferative neoplasm characterized by the presence of the Philadelphia chromosome, resulting from a reciprocal translocation between chromosomes 9 and 22 [t(9;22)(q34;q11)], leading to the BCR–ABL1 fusion gene and constitutive tyrosine kinase activation.[1] CML accounts for approximately 15–20% of all adult leukemias, with a global incidence of 1–2 cases per 100,000 population per year.[2]

The disease typically presents in the chronic phase with nonspecific constitutional symptoms such as fatigue, weight loss, night sweats, and abdominal fullness due to splenomegaly.[3] Many patients are diagnosed incidentally through routine blood counts revealing leukocytosis. Ocular involvement in CML is rare and often overlooked, but it may be the first manifestation of the disease in some cases.[4]

Ophthalmic presentation of leukemia can occur due to direct leukemic infiltration of ocular tissues or, more commonly, secondary to hematologic abnormalities such as anemia, thrombocytopenia, hyperviscosity, and leukostasis. Reported findings include retinal hemorrhages, Roth spots, cotton wool spots, optic disc swelling, and vascular occlusions.[5] Vitreous hemorrhage and retinal detachment as the initial presentation of CML are uncommon, with only a few isolated case reports available in the literature.

Recognition of such ocular complications is clinically important, as they may lead to irreversible visual impairment if not diagnosed and managed promptly. Early ophthalmic evaluation in patients with hematologic malignancies presenting with visual complaints is therefore important. We present a case of newly diagnosed chronic-phase CML with initial presentation of vitreous hemorrhage and retinal detachment, highlighting the importance of prompt multidisciplinary intervention.

CASE PRESENTATION

A 29-year-old female with no significant past medical or surgical history presented with a two-week history of progressive fatigue, intermittent headache, dizziness, discomfort in the left upper quadrant of the abdomen and blurring of vision of the right eye. There was no history of fever, weight loss, night sweats, ocular trauma, or bleeding from other sites.

On examination, she was afebrile and hemodynamically stable. Pallor was present but no icterus, lymphadenopathy, or skin rash.There was mild splenomegaly which was palpable 3 cm below the left costal margin but no hepatomegaly on abdominal examination. Neurological examination was normal.

Initial laboratory investigations showed marked leukocytosis with a total white blood cell count of 480 × 10^9/L, hemoglobin of 10.5 g/dL, and platelet count of 180 × 10^9/L. Peripheral blood smear has marked leukocytosis with a full spectrum of myeloid precursors and occasional myeloblasts (<2%). Bone marrow aspiration was hypercellular with marked granulocytic hyperplasia and no excess blasts. Cytogenetic analysis demonstrated the Philadelphia chromosome t(9;22)(q34;q11), and reverse transcription polymerase chain reaction (RT-PCR) confirmed the presence of the BCR–ABL1 fusion transcript.

A diagnosis of chronic-phase chronic myeloid leukemia (CML) was made, and the patient was initiated on oral imatinib 400 mg daily, with supportive measures including hydration and allopurinol prophylaxis.

Two weeks after starting treatment, the patient developed sudden, painless loss of vision in the right eye associated with the perception of multiple red floaters. There was no ocular pain, redness, or photophobia.

Ophthalmic evaluation (Figure 1) at this time revealed a best corrected visual acuity of 6/60 in the right eye and 6/12 in the left eye. Anterior segments were unremarkable in both eyes. Fundus examination of the right eye showed dense vitreous hemorrhage obscuring the macula, with a superior retinal detachment visible in the peripheral retina. The left eye fundus showed disc edema with roth spots with focal and diffused vascular attenuation.

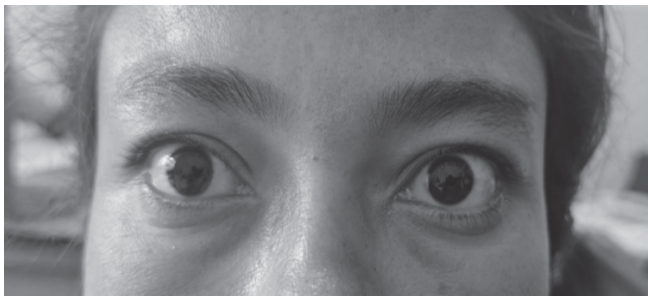

Figure 1. Showing patient's bilateral eye at the time of examination

Ocular imaging with B-scan ultrasonography and Scanning Laser Ophthalmoscopy (SLO) of the right eye confirmed the presence of dense vitreous hemorrhage with partial superior retinal detachment; (Figure 2) the left eye was normal.
Contrast-enhanced magnetic resonance imaging (CE MRI) of the brain and orbit was performed to rule out any orbital or intracranial pathology;showed features of right retinal detachment. Both MRI and Magnetic resonance venography (MRV) were unremarkable, showing no evidence of mass lesions, optic nerve involvement, or venous sinus thrombosis.

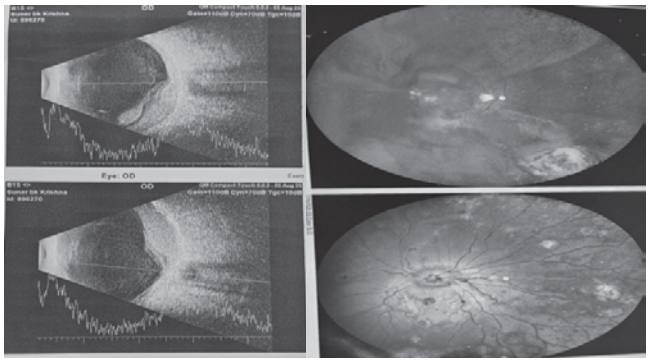

Figure 2. Ocular imaging with B-scan ultrasonography and Scanning Laser Ophthalmoscopy confirmed the presence of dense vitreous hemorrhage with partial superior retinal detachment.

Renal function tests were within normal limits, including serum creatinine at 0.9 mg/dL and blood urea nitrogen at 15 mg/dL. Liver function tests were normal with total bilirubin of 0.8 mg/dL, ALT 28 U/L, and serum albumin 4.1 g/dL. The coagulation profile was also normal, including bleeding time, clotting time, prothrombin time, INR, aPTT, fibrinogen, and von Willebrand factor levels. Thyroid function tests and vitamin D levels were within normal ranges. Echocardiography showed normal cardiac function, cerebrospinal fluid analysis was normal without evidence of CNS involvement, and infectious serology was negative.

The ophthalmology team planned for conservative management of the ocular findings, with close observation until hematologic stabilization was achieved. At the time of ophthalmic evaluation, the patient's WBC count had already decreased to 9.2×10^9/L.

At 1 month follow-up, there was partial clearing of the vitreous hemorrhage with improvement in right eye visual acuity to 6/24, and the retina remained attached. The left eye vision remained unaffected throughout the course.

DISCUSSION

Ocular involvement in chronic myeloid leukemia (CML) is an under-recognized but important clinical entity. While ocular manifestations are reported in 30–50% of leukemic patients during the disease course, they are rarely the initial or early complication in chronic-phase CML.[6] The most common findings include retinal hemorrhages, cotton wool spots, and venous tortuosity, with vitreous hemorrhage and retinal detachment considered unusual. [7,8]

Our patient presented with hyperleukocytosis (WBC 480×10^9/L) and developed vitreous hemorrhage with tractional retinal detachment within two weeks of initiating imatinib, despite being in chronic phase and having a normal platelet count. This presentation differs from many case series in which such severe ocular events occurred in blast crisis or with coexisting thrombocytopenia or coagulopathy. [9,10] For example, Reddy et al[7] reported vitreous hemorrhage in only 1–2% of leukemic eyes, predominantly in acute leukemias or advanced CML. Saeed et al[8] described a CML case with vitreous hemorrhage where platelet count was low, suggesting hemorrhagic risk from thrombocytopenia, a factor absent in our case.

The preserved platelet count suggests that vascular injury in our patient was more likely related to hyperleukocytosis-induced leukostasis. Leukostasis causes microvascular occlusion, endothelial damage, ischemia, and increased vascular fragility.[11] Kincaid and Green described histopathologic evidence of intravascular leukemic aggregates in the retinal vasculature, supporting this mechanism. In hyperleukocytic states, blood viscosity increases disproportionately with leukocyte count, impairing retinal perfusion and predisposing to neovascularization and hemorrhage.[12]

The onset of ocular symptoms soon after imatinib initiation raises the possibility that rapid cytoreduction contributed to vascular events. Similar phenomena have been described in tumor lysis-related leukostasis, where abrupt changes in hematologic parameters alter microvascular hemodynamics.[13] Chan et al noted that TKI-related ocular events, though rare, may occur due to changes in vascular permeability during early treatment.[10]

Retinal detachment in leukemia is typically tractional, secondary to proliferative retinopathy from ischemia and recurrent vitreous hemorrhage.[14] Maganti et al recently reported rhegmatogenous retinal detachment in CML, emphasizing that both ischemic and mechanical factors can be involved.[15] In our case, the detachment likely resulted from fibrovascular traction after vitreous hemorrhage in an ischemic retina.

Prompt systemic control of CML with TKIs or cytoreductive therapy is critical, but visual prognosis depends on the extent of irreversible ischemic damage.[14,16] Leong et al reported partial visual recovery after leukapheresis in a patient with leukostasis retinopathy, but outcomes were poor in cases with established detachment or extensive hemorrhage.16 Early ophthalmology consultation is essential, and surgical intervention such as pars plana vitrectomy may be required in selected cases.

This case highlights that severe ocular complications can occur even in chronic-phase CML without thrombocytopenia. Hyperleukocytosis and leukostasis remain important mechanisms for vision-threatening events. Rapid hematologic changes during early TKI therapy may precipitate or unmask ocular pathology. A high index of suspicion and multidisciplinary management are essential to preserve vision.

This case adds to the limited but growing body of literature emphasizing that vigilance for ocular symptoms is necessary in CML patients, and prompt multidisciplinary management is essential to prevent irreversible vision loss.

CONCLUSION

Sight-threatening ocular complications such as vitreous hemorrhage and retinal detachment, although rare, can occur even in chronic-phase CML without thrombocytopenia or blast crisis. Hyperleukocytosis and leukostasis are likely key pathogenic mechanisms, and rapid hematologic shifts during early tyrosine kinase inhibitor therapy may precipitate or unmask these events. This case underscores the need for heightened clinical vigilance, prompt ophthalmic evaluation, and a multidisciplinary approach in CML patients presenting with new visual symptoms. Early recognition and timely intervention are essential to optimize both systemic and visual outcomes.

CONFLICT OF INTERESTS

None

REFERENCES

1 Rowley JD. A new consistent chromosomal abnormality in chronic myelogenous leukemia identified by quinacrine fluorescence and Giemsa staining. Nature. 1973;243 (5405):290-3.

2. Hochhaus A, Saussele S, Rosti G, Mahon FX, Janssen J, Hjorth- Hansen H, et al. Chronic myeloid leukemia; ESMO Clinical Practice Guidelines. Ann Oncol. 2020;31(6);701-13.

3. Bowe H, Bjorkholm M, Dickman PW, Hoglund M, Lambet PC, Anderson TM. Life expectancy of patients with chronic myeloid leukemia approaches the life expectancy of the general population. J Clin oncol. 2016;34(24):2851-7.

4. Russo V, Scott IYU, Querques G, Stella A, Barone A, Delli noci N. Ocular manifestations of leukemia: a case report and review of the literature. BMC Ophthalmol.2010;13:13.

5. Rosenthal AR.Ocular manifestation of leukemia: a review. Surv Opthalmol 1975;19(5):394-408.

6. Kincaid MC, Green WR. Ocular and orbital involvbement in leukemia. Surv Opthalmol. 1983;27(4):211-32.

7. Reddy SC, Takkar B, Singhal S, Sahni S, Garg SP. Ocular manifestations of leukemia: A review. Indian Opthalmol. 2021;69(6):1440-8.

8. Saeed MU, Mukhtar F, Shaikh AS, Sheikh ZI, Channar MS. Leukemic retinopathy and vitreous hemorrhage in chronic myeloid leukemia. J Pak Med Assoc. 2015;65(5):530-2.

9. Mahendra C, Mahendra B, Kumar G, Rudrappa S. Ocular manifestations in leukemia patients: a prospective study. Ann Hematol. 2013;92(4):473-9.

10. Chan W, Lai TY, Liu DT, Lam DS. Ocular manifestations of leukemia: a review. Hong Kong Med J. 2004;10(5):331-6.

11. Beltrami G, Barbui T, Finazzi G. Leukostasis: clinical features, pathogenesis and management. Hematol Occol Clin North Am. 2003;17(3):749-71.

12. Kaur C, Ling EA. Blood–retinal barrier in hypoxic and ischemic conditions: Basic concepts, clinical features and management. Prog Retin eye Res. 2008;27(6):622-47.

13. Hegde V, Puthuran GV, Dada T, Pathengay A, Sachdeva S, Hegde R, et al. Rapid cytoreduction and ocular complications in CML: A case series. Eur J Haematol. 2016;96(1):84-7.

14 Shields JA, Shields CL. Intraocular involvement of leukemia: a review,Cancer Control. 1997;4(4):287-95.

15. Maganti N, Crabtree GS, Chang JS, Schildroth KR. A case of rhegmatogenous retinal detachment in chronic myeloid leukemia. Retin Cases Brief Rep. 2024 Mar 4.

16. Leong B, Lim L, Tan A. Acute vision loss due to CML leukemic retinopathy reversed with leukapheresis: a case report. Clin Case Rep. 2023;11:e7441.