

Improvement of Leukemic Retinopathy After Leukapheresis In Chronic Myelogenous Leukemia with Leukostasis: A Case Report

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ABSTRACT

Introduction: to report a case of bilateral leukemic retinopathy due to leukostasis that was successfully managed by leukapheresis.

Case Presentation: 31-year-old male with mild visual disturbance was referred to ophthalmology department. He suffered from Chronic Myelogenous Leukemia (CML) with white blood cell (WBC) count 533.900/microL. He was started on hydroxyurea, allopurinol, and once leukapheresis. Ophthalmologic evaluation revealed visual acuity of 4/4 in the right eye and 4/6,3 in the left eye. Funduscopy examination showed the presence of bilateral papilledema, venous engorgement, tortuosity, and retinal hemorrhages. Then this patient continued with second leukapheresis.

Result: Visual acuity, laboratory examination, and funduscopy finding was evaluated. His visual acuity was improved, papilledema and retinal blood vessels abnormality had markedly reduced concurring with the patient's hematological remission. Decreasing WBC count after leukapheresis has improved blood flow that reflected from the retinal findings and visual acuity improvement.

Conclusion: Leukapheresis treatment is sufficient to improved clinical condition for leukemic retinopathy caused by CML with leukostasis.

Keywords: chronic myelogenous leukemia (CML), hyperleukocytosis, leukostasis, leukemic retinopathy

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INTRODUCTION

Chronic Myelogenous Leukemia (CML) which is also known as Chronic Myeloid Leukemia is a myeloproliferative disorder characterized by increased proliferation of granulocyte cells. CML occupies 20% of all leukemia in adults.¹ The white blood cell counts of patients diagnosed with chronic myeloid leukemia (CML) vary greatly. Study by Koshy et al., revealed that leukemic ophthalmic lesions were found in 43.8% patients with acute and chronic leukemias. Ocular involvement is more often seen in adults, acute and myeloid leukemias.²

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Leukostasis is characterized by the occlusion of the microvasculature by white blood cells.³ We report the case of a previously healthy man with mild visual loss, striking fundus changes, and an extremely high white blood cell count who had significant recovery of vision after leukapheresis.

CASE PRESENTATION

A 31-year-old man was diagnosed blast phase of CML from internal medicine department and referred to ophthalmology department after reporting mild visual loss in both eyes for 3 months. Additional history revealed epistaxis, back pain, night sweats, and weight loss. A complete blood count revealed white blood cell (WBC) count of 533.900/mm³ (reference range: 3.500 to 11.000/mm³), platelet count of 139.000 (reference range: 150.000 to 400.000/mm³), and hemoglobin concentration of 8.7 g/dL (reference range: 14-18 g/dL).

The patient was treated with hydroxyurea, allopurinol, and two sessions of leukapheresis. At his first visit, after the first leukapheresis, examination showed best-corrected visual acuity (BCVA) of 4/4 in the right eye and 4/6,3 in the left eye. Anterior segment examination by slit lamp biomicroscope showed no abnormality. Posterior segment examination revealed papilledema, dilated retinal veins, diffuse retinal hemorrhages, and white-centered hemorrhages on both eyes (Figure 1).



Figure 1. Fundus photographs of the right and left eyes illustrate the present of papilledema, dilated retinal veins and tortuosity, diffuse retinal hemorrhages, and white-centered hemorrhages (Roth's spot).

The patient was diagnosed with leukemic retinopathy in both eyes. There was no specific therapy from ophthalmology department. Second examination was done 3 weeks after the second leukapheresis. A complete blood count revealed WBC count of 112.770/mm³, platelet count of 522.000, and hemoglobin concentration of 10,3 g/dL. His visual acuity was improved (4/5 on left eye), papilledema and retinal blood vessels abnormality had markedly reduced concurring with the patient's hematological remission (Figure 2).

DISCUSSION

The manifestation of leukemia in the eye can be in the form of direct infiltration of leukemia cells or is the impact of leukocytosis, anemia, and thrombocytopenia.⁴ It can involve in various ocular tissue. Conjunctival involvement, is not a common presentation of leukemias. Cellular involvement is found at all levels of the substantia propria and can be diffuse or patchy, tending to concentrate along blood vessels.

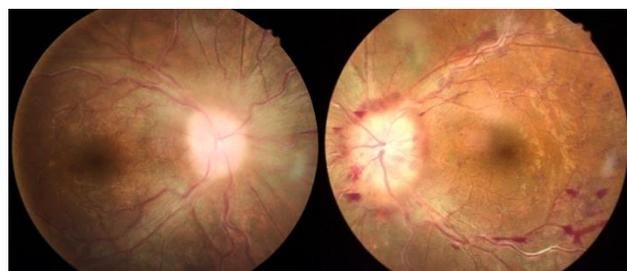


Figure 2. Fundus photographs of the right and left eyes showed reduced papilledema and retinal blood vessels abnormality 3 weeks after the second leukapheresis.

The involvement of choroid and ciliary body may occur in anterior segment. Clinically, it is characterised by a change in iris colour, and a pseudohypopyon. The intra-ocular pressure (IOP) can be high enough to cause signs and symptoms of acute glaucoma with normal depth anterior chamber. Orbital infiltration in leukaemia presents with exophthalmos, lid oedema, chemosis and pain. All types of leukaemia may involve the orbit; however, anterior segment and

orbital involvement is more common in acute and lymphoid leukaemias.⁵

Leukemic retinopathy is a common complication in both acute and chronic forms.⁶ Fundus examination can illustrate the effects of CML in the body. Its pathophysiology is still unclear but other mechanisms that may play a role in CML include: anemia; leukostasis; hyperviscosity syndrome; leukoembolization; endothelial lesions and local thrombosis secondary to toxic products released by leukemia cells; angiogenic factors released by the ischemic retina; and elevated serum levels of angiogenic growth factors.⁷ Treatment of the underlying causes of the ocular findings could actually result in their improvement or even resolution.

This patient has received cytoreduction treatment with hydroxyurea. This medication has rapid onset and short duration so that it can manage hyperviscosity syndrome with shorter period of myelum suppression. To anticipate hyperuricemia caused by hydroxyurea therapy, this patient was given oral allopurinol to altered uric acid production. Leukapheresis is the best option for patient with hyperleukocytosis and not pregnant, combined with hydroxyurea to give the best result in decreasing leukocyte.⁸ Three weeks after second leukapheresis, visual acuity left eye become 4/5. Fundus examination showed improvement along with decreasing leukocyte. The results obtained in this case report are inversely proportional to the results of the study by Soman *et al.* where statistically there was no correlation between the number of leukocytes and ophthalmic manifestations.⁹

The ocular abnormality found in leukemic retinopathy can also mimic some other disease such as retinal vein occlusion, congenital or hereditary vein tortuosity, papilloedema, or even

infection. It is recommended that ophthalmologist have to run other supportive diagnostic test (color vision test, contrast sensitivity test, optical coherence tomography, visual field examination, brain imaging) if the underlying systemic disease is unknown.

CONCLUSION

This case report underlined the importance of routine eye examinations in patients with leukemia and vice versa in healthy patients with signs of leukemic retinopathy to complete blood tests and referral to hematologist for further management, because the key is to treat the underlying malignancy.

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