International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Unique Case of Unilateral Leukemic Retinopathy in a k/c/o Chronic Myeloid Leukemia (Remission on Stopping the Treatment)

Dr. Prateek Maurya, Dr. Shikha Bhardwaj, Dr. Mridul Trisal

Abstract: ***Chronic myelogenous leukemia (CML), also known as chronic myeloid leukemia, is a myeloproliferative disorder characterized by increased proliferation of the granulocytic cell line without the loss of their capacity to differentiate. Consequently, the peripheral blood including occasional blast cells. CML accounts for 20% of all leukemias affecting adults. Retinal lesions are the most common ocular manifestation of leukemia. Often in adults and in patients with myeloid leukemia. Despite the significant. Ophthalmologic manifestations are among the therapeutic challenge. Here we present a case of CML (chronic phase) with ophthalmologic manifestations, trying to shed light on this important type of presentation.***

Keywords: Leukemic retinopathy**,** chronic myeloid leukemia, Optic neuritis

1. Introduction

Cell profile shows an increased number of granulocytes and their immature precursors.

2. Purpose

To report a case of unilateral leukemic retinopathy (OS) secondary to diagnosed case of chronic myeloid leukemia on halting the treatment.

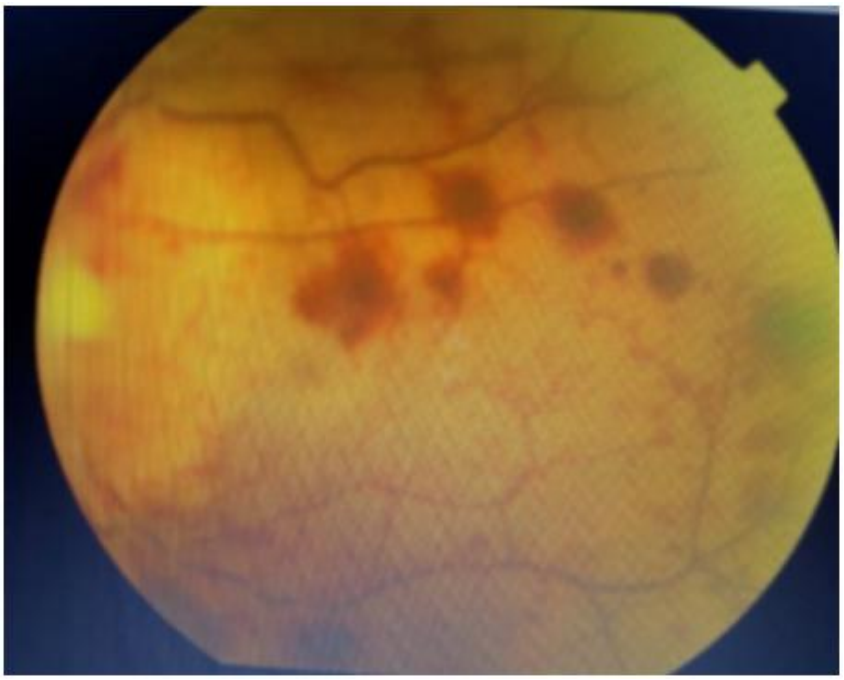
3. Observation

A 59 - year - old male patient who is a known case of PLHA, HBsAg positive and diagnosed case of Chronic myeloid Leukemia for 2 years with Cytogenetic test detected Abelson murine leukemia (ABL) – breakpoint cluster region (BCR) fusion transcript, HLA DR +ve, CD 13, CD79, CD 34. Patient was on treatment which he stopped from last 3 months. Patient then developed diminution of vision left eye for 1 month. Ophthalmic examination of OD has 6/9, OS has Perception of light with conjunctival injection, chemosis and RAPD. Extraocular movements were restricted in all directions (OS) Dilated Fundus examination was OD wasWNL; OS was suggestive of Grade 5 papilloedema (Frisenscale) with high exudate infiltration of temporal disc margin, vasculartortuosity, multiple haemorrhages and roth spots, haemorrhages involving the macula. OCT showed multiple intra - retinal hyper - reflective foci corresponding to intra - retinal hemorrhages, and outer retinal hyper - reflective foci in area corresponding to retinal infiltrate.

MRI brain with orbit showed – bulkiness of left optic nerve with intrasubstance signal suggestive of Left Optic neuritis.

Further assessment including blood pressure and pulse rate measurement were consistently normal when taken on four different occasions about the same time. Fasting blood sugar, FBS was also within normal range (5.1 mmol/L), however, glycosylated hemoglobin of 6.5% indicated prediabetic tendencies (normal: 4–6%, prediabetes 6.1–6.4%). An initial complete blood count showed a low hematocrit level (38 % as against an expected normal of 40–52%) and a high red blood cell distribution width of 17% (normal: 10–14%). Red blood cells levels were low (4.0×10^{12}/L as against the expected normal of $4.40–5.90 \times 10^{12}$/L) and white blood cells count was high 56.8×10^{9}/L (normal: $3–10.6 \times 10^{9}$/L). Lymphocytes count was 20.00×10^{9}/L (normal: $0.8–3.9 \times 10^{9}$/L) and granulocytes count was 35.10×10^{9}/L (normal: $1.1–7.8 \times 10^{9}$ L).

Patient was immediately referred to Oncology department but patient was lost to followup.



4. Discussion

Ocular manifestations of leukemia are common, occurring in as many as 80% of eyes from patients with the disease examined at autopsy. Clinical studies have documented ophthalmic findings in as many as 40% of patients at diagnosis of the leukemia. Patients may be asymptomatic, or they may report blurred or decreased vision. Clinically, the retina is the most commonly affected intraocular structure. Leukemic retinopathy is characterized by intraretinal and subhyaloid hemorrhages, hard exudates, cotton - wool spots, and white - centered retinal hemorrhages, also known as pseudo–Roth spots (Fig 20 - 13). (Classically, white - centered hemorrhages associated with endocarditis are termed Roth spots; they may be called pseudo–Roth spots when associated with other diseases.) In leukemia, these findings are usually the result of associated anemia, hyperviscosity, and/or thrombocytopenia. True leukemic infiltrates are less common and appear as yellow - white deposits in the retina and the subretinal space. Perivascular leukemic infiltrates produce gray - white streaks in the retina. Vitreous involvement by leukemia is rare and most often results from direct extension via retinal hemorrhage. If necessary, a diagnostic vitrectomy can be performed to establish a diagnosis. Although the retina is the most commonly affected ocular structure clinically, histologic studies have shown that the uveal tract is more commonly affected by leukemia than is the retina. The uveal tract may serve as a “sanctuary site” for leukemic cells, making the eye more likely to be a site of recurrent disease. Choroidal infiltrates may be difficult to detected with indirect ophthalmoscopy; they may be better detected on ultrasonography as diffuse thickening of the choroid. Serous retinal detachments may overlie these infiltrates. Leukemic involvement of the iris manifests as a diffuse thickening with loss of the iris crypts, and small nodules may be seen at the margin of the pupil in some cases. Leukemic cells may invade the anterior chamber, forming a pseudo hypopyon. Infiltration of the angle by these cells can give rise to secondary glaucoma. With leukemic infiltration of the optic nerve (Fig 20 - 14), the patient may present with severe vision loss and optic nerve edema. One or both eyes may be affected. This is an ophthalmic emergency that requires immediate treatment to preserve as much vision as possible. Systemic imaging, CNS assessment including lumbar puncture with cytology, and bone marrow evaluation are necessary to confirm the diagnosis. Urgent external beam radiation to the optic nerves is typically used along with combined systemic and intrathecal chemotherapy. Leukemic infiltrates may also involve the orbital soft tissue, with resultant proptosis. These rare tumors are referred to as granulocytic sarcomas or chloromas because they are solid masses of granulocytic precursors, including myeloblasts and myelocytes from myelogenous leukemias. The tumors appear to have a greenish hue on direct visualization. The term granulocytic sarcoma is a misnomer as this tumor is not a true sarcoma. These tumors have a predilection for the lateral and medial walls of the orbit. Involvement of the eye may be seen at initial diagnosis or relapse of leukemia; treatment typically consists of systemic chemotherapy. Depending on the response, low - dose radiation to the eye may be included; however, this should be done with caution as it may limit future use of radiation therapy. The exception is optic nerve infiltration with acute vision loss, for which radiotherapy is mandatory for treatment. The prognosis for vision depends on the particular subtype of leukemia and the extent of ocular involvement.

Several processes have been implicated in the ocular manifestations of CML since they epitomize the effects of this disease. Multiple mechanisms may have independently contributed to the observed manifestation. Such mechanisms include: reduced blood flow, vascular stagnation, retinal capillary dropout, and ischemia. Others associate the ocular findings to anemia; leukostasis; hyperviscosity syndrome; leukoembolization; endothelial lesion, and localized thrombosis secondary to toxic products released by the leukemic cells. Angiogenic factors caused by the ischemia resulting in increased serum levels of angiogenic growth factors, comprising elevated levels of vascular endothelial

growth factor, fibroblast growth factor 2, hepatocyte growth factor, and matrix metalloproteinases have all been implicated.

There is no direct treatment for leukemic retinopathy, however, chemotherapy, immunotherapy, and radiotherapy are usually employed to manage the underlining systemic cause. Chemotherapy that is best suited for intraocular leukemic infiltration, while application of external beam radiation is cut out for optic nerve or orbital lesions. The treatment of CML is informed by the specificity of the disease phase. That is, the chronic stable phase, accelerated phase, or the blast crisis. At the chronic phase, BCR - ABL tyrosine kinase inhibitors (TKIs) for example, imatinib and interferon alpha, cytotoxic agents for example, hydroxyurea, and allogeneic hematopoietic cell transplantation (HCT) may be useful. Although allogeneic HCT remains the sole curative therapy, initial treatment with imatinib has been suggested. In most cases, initial imatinib treatment for chronic phase chronic myeloid leukemia halts the disease progression.

5. Results

White cell count (WCC) were elevated. Relapse of CML as ophthalmic manifestation on stopping the treatment for 3 months.

6. Conclusion

Patient was referred to Cancer Institute for immediate start of chemotherapy. Patient was then presented after one month but with no improvement in vision. Treatment may improve patients' survival and symptomatic relief but vision could not be improved.

References

[1] Szeto SK, Mohamed S, Tsang CW, Chan CK. Massive retinal infiltrates as the presenting sign of chronic myeloid leukemia: Clinical and imaging features of leukemic retinopathy. European Journal of Ophthalmology. October 2021. doi: 10.1177/11206721211048792
[2] Vicini G, Nicolosi C, Malandrino D, Tozzetti C, Rizzo S, Sodi A. Leukostasis retinopathy with leukemic infiltrates as onset manifestation of chronic myeloid leukemia: a case report. European Journal of Ophthalmology.2021; 31 (5): NP116 - NP121. doi: 10.1177/1120672120930679
[3] AAO Ocular pathology and oncology 2020 - 2021
[4] Gupta, A. K., Basnet, R., Yadav, R. D., & Singh, S. (2021). Leukemic retinopathy, a rare entity: A case report. *Asian Journal of Medical Sciences*, *12* (9), 160–165. https: //doi. org/10.3126/ajms. v12i9.37021
[5] Awuah A, Asiedu K, Adanusa M, Ntodie M, Acquah E, Kyei S. A case of leukemic retinopathy mimicking common ischemic retinopathies. Clinical Case Reports.2016 Feb; 4 (2): 133.