Presentation of Chronic Myeloid Leukaemia in Accelerated Phase Presenting with Bilateral Leukemic Retinopathy – A Case Report

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Abstract

The leukemias are malignant neoplasms of the hematopoietic stem cells, characterized by diffuse replacement of the bone marrow by neoplastic cells. There are 3 stages in Chronic Myeloid Leukemia-chronic phase, accelerated phase and blast phase.

Ophthalmic manifestation can be classified into two categories: primary or direct leukemic infiltration and secondary or indirect involvement. Screening for ocular manifestations of leukemia, although not a routine practice, is important as they may antedate systemic disease or form an isolated focus of its relapse. The prevalence of ocular involvement in leukemic patients has been reported to be 9% to 90% in various studies. To highlight the important ophthalmic manifestation in patients of leukemias we are presenting a case report on chronic myeloid leukemia in accelerated phase of a young male patient referred from medicine OPD with chief complaints of blurring of vision in both eyes since 2 days.

Keywords: Leukemia; Ophthalmic manifestation; Chronic myeloid leukemia

Abbreviations: CML: Chronic Myelogenous Leukemias

Introduction

Chronic myeloid leukemia is a classic chronic myeloproliferative disorder. It is a clonal stem cell disorder characterized by acquisition of an oncogenic BCR/ABL fusion protein. (Usually result of a reciprocal translocation (9;22)q34;q11) [1]. Incidence of CML increase with age, with a peak incidence of 53 years [2]. It was the first disease (a) in which the term leukemia was utilized, (b) to be associated with a consistently recurring chromosomal abnormality, (c) to be recognized as the result of material reciprocally translocated from one chromosome to another, (d) to be direct result of a specific gene fusion (as a result of translocation), and (e) to have a therapy particularly targeted against the fusion protein.

Case presentation

Our patient is 35 years old patient who was first diagnosed with CML after presenting to the Medicine OPD. During his investigation it was found to have an elevated blood cell count (50 × 10^9/L). The Red blood corpuscles were microcytic and hypochromic. Platelets were reduced to 89,000 cells/mm³. Hb was 9.8 gm/dl. Differential leukocytes counts were-Myeloblast 12%, Promyelocytes 12%, Myelocytes 10%, Neutrophils 45%, Eosinophils 4%, Basophils 6%, Monocytes 0%. Bone marrow cytology suggestive of Myeloblast 16%, Promyeloblast 12%, Myelocyte 20%, Metamyelocytes 27%. Megakaryocytes were normal in morphology, M:E ratio=10:1 which was suggestive of chronic myeloid leukemia (Accelerated phase).

Diagnosis of accelerated phase of CML was made when Blasts 10-19% in the peripheral blood and/or bone marrow, Basophils ≥ 20% in the peripheral blood, Persistent thrombocytopenia, Increase in spleen size and white blood cell count despite therapy, Cytogenic evidence of clonal evolution (Diagnosis is made when one or more of the listed features is present) (Figures 1 and 2).

Figure 1: Peripheral blood smear of chronic myeloid leukemia in accelerated phase.
Figure 2: Bone marrow slide suggestive of chronic myeloid leukemia in accelerated phase.

Patient came to ophthalmology OPD with complaints of diminution of vision in both eyes since 2 days. On taking history it was a sudden painless in nature with Best corrected visual acuity of 20/60 both eyes for distant and near N [3] both eyes. Anterior segment on torch light and slit lamp examination case out to be within normal limit in both eyes. On dilating the patient pupil with eye drop tropicacyl plus fundus examination was suggestive of disc edema with vascular sheathing near the disc. Blood vessels were tortuous and dilated. Superficial haemorrhage and deep haemorrhage is present all over fundus in both eyes. Foveal reflex was absent in both eyes.

Patient was started on started on Tab Imatinib (400 mg/day) and Tab Hydroxyurea (500 mg/day). Imatinib is an ABL-specific tyrosine kinase inhibitor that inhibits proliferation of CML cell lines by inhibiting BCR-ABL kinase activity. Hydroxyurea is an inhibitor of ribonucleotide reductase, can lower blood count within 1 to 2 days. Due to loss of further follow up patient did not come to ophthalmology OPD (Figures 3 and 4).

Figure 3: Fundus of left eye showing leukemic retinopathy.

Figure 4: Fundus of right eye showing leukemic retinopathy.

Discussion

Knowledge of ocular involvement in leukemia is important because eye is the only site where the leukemic involvement of nerves and blood vessels can be directly observed [4]. This is so because the eye symptoms may be the initial mode of presentation of systemic illness, or the first manifestation of relapse after remission inducing chemotherapy [5]. A prompt recognition of the ocular manifestation and their importance as a sign of possible extra medullary disease is crucial if appropriate therapy is to be initiated [6]. This definitely has implications for ophthalmologist involved in the care of leukemic patients that mandatory and periodic eye check-ups at least every 6 months are a must despite the apparent non-involvement of the eye [10].

The early manifestations are venous dilatation and tortuosity. Haemorrhages may occur in all levels of the retina, usually the posterior pole, and may extend into the vitreous [11]. They may be round or flame shaped, and often has a white component known as roth spot. Cotton wool spots may be seen and often due to ischaemia from anemia, hyperviscosity, or leukemic infiltration [12,13]. Less
common manifestation includes microaneurysm which is probably related to increased blood viscosity from elevated white blood cell count.

Conclusion

We presented this case report to highlight the ocular manifestations of CML. The secondary involvement of retina is the most common eye changes in leukemia. Ocular involvement is more common in acute leukemias and myeloid leukemias. The retina is involved in leukemia more often than any other ocular tissue.

Although the ophthalmologist has a secondary role in treatment of leukemias, a prompt recognition of ocular manifestation is crucial because of poor prognosis associated with ocular involvement [14] and to identify possible extra medullary diseases [15,16]. All patients diagnosed with leukemia should be sent to ophthalmologist for proper examination. They should be kept in follow up after initiation of treatment of the disease. A periodic ophthalmic examination should be mandatory for all leukemic patients, as ocular changes are often picked up in asymptomatic patients.

Acknowledgement

Written consent was obtained from the patient for publication of the case report and any accompanying images.

References