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Posterior Segment Involvement as Initial Presentations of Patients Withchronic Myeloid Leukemia

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Chronic myeloid leukemia (CML) is a myeloproliferative neoplasm. Ophthalmologic manifestations are among the therapeutic challenges. Here we present two cases of CML (chronic phase) with posterior segment involvement as initial presentation, trying to shed light on this important type of presentation and view of the current literature.

Keywords: Chronic myeloid leukemia; ophthalmologic manifestations.

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1. INTRODUCTION

Chronic myeloid leukemia is a myeloproliferative neoplasm that can present in different ways; it varies from incidental finding in routine complete blood count to symptomatic presentations such as splenomegaly.

Ocular involvement in leukemia is considered rare [1,2]. First described by Liebreich in 1860. Among the uncommon presentations of CML are ophthalmologic manifestations, which can vary from incidental finding during eye examination to blurred vision and partial/total loss of vision.

The ophthalmic manifestations of CML are florid, with vascular changes such as retinal vein tortuosity or obstruction, flame-shaped hemorrhages, dot-and-blot hemorrhages, Roth spots, and even optic nerve edema.

Patients with ophthalmic manifestation of chronic myeloid leukemia (CML) have been reported to have lower 5-year survival than those without [3].

2. CASE PRESENTATION

We present two cases of CML (chronic phase) with ophthalmologic manifestations as initial presentation.

CASE 1:

A 54-year-old male, with no medical history,was admitted for asthenia and a painless rapidly progressive loss ofvisual acuity decreasing and blurring vision. The visual acuity was at counting fingers in both eyes, with a normal IOP and anterior segment examination.

-Fundus examination revealed the presence of bilateral papilledema, significant macular edema, whitecenteredhemorrhages and multiple hemorrhages with vascular tortuosity (Fig. 1).

OCT confirmed macular edema, FA highlighted hemorrhages, disk swelling with no signs of ischemia (Fig. 2).

A completebloodpictureshowed high white bloodcell (WBC) levels (Over 500 × 103/μL). The myelogramfounda blast rate lessthan 5% and bonemarrowexaminationwas suggestive of CML. Cytogeneticsconfirmed the presence of Philadelphia chromosome and RT-PCR found positive BCR ABL (92%), confirming the diagnosis of CML.

The patient wastreatedwithImatinib (600mg daily) and rehydratation.

Within a month, we had an improvement of visual acuity to 3/10 OU with regression of ocular signs. Patient is being followed-up.

CASE 2:

A 37-year-old women on followe up for breast carcinoma for 3 years on oralTamoxifen, presented with 20 days of progressive decrease in visual acuity, aggravated by a bilateral visual blindness with negative light perception.Physical examination found a splenomegaly.

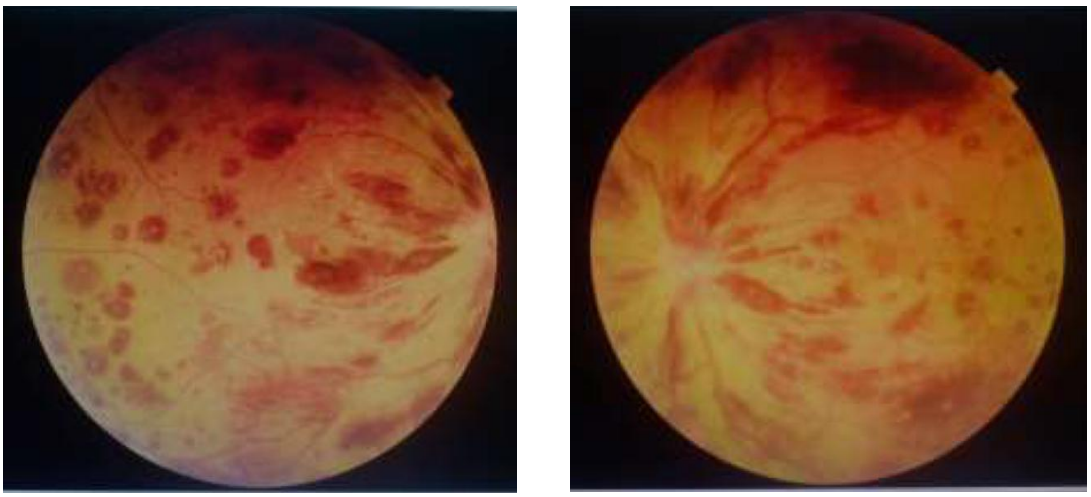

Fig. 1. Fundus photograph showing bilateral papilledema, macular edema with Roth's spot hemorrages and vascular tortuosity

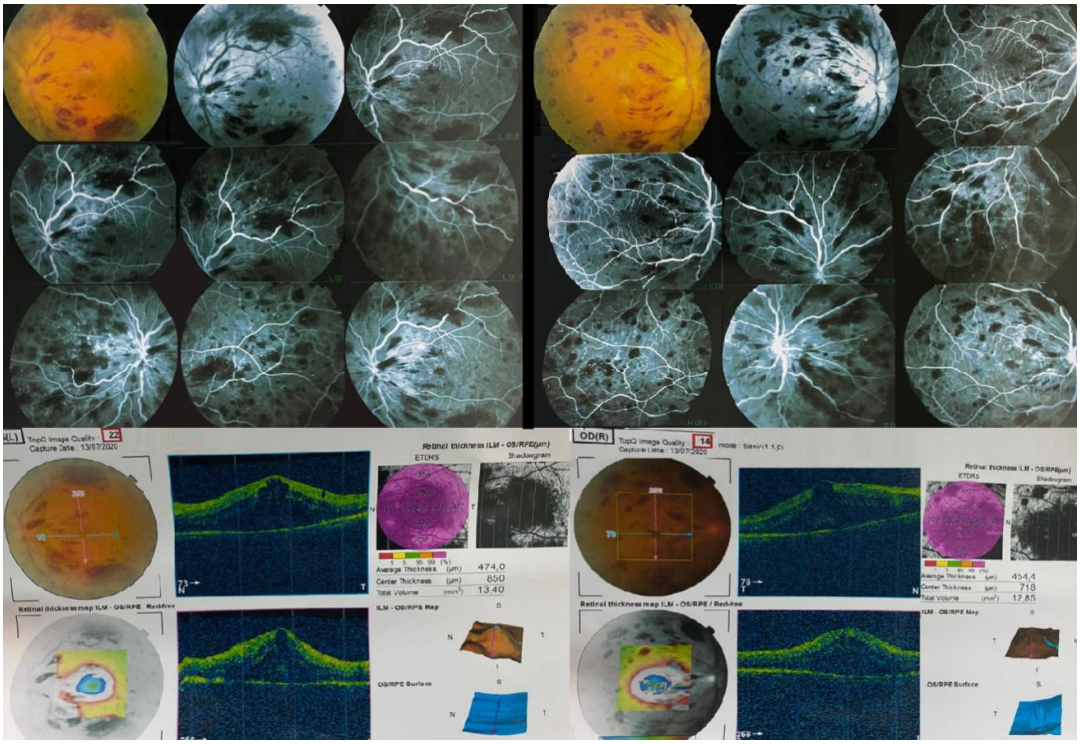

Fig. 2. Up: fluorescein angiography showing bilateral occlusion of the central retinal vein with no signs of ischemia. Down: optical coherence tomography confirmed the macular edema

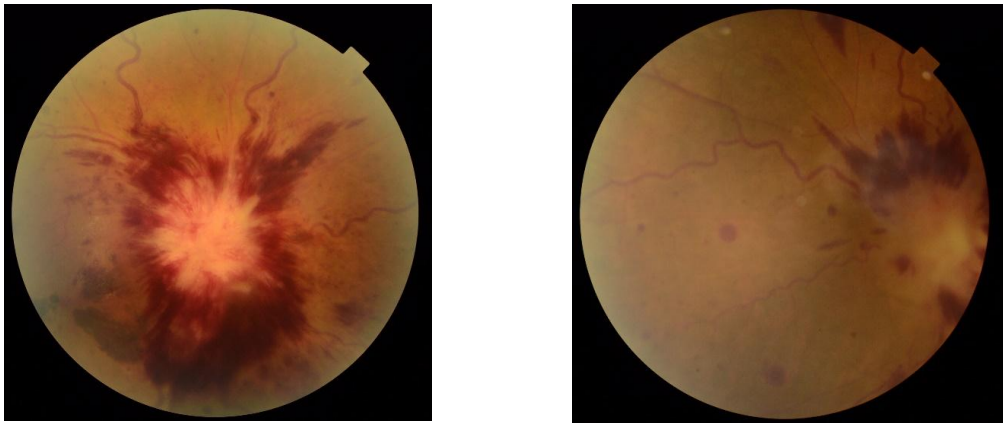

Fig. 3. Fundus images showing bilateral papilledema at the stage of optic atrophy and retinal hemorrhages

The anterior segment examination showed an abolished direct and consensual photo-motor reflex, the fundus examination showed bilateral papilledema at the stage of optic atrophy, retinal haemorrhages and tortuous veins (Fig. 3).

Magnetic resonance imaging (MRI) of the brain and orbits with and without contrast did not show signs of signal abnormalities of the supratentorial and subtentorial brain parenchyma as well as the absence of abnormalities on the diffusion sequence. Midline structures are without abnormality.The complete blood count CBC showed hyperleukocytosis at 250,000/mm3, anemia with hemoglobin at 9g/dl and thrombocytopenia with platelet count at 60,000/mm3.

The myelogramfounda blast rate lessthan 5%. Blood smear and bonemarrowexaminationwas suggestive of CML. Cytogeneticsconfirmed the presence of Philadelphia chromosome and RT-PCR found positive BCR ABL (87%), confirming the diagnosis of CML.

The patient wastreatedwithImatinib (600mg daily) and rehydratation but died short time after the beginning of the treatment.

3. DISCUSSION

Studies have shown that only 5%–10% of CML patients present with eye symptoms at initial diagnosis [4].

Ophthalmologic manifestations have a wide range of presentations and findings upon fundoscopy and eye examination. The optic nerve or retina might be affected.

Visual disturbance or blindness could be the first presentation of CML. It wasreportedthat 5-year survival of patients withophthalmic manifestation of CML was 21.4% compared to 45.7% of thosewithout [5].

An increased level of WBC, such as 300,000, will present with a picture of hyperviscosity.

Fundoscopy of these patients might reveal papilledema, venous obstruction, and hemorrhages [6] like in our two cases.

Optic nerve involvement typically leads to relatively rapid and potentially irreversible vision loss [7] like in our second case. Theoptic disc edema or pallor can be due to autoimmune, infectious, inflammatory, infiltrative, and medication induced etiologies. An MRI of the brain and orbit and a CSF study to look for leukemic cells can help guide urgent management [8, 9].

Patients with leukemic optic neuropathy LON may require intrathecal chemotherapy and orbital radiation, and a prompt oncology consultation is recommended for all cases.

Features of leukemic retinopathy include yellowish-white masses of variable size, multiple pre-retinal and intraretinal hemorrhages, cotton wool spots, perivascular sheathing, and neovascularization. Roth's spot hemorrhages may represent small areas of retinal leukemic infiltration [10]. Leukemicretinopathyis more common in the acute leukemias and isusually a poorprognosticindicator.

Leukemic infiltrates most often affect the choroid; however, they may present with a serous or exudative retinal detachment [11].

Leukemic infiltration of the eye is best treated with systemic chemotherapy appropriate for the type and stage of the leukemia. Of note, the various chemotherapeutic agents used to treat leukemia may cause ocular toxicity such as cataract development, cranial nerve palsies, optic atrophy, and intraocular inflammation [12,13].

4. CONCLUSION

These 2 cases illustratepeculiar and varied clinical features of chronic myéloid leukemia, where an Ophthalmologist'srole can be primordial. It is important to recognize early fundus changes in patients who do not present with the usual signs and symptoms of CML. A suspicious ocular finding should be followed up with appropriate systemic evaluation and workup, including a full blood count.

We recommend baseline fundoscopy and eye examination by an ophthalmologist for all newly diagnosed CML.

DISCLAIMER

The products used for this research are commonly and predominantly use products in our area of research and country. There is absolutely no conflict of interest between the authors and producers of the products because we do not intend to use these products as an avenue for any litigation but for the advancement of knowledge. Also, the research was not funded by the producing company rather it was funded by personal efforts of the authors.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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