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Sudden central vision loss in a young patient with chronic myeloid leukemia

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Introduction

Chronic myeloid leukemia (CML) is a myeloproliferative neoplasm characterized by a fusion of *ABL1* and *BCR* genes. It leads to the malignant transformation of cells with a myeloid phenotype [1]. In Western countries, the median age for a CML diagnosis is 56–57 [2, 3]. About half of CML patients are asymptomatic at diagnosis. In other patients, the most common symptoms are caused by splenomegaly and anemia. General symptoms such as weight loss, night sweats, and fatigue may also occur. In some individuals, especially those with white blood cell (WBC) counts exceeding 100 G/L, the clinical manifestation of CML is mainly attributed to leukostasis (confusion, drowsiness, hearing difficulties, dyspnea) [4]. The standard first-line CML treatment involves tyrosine kinase inhibitors (TKIs). Four TKIs, imatinib, dasatinib, nilotinib, and bosutinib, are registered and approved by the US Food and Drug Administration (FDA) and the European Medicines Agency (EMA) [5, 6]. Ophthalmic manifestations are relatively uncommon in CML, although some case reports have been described in the literature [7]. Ophthalmic examination may lead to a diagnosis of CML [8]. Potentially, any ocular structure may be involved. This case report illustrates the importance of the ophthalmologist recognizing various ocular signs during the treatment of CML patients.

Case report

A 20-year-old female with no significant medical history was referred to the hospital because of fatigue, night sweats, discomfort within the abdomen, and tinnitus. She also reported occasionally seeing spots, especially when standing up from being seated or prone. Complete blood count revealed a white blood cell count (WBC) of 564 G/L, hemoglobin 5.8 g/dL, and platelets 836 G/L, while peripheral blood smear was remarkable for immature granulocytes and blasts percentage (4%). Ultrasound examination revealed a greatly enlarged, immeasurable spleen, and a liver of the borderline size. Chest X-ray was unremarkable. Suspicion of CML was postulated, and the patient was admitted to the hematology department. On admission, cytogenetic and molecular tests to confirm the CML diagnosis were planned. Due to hyperleukocytosis, cytoreductive treatment with hydroxyurea 3 grams daily, allopurinol, and intravenous hydration were immediately started, while leukapheresis was started the next day. Additionally, two units of red cells were transfused.

Over the next two days, with the treatment with leukapheresis and hydroxyurea, continued gradual reduction of WBC and platelet counts was observed. The patient's condition remained stable, with no new symptoms. However, on day 3 after admission, she reported a rapid loss of central vision in her right eye. The patient's WBC on this day decreased to 392 G/L. An ophthalmological examination revealed consequences of leukostasis, including intraretinal hemorrhages, extremely tortuous retinal vessels, white-centered hemorrhages (Roth spots), cotton wool spots, macular hemorrhage in the right eye, and bilateral optic disc swelling (Fig. 1). Visual acuity was 5/25 in the right eye and 5/5 in the left eye. No indication for ophthalmological intervention

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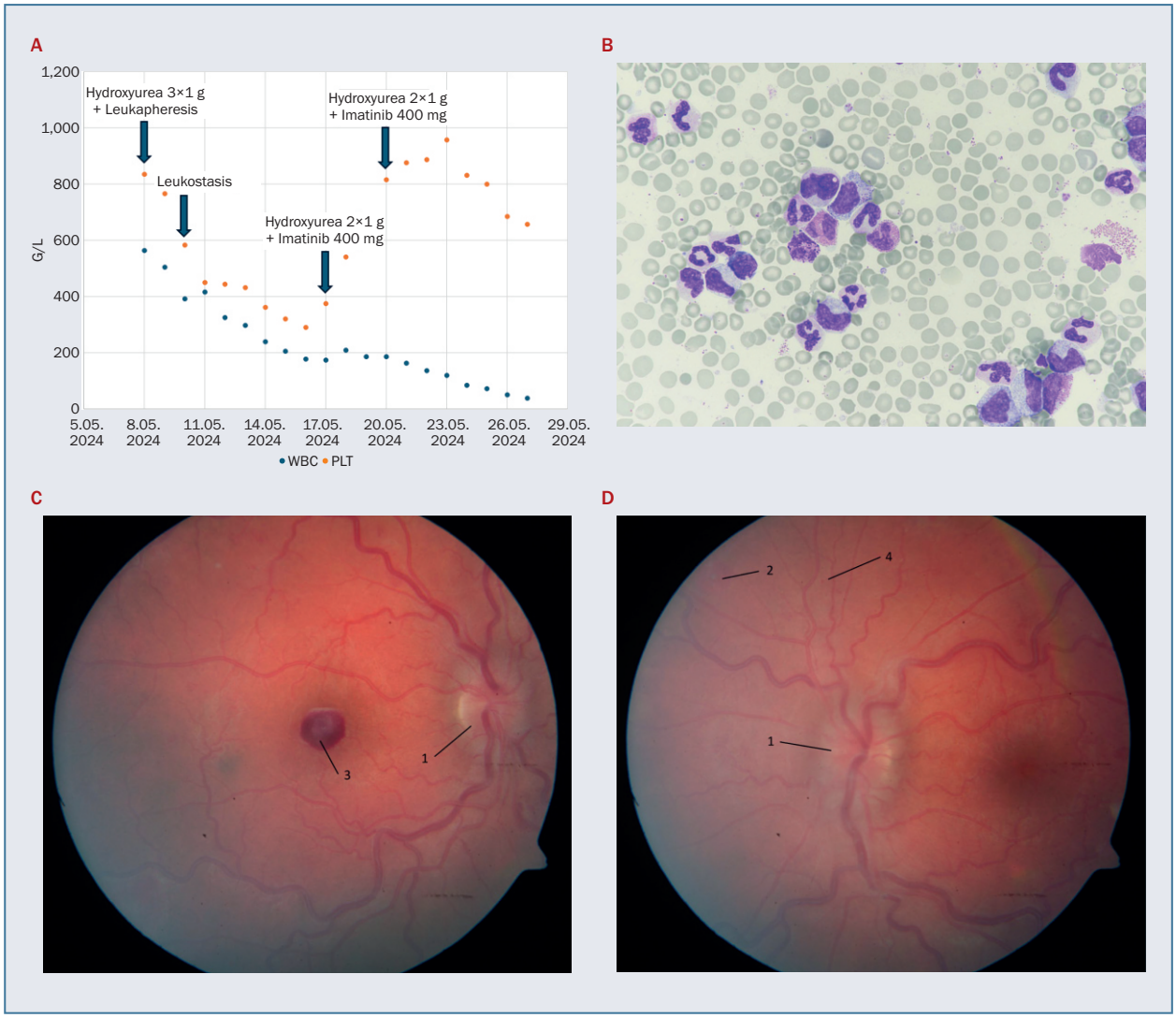

Figure 1. A. Plot shows WBC and PLT counts in timeline. Start days of various treatments and symptoms are indicated; **B.** Peripheral blood smear shows immature granulocytes typical for CML; **C.** Right eye fundoscopy shows extremely tortuous retinal vessels, optic disc swelling (1), and macular hemorrhage (3); **D.** Left eye fundoscopy shows extremely tortuous retinal vessels, intraretinal hemorrhages (4), white-centered hemorrhages (Roth spots) (2), optic disc swelling (1)

was stated. A CT scan showed no signs of bleeding or thrombosis, and in magnetic resonance imaging (MRI), a slightly increased amount of fluid was seen in the optic nerve sheaths. As treatment continued over the following days, an ophthalmologist regularly consulted the patient. Her central vision in the right eye gradually improved, but without complete resolution. On day 7 after admission, a positive result for the BCR/ABL1 fusion polymerase chain reaction test was obtained, and imatinib at 400 mg daily was commenced with a gradual decrease of hydroxyurea dose.

Subsequently, further results of bone marrow examinations, including smear and cytogenetics, confirmed the chronic phase of CML. Her general condition was good on day 34 after admission, and splenomegaly had completely resolved. WBC was 3.5 G/L and hemoglobin 9.2 g/dL. It was decided to discontinue hydroxyurea, and the patient has since continued treatment with imatinib.

Discussion

The most common ocular findings in CML patients are localized in the posterior segment, referring to the retina and choroid. In such patients, leukemic retinopathy consists of Roth spots, cotton wool spots, leukemic infiltrates, hemorrhages (preretinal, intraretinal, subretinal), dilated and tortuous vessels, and optic disc hyperemia. The condition develops mainly secondary to vascular stasis due to high WBC count [8, 9]. Optic nerve swelling and infiltration have also been reported and can cause severe loss of vision.

In our case, the patient reported occasionally seeing spots on admission, but it is worth underscoring that central vision loss started some days after the start of cytoreductive treatment and leukapheresis. In patients with very high leukocytosis, prompt ophthalmological consultation with dilated fundus examination should be performed even in the absence of symptoms in order to rule out ophthalmological manifestations of the disease [10]. In the above-mentioned case the vision loss resolved, but not completely, probably due to macular involvement and optic nerve swelling. In some cases, such visual impairment may be irreversible.

Article information and declarations

Ethics statement

Authors declare that informed consent for publications was not obtained, as published data does not allow for patient identification.

Authors' contributions

MJ — manuscript preparation, data collection, literature search; PK — manuscript preparation, data collection; MP-K — data collection, KJ — manuscript preparation, idea, critical review.

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Conflicts of interest

The authors declare no conflict of interest.

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Supplementary material

None.

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