

Hemorrhagic Chemosis in Acute Myeloid Leukemia: A Case Report

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Abstract

Purpose: Ocular manifestations of leukemia are common and can present at any time, resulting from either infiltration by leukemic cells or from effects of the disease and treatment. Despite the prevalence of ocular manifestations in patients with leukemia, hemorrhagic chemosis in pediatric patients with leukemia has not been well described.

Observations: We describe a patient with acute myeloid leukemia (AML) who developed hemorrhagic chemosis secondary to leukostasis, thrombocytopenia, and hyperhydration that was successfully surgically corrected after the completion of chemotherapy.

Conclusions: This case highlights the importance of an interdisciplinary approach to the care of patients with AML.

Keywords: Hemorrhagic chemosis; Acute myeloid leukemia; AML with ocular manifestations; AML and lid proptosis; AML and hemorrhagic chemosis

Introduction

Ocular manifestations of leukemia are an important and prevalent finding with a variety of involved structures and presentations. Ophthalmic involvement can occur in up to 90% of patients and can develop before, after, or concurrently with the hematologic manifestations of leukemia^[1,2]. These findings result from either direct infiltration of the orbital structures by leukemic cells or, more commonly, secondary to the course of disease and treatment^[1,3]. Although lesions most commonly affect the retina and choroid, virtually any ocular structure can be affected. Depending on the mechanism of involvement and the structure affected, orbital manifestations can present with symptoms including proptosis, lid edema, chemosis, or can be asymptomatic^[1,2]. In most cases, the ophthalmic manifestations of leukemia improve with chemotherapy or limited-field radiation; however, the presence of ocular involvement has been associated with a worse prognosis and higher risk of relapse compared to patients without ocular lesions^[1,3]. While both mechanisms contribute to a worse prognosis in leukemia patients with ocular involvement, lesions directly caused by ocular infiltration by leukemia cells are associated with a higher frequency of leukemic relapses compared to secondary lesions^[4]. Chemosis (conjunctival edema) is a presentation that often raises concern for direct ocular infiltration but can also occur as a secondary mechanism of the disease, especially in the setting of hemorrhage, a common hematologic complication of leukemia^[2,4]. We describe a case of severe, bilateral hemorrhagic chemosis in a pediatric patient with

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AML. We believe the hemorrhagic chemosis in this patient was secondary to hematologic manifestations of leukemia including leukostasis and thrombocytopenia; supportive care measures including aggressive hydration also contributed to worsening of his ocular disease.

Case Report

An 11 year old Caucasian male presented with hyperleukocytosis (white blood cell (WBC) count of 189,000/ μ L), anemia (hemoglobin of 6.1 g/dL), and severe thrombocytopenia (platelet count of 15,000 u/L). His prothrombin time (PT), activated partial thromboplastin time (aPTT), and fibrinogen were normal. He was diagnosed with acute myelomonocytic leukemia by flow cytometry; karyotype and FISH analyses both revealed inv(16) (p13.1;q22). Initial cerebrospinal fluid (CSF) analysis was positive for blasts, which cleared after three intrathecal treatments with cytarabine. He was treated up front with leukocytapheresis, followed by systemic AML chemotherapy and aggressive supportive care.

Although his initial eye exam was normal, with no lid edema or proptosis, after two days of hospitalization, he developed bilateral subconjunctival hemorrhages. [Figure 1A] Magnetic resonance imaging (MRI) of the brain did not demonstrate a myeloid sarcoma. By day 7, he had begun to develop severe hemorrhagic subconjunctival chemosis of both eyes. [Figure 1B] On ophthalmic exam, he was also noted to have intra-retinal hemorrhage. He was treated with artificial tears and Lacri-Lube, and every effort was made to maintain his platelet count $>50,000/\mu$ L. His severe subconjunctival chemosis persisted throughout his full course of chemotherapy, and worsened with thrombocytopenia and with the aggressive hydration associated with chemotherapy infusions. [Figure 1C] His vision remained normal.

Due to persistent chemosis, approximately 1 month after completion of chemotherapy, he underwent sequential tarsorrhaphy of both eyes, with excellent results. [Figure 1D-E] Biopsies taken at the time of surgery were negative for leukemia. To date, he remains in remission, with normal conjunctivae on examination, and no visual defects.

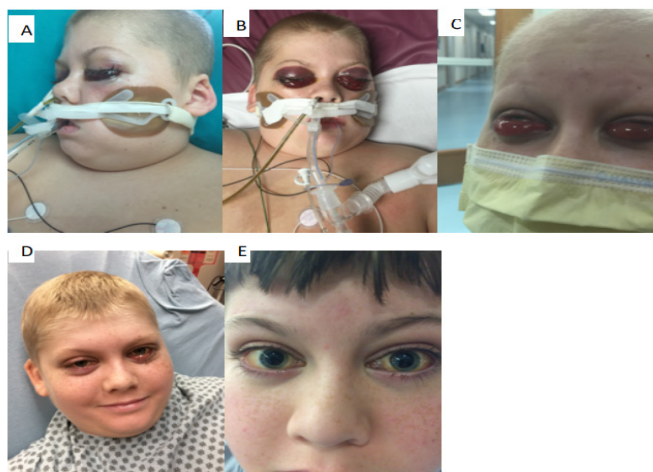


Figure 1: Progression before and after surgical intervention of hemorrhagic chemosis secondary to newly diagnosed AML.

A. 2 days after diagnosis of AML.

B. 1 week after diagnosis.

C. Prior to start of subsequent chemotherapy cycles.

D. Approximately 1 month after right eye tarsorrhaphy.

E. 1 month after left eye tarsorrhaphy.

Discussion

Ocular lesions are the third most common extramedullary location of acute leukemia and are four times more likely to be involved in acute leukemia than in all of the chronic leukemias and leukosarcomas combined, according to an autopsy study performed at UCLA of patients affected with leukemia or leukosarcoma^[4,5]. In addition, this study noted a bimodal distribution of patients with ocular manifestations of leukemia, peaking first in ages 11-20, then again in the late 60s to early 70s. In this study, hemorrhage in one or more structures of the eye seems to exclusively occur in acute leukemia (in all but 1 case)^[5].

Hemorrhage in an orbital structure is considered a secondary complication of the disease course rather than a manifestation of direct infiltration. Leukemic retinopathy, a well-described and common ocular manifestation, is a result of anemia, thrombocytopenia, and hyperviscosity leading to retinal changes, including hemorrhage^[1,4]. Subconjunctival hemorrhage, as seen in our patient, is another potential secondary complication of the abnormal hematologic parameters associated with leukemia. Hyperleukocytosis can lead to leukostasis and/or hyperviscosity and cause veins to become tortuous and dilated, and ultimately develop ischemia or hemorrhage^[2,4]. This was the likely mechanism of the ocular manifestations described in our patient which were subsequently worsened by thrombocytopenia and aggressive hydration. Initially there was concern for leukemic infiltration also contributing to our patient's presentation; however, biopsies showed no evidence of infiltration, further implicating the primary role of leukostasis as the mechanism for the development of this patient's hemorrhagic chemosis and proptosis.

Ocular involvement in leukemia is an important finding as it can be the presenting feature of disease, can indicate relapse, and has been shown to affect survival rates^[4]. A study by Ohkoshi and Tsiaras found that the 5-year survival was worse for patients with ocular manifestations than patients without eye abnormalities (21 vs. 46%). This negative effect on prognosis was attributed to the concomitant CNS involvement or bone marrow relapse that was seen in the majority of cases exhibiting ophthalmic involvement^[6]. Due to the possibility of ophthalmic lesions remaining asymptomatic, routine eye exams are an important component in the initial work-up of a newly diagnosed patient and in patients who have completed treatment as several studies found ocular manifestations can be one of the first signs of disease relapse.^[1]

Conclusions

We report a patient with AML who developed hemorrhagic chemosis after initial presentation secondary to complications of hematologic derangement, requiring surgical resection by tarsorrhaphy after completion of chemotherapy. Given the high incidence of ocular manifestations in AML, an eye exam by an ophthalmologist should perhaps be part of the initial physical exam and disease evaluation. In our patient, the symptoms worsened in the context of thrombocytopenia and leukostasis,

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as well as with hyperhydration as part of the supportive regimen for chemotherapy. This case highlights the importance of a multidisciplinary approach to the patient with AML, as evidenced by the preservation of this patient's eyesight and excellent cosmetic outcomes after surgical correction.

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