

Adnan AKSOY Murat ASLANKURT Lokman ASLAN Gökhan ÖZDEMİR

Kahramanmaraş Sütçü İmam Üniversitesi, Tıp Fakültesi, Göz Kliniği Anabilim Dalı, Kahramanmaraş, TÜRKİYE

**OLGU SUNUMU** 

F.Ü.Sağ.Bil.Tıp Derg. 2014; 28 (2): 77 - 79 http://www.fusabil.org

# Retina and Optic Disc Infiltration in a Young Patient with Acute Lenfoblastic Leukemia

We would like to draw attention to ocular involvement and regression with chemotherapy in a young patient with Acute Lenfoblastic Leukemia (ALL) in this case study. Sixteen years old female patient was admitted with loss of vision in right eye obtaining for one month. The patient had diagnosis of ALL for 4 years and and have been on remission for the last one year. Best corrected visual acuity (BCVA) on the right eye was counting fingers from two meters and 10/10 on the left eye. Ocular metastasis of ALL was considered as a result of investigations and anamnesis of patient. Systemic chemotherapy was started again and after a few days, visual acuity improved to 2/10 on snellen acuity.

Key Words: Acut Lymphoblastic Leukemia, retina, optic disc.

#### Akut Lenfoblastik Lösemi'li Genç Bir Hastada Retina ve Optik Disk Infiltrasyonu

Biz bu olgu çalışmasında kemoterapiyle regrese olmuş genç bir Akut Lenfoblastik Lösemi (ALL) hastasında oküler tutuluma dikkat çekmek istedik. 16 yaşındaki bayan hasta bir aydır sağ gözde görme azlığı şikayetiyle polikliniğe başvurdu. Hasta, ALL tanısını 4 yıl önce almıştı ve son bir yıldır remisyondaydı. Sağ gözde görme 2 metreden parmak sayma seviyesindeydi. Sol gözün görmesi tamdı. Göz bulguları yapılan araştırmalar ve hasta hikayesiyle ALL metastazı olarak değerlendirildi. Tekrar başlanan kemoterapi tedavisinden birkaç gün sonra görme keskinliği snellen eşeliyle 2/10 olarak tesbit edildi.

Anahtar Kelimeler. Akut lenfoblastik lösemi, retina, optik disk.

#### Introduction

Leukemia develops by acquiring malignant character of hematopoietic cells. The most common childhood malignancies are acute leukemias and 75-85% of this group represents as ALL. The prevalence of ocular involvement in patients with leukemia is 9-90% in the literature (1-3). Ocular involvement has been reported at the beginning of the disease in some studies (1, 2), at various stages of the disease in the others (4, 5). It should be kept in mind that visual acuity decrease in patients with ALL may be caused by leukemic retinal and optic disc infiltration, confused with other retinal diseases and may regress with chemotherapy.

### Case Report

A sixteen years old female was admitted to our clinic with the complaint of decreased vision in her right eye obtaining for one month. The patient was diagnosed as ALL four years ago. After chemotherapy, remission was achieved one year ago. Best corrected visual acuity (BCVA) was counting fingers from two meters in the right eye and 10/10 in the left eye. Intraocular pressure was 16mmHg in the right eye and 14 mmHg in the left eye. Biomicroscopic examination of the right eye revealed a normal anterior segment and vitreous. Fundus examination of the right eye revealed approximately 3-disc sized, elevated white membranous structure on the optic disc (Figure 1). Widespread leukemic infiltration in the retina and optic disc with vascular dilatation were determined. The left eye was normal.

In the examination of viral infection for etiology, EBV VCA-IgM, CMV-IgG and CMV-IgM, rubella IgG were positive but the presence of two seropositivity were considered as cross reaction. There were atypical lymphoid cells in the cytological evaluation of Cerebrospinal Fluid (CSF). Patient's orbital Magnetic Resonance Imaging (MRI) and Computed Tomography (CT) revealed as normal. Optic Coherence Tomography (OCT) showed serous retinal detachment in the right eye.

# **Geliş Tarihi**: 03.04.2014 **Kabul Tarihi**: 13.06.2014

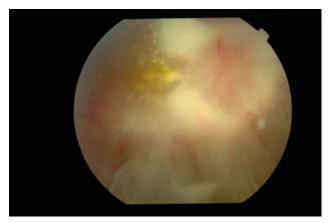
### Yazışma Adresi Correspondence

#### **Adnan AKSOY**

Kahramanmaraş Sütçü İmam Üniversitesi, Tıp Fakültesi, Göz Kliniği Anabilim Dalı, Kahramanmaraş-TÜRKİYE

## dradnanaksoy@hotmail.com

The patient was regarded as early isolated Cenral Nervous System (CNS) relaps ALL and started therapy according to the ALL Rez BFM protocol at the pediatric Dexamethasone. hematology clinic. vincristine, asparaginase, methotrexate, cytorabine, ifosfamide, daunorubicine and intrathecal therapy were given. Posttreatment BCVA of the right eye was 2/10 and BCVA of left eye was 10/10. Bilateral biomicroscopic examination evaluated as normal. Intraocular pressure was measured as 16mmHg on the sheathing at the nasal side by the final examination. Resorbed infiltration at the final examination (Figure 2). OCT revealed folding on epiretinal surface (Figure 3).



**Figure 1.** Fundus examination of the right eye revealed approximately 3-disc sized, elevated white membranous structure on the optic disc

#### Discussion

Acute lenfoblastic leukemia is an important component of hematopoietic system malignancy that effect young population. Since 1860 Liebreich's first described the leukemic retinopathy form, it has shown that almost all ocular structures were invaded in acute and chronic leukemia (1, 2). Ophthalmopathy is common in leukemia especially during periods of active disease in clinical and hematological period<sup>6</sup>. In postmortem autopsy studies, ocular involvement was found in 82% of acute leukemias and in 75% of chronic leukemia (3). This ratio alters between 9-90% on clinical series (2). Many findings were reported such as infiltration of choroid, leukemic retinopathy, leukemic cells in the anterior chamber and orbital involvement in patients with leukemia (2).

Involvement of eye and adnexia in patients with leukemia may be in a variety such as secondary hemorrhage and infarctions due to anemia or hiperviscosity (6).

Although some patients are symptomatic with respect to ocular findings, the majority of patients are asymptomatic. Ocular symptoms of leukemia are decreased visual acuity, leukemic cell infiltration of vitreous and sudden unilateral loss of vision (6).

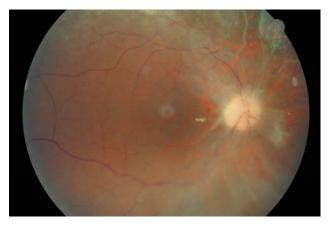


Figure 2. Resorbed infiltration at the final examination

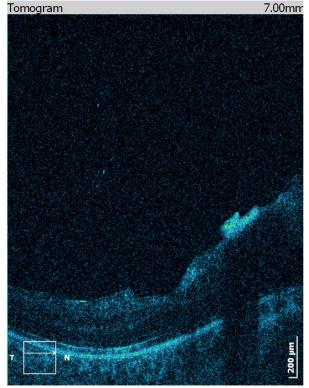


Figure 3. OCT revealed folding on epiretinal surface

In patients of leukemia, the most common relevant ocular tissue is retina. One of the first findings on fundoscopic examination is dilatation and tortuosity of the veins. Depending on decreased red cell count and increased white cell count, more yellowish appearance in both arteries and veins and retinal vascular sheathing can be seen (6). Retinal hemorrhages can be seen at every level of the retina. In addition, it was white centred hemorrhage and it included leukemic cells and debris, fibrin, platelet aggregates. Less common retinal findings like capillary microaneurysms (7), peripheral retinal neovascularization (8, 9) and leukemic epitheliopathy tend to be peripherally located. Another finding in leukemia is retinal cotton wool spots. Ischemia may be

caused by anemia, hyperviscosity or leukemic infiltration (6, 10, 11).

Microaneurysms in leukemia may be due to an elevated white blood cell count, in some cases leukocytosis and microaneurysms has not been determined. Increased blood viscosity due to an increased number of white blood cells leads to neovascular activity response and blockage of the terminal arterioles. This is thought to be responsible for the peripheral retinal neovascularization (12).

As a result of leukemia treatment, survival rate has increased in recent years. Therefore the CNS involvement is more common (3, 12). Optic nerve involvement may be a diffuse involvement due to CNS invasion. CNS is infiltrated and inracranial pressure (ICP) increases and causes papilledema. Direct infiltration of leukemic cells in the optic nerve can be seen without

#### References

- Guyer DR, Schachat AP, Vitale S, et al. Leukemic retinopathy. Relationship between fundus lesions and hematologic parameters at diagnosis. Ophthalmology 1989; 96: 860-862.
- Schachat AP, Markowitz JA, Guyer DR, et al. Ophthalmic manifestations of leukemia. Arch Ophthalmol 1989; 107: 697-700.
- 3. Kincaid MC, Green WR. Ocular and orbital involvement in leukemia. Surv Ophthalmol 1983; 27: 211-232.
- Ridgway EW, Jaffe N, Walton DS. Leukemic ophthalmopathy in children. Cancer 1976; 38: 1744-1749.
- Ohkoshi K, Tsiaras WG. Prognostic importance of ophthalmic manifestations in childhood leukaemia. Br J Ophthalmol 1992; 76: 651-655.
- Clayman HM, Flynn JT, Koch K, et al. Retinal pigment epithelial abnormalities in leukemic disease. Am J Ophthalmol 1972; 74: 416-419.

intra cranial pressure elevation (3). Eye findings of CNS involvement of leukemia can lead to diplopia, blurred vision, or involvement of extraoculer muscle and papilledema, Nikaido et al. reported optic nerve invasion in acute leukemia 13% (12). Optic nerve involvement is often associated with CNS or bone marrow involvement in relapses. First relapse may be in the optic nerve when the patient was in remission (4). With total remission retinal findings were cured and vision was increased in our case.

Leukemic infiltration in the OCT findings provides unique images. In our case, OCT findings were consistent with retinal invasion.

As a result, acute relapse may be seen in retina initially. With increased treatment modalities, ocular involvement is becoming increasingly common; with prompt treatment salvage is possible.

- Jampol LM, Goldberg MF, Busse B. Peripheral retinal microaneurysms in chronic leukemia. Am J Ophthalmol 1975; 80: 242-248.
- Morse PH, McCready JL. Peripheral retinal neovascularization in chronic myelocytic leukemia. Am J Ophthalmol 1971; 72: 975-978.
- Wheatcroft S, Watts P, McAllister J. Leopard spot retina. Eye (Lond) 1993; 7: 189-190.
- Soylu M, Tanyeli A, Ozdemir N, Eroğlu A, Ersöz TR. Ocular involvement in childhood leukemias. Turk J Pediatr 1994; 36: 35-41.
- Clayman HM, Flynn JT, Koch K, et al. Retinal pigment epithelial abnormalities in leukemic disease. Am J Ophthalmol 1972; 74: 416-419.
- 12. Nikaido H, Mishima H, Ono H, et al. Leukemic involvement of the optic nerve. Am J Ophthalmol 1988; 105: 294-298.
- 13. Christmas NJ, Mead MD, Richardson EP, et al. Secondary optic nerve tumors. Surv Ophthalmol 1991; 36: 196-206.