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Case Report

# Lymphocytic leukemia presenting as acute Vogt–Koyanagi–Harada disease

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## Abstract

Leukemia commonly involves eyes and adnexae. It is unusual, however for leukemia to present with visual complaints. There are only rare case reports of a leukemic patient presenting with bilateral exudative retinal detachment. This report describes a case of bilateral exudative retinal detachment associated with prodromal symptoms simulating the presentation of acute Vogt–Koyanagi–Harada disease that was eventually diagnosed as acute lymphocytic leukemia. There was rapid settling of the exudative detachment and improvement in vision when the patient received chemotherapy. Bilateral exudative retinal detachment associated with neurologic and auditory abnormalities may be a presenting sign of acute lymphocytic leukemia in an otherwise healthy young adult. Clinicians should be aware of the possibility of leukemia in such patients. A simple blood investigation such as complete blood profile confirms the diagnosis.

**Keywords**: Exudative retinal detachment, Acute lymphocytic leukemia, Vogt–Koyanagi–Harada disease


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## Introduction

Retinal manifestations in patients with acute leukemia include large spectrum of small vessel disease, ranging from mild ischemia with scattered cotton wool spots, and few retinal hemorrhages (round or flame shaped, often with a white component, which consists of leukemic cells and debris, platelet fibrin aggregates, or septic emboli) to proliferative retinopathy.[1]

Exudative retinal detachment, a very rare ocular finding in leukemia, has only been described in a few cases.[2–6] This report describes a case of acute lymphocytic leukemia presenting with bilateral exudative retinal detachment associated with prodromal symptoms of neurologic and auditory abnormalities simulating the presentation of acute stage Vogt–Koyanagi–Harada (VKH) disease.

## Case report

A previously healthy 39-year-old female presented with gradual visual loss in both eyes over 2 weeks. Her best corrected visual acuity (BCVAs) were 20/100 and 20/50 in the right and left eyes, respectively. Her medical history was obvious for intermittent headache, neck stiffness and tinnitus for 3 weeks prior to presentation. There was no history of any systemic disease, steroid intake, ocular trauma or surgery. There was no anterior chamber reaction or vitreous cells in either eye. Fundus examination showed multifocal bilateral exudative retinal detachment in both eyes (Fig. 1). Optical coherence tomography (OCT) revealed macular subretinal serous fluids in both eyes, with foveal thickness of 551 μm in the right eye and 423 μm in the left eye (Fig. 2). Fluorescence angiography (FA) in the early phase showed delayed choroidal filling and hypofluorescence in the macula,

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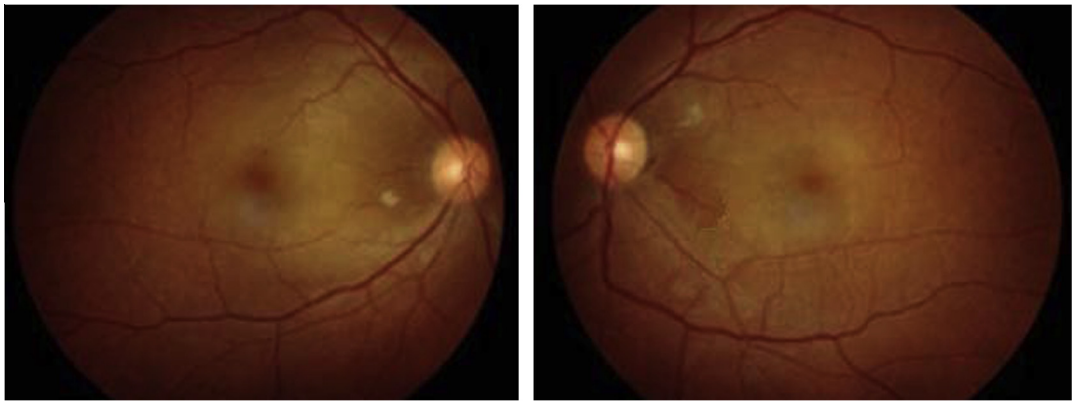

**Figure 1.** Initial presentation. Fundus photo of both eyes showing exudative retinal detachment.

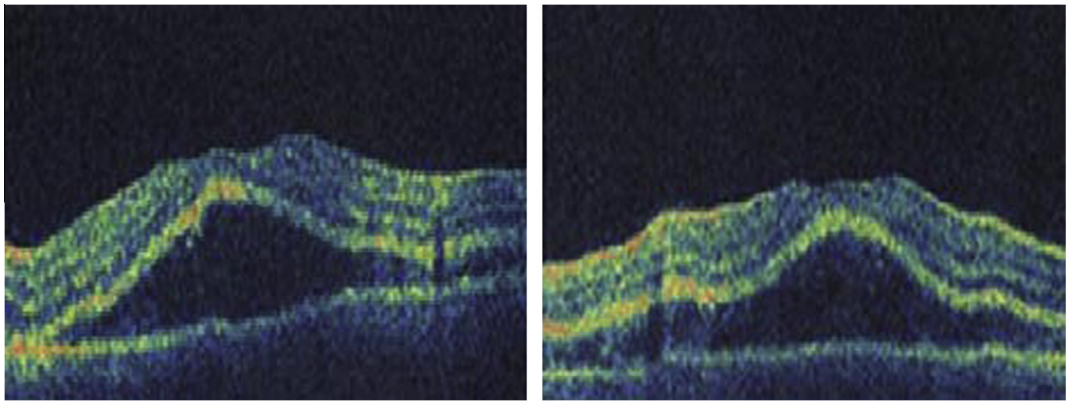

**Figure 2.** Optical coherence tomography of the macula in both eyes showed large amount of subretinal exudative fluid in the posterior pole. No subretinal septa were observed.

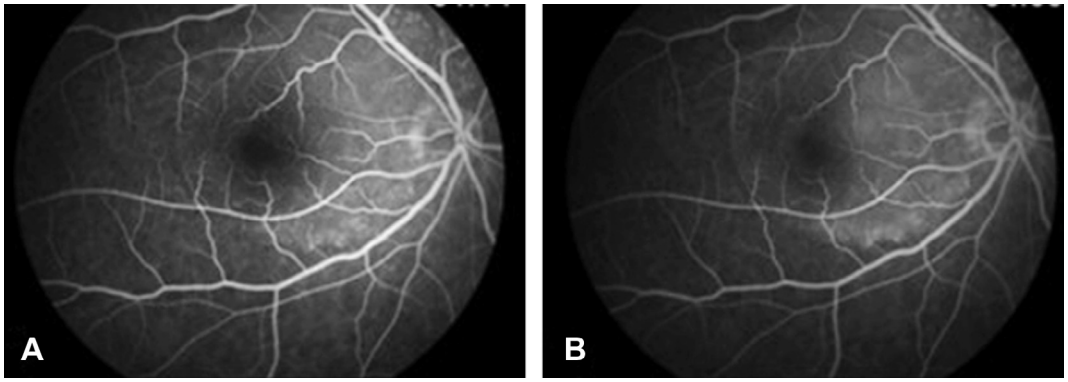


**Figure 3.** Fluorescence angiography (FA) of the right eye. (A) Mid-phase FA showed patchy choroidal filling and multiple punctate hyperfluorescent lesions. (B) Late-phase FA showed diffuse leakage and unilobular fluorescence pooling.

followed by mid-phase punctuate hyperfluorescence at the level of the retinal pigment epithelium (Fig. 3A). Late phase FA was apparent for diffuse late leakage and fluorescence staining in the area of exudative retinal detachment (Fig. 3B).

A clinical diagnosis of incomplete type VKH was considered. However, systemic evaluation revealed increased white blood cell counts (210,000/μL), with an abnormally increased percentage of blast cells (86%). Bone marrow examination confirmed the diagnosis of precursor B cell lymphoblastic leukemia. She received induction chemotherapy of vincristine, prednisolone, daunorubicin, L-asparaginase (VPDL) by the oncologist.

Two weeks later, her BCVAs were 20/25 in both eyes. The serous retinal detachments were nearly absorbed in both eyes (Fig. 4).

## Discussion

Visual loss after choriocapillary occlusion in leukemia was first reported by Zimmerman in 1964.[2] Beyond that,

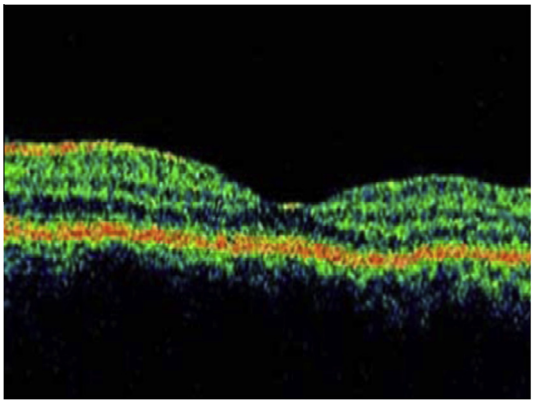

**Figure 4.** Two weeks after chemotherapy. Optical coherence tomography showed nearly absorbed serous retinal detachments of right eye.

exudative retinal detachment in leukemia has been reported in only a few cases world-wide, as a presenting sign of the disease or the first sign of relapse.[2–6] This case showed characteristic prodromal symptoms of neurologic and auditory abnormalities. The presence of such symptoms preceding bilateral exudative retinal detachment is a typical feature of VKH disease, fulfilling the diagnostic criteria of incomplete VKH disease as long as there is no underlying systemic disease.[7]

Bilateral exudative retinal detachment in a young individual can be due to central serous chorioretinopathy (CSC), however, this patient did not show any features of CSC leaks on FA. She did not report any risk factor for CSC such as a history of steroid usage. Other causes for exudative retinal detachment such as renal or cardiac dysfunction have been ruled out by systemic investigations.

The mechanism of serous retinal detachment in leukemia has been thought to be choroidal ischemia and secondary retinal pigment epithelial dysfunction.[3–6] The choroid is the most frequently involved ocular tissue in leukemia. Leukemic cell infiltration or hematologic disturbances may cause partial occlusion of the choriocapillaries and delay of choroidal circulation.[8] Secondary dysfunction of the Bruch's membrane and retinal pigment epithelium may ultimately develop into serous retinal detachments of the macula.[3,9] Ocular histologic findings by autopsy in leukemic patients have documented the features of choroidal thickening and leukemic cell infiltration that lead to choroidal ischemia.[8] Despite no histologic evidence, the fact that systemic chemotherapy induced a rapid remission of exudative retinal detachment, consistent with previous reports, suggests leukemic cell infiltration as the underlying pathology.[3,4,6]

Prodromal symptoms of headache, neck stiffness and tinnitus, such as in this case, followed by bilateral serous retinal detachment, may misdirect to the clinical diagnosis of incomplete type VKH disease.[7] However, several features are helpful in making the differential diagnosis. First, anterior uveitis was absent in this case, which is frequently seen in VKH disease.[10] Second, FA findings of mid-phase multiple hyperfluorescent dots were clustered along the boundaries of the detached retina, sparing the macula; in VKH disease such dots are usually distributed in the posterior pole.[11] Third, late-phase dye pooling in FA showed a unilobular CSC-like feature, different from the multi-lobulated pooling seen in VKH disease.[12] Fourth, OCT did not reveal any of the subretinal septa generally observed in VKH disease.[11] The prodromal symptoms that misdirected the diagnosis may have been caused by leukemic infiltration into the cerebrospinal fluid and microvasculopathy due to hyperleukocytosis.[10]

Leukemia may present when there is a sudden visual loss due to exudative retinal detachment, and the ophthalmologist may be the first clinician to encounter such patients. A thorough systemic evaluation should be performed in patients presenting with bilateral exudative retinal detachment, especially in those who report certain systemic symptoms.

## Conflict of interest

The authors declared that there is no conflict of interest.

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