

Acute Lymphoblastic Leukemia Masquerading as Vogt-koyanagi-harada Disease

Ana Arce (✉ canadarce@gmail.com)

Hospital Galdakao-Usansolo <https://orcid.org/0000-0002-1779-2169>

Naiara Relaño

Hospital Universitari Sant Joan de Reus

Joseba Artaraz

Hospital Universitario Cruces

Alex Fonollosa

Hospital Universitario Cruces

Brief report

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Abstract

Purpose: To report a case of Acute Lymphoblastic Leukemia masquerading as Vogt-Koyanagi-Harada disease.

Case Report: A 61-year-old man presented with decreased visual acuity in both eyes. Optical coherence tomography showed bilateral exudative retinal detachment and fluorescein angiography pinpoint leakage. The patient was diagnosed with Vogt-Koyanagi-Harada disease. Blood analysis revealed increased percentage of blast cells (69%) and bone marrow biopsy confirmed the diagnosis of acute B cell lymphoblastic leukemia. He received chemotherapy treatment with multiple drugs including asparaginase, which caused hepatic disease and patient's death despite ocular findings improvement.

Conclusions: Acute lymphoblastic leukemia may present as a bilateral exudative retinal detachment and can mimic VKH. Therefore, clinicians should take into consideration the possibility of leukemia in such patients. This fact stresses out the importance of multidisciplinary management of uveitis conditions.

Introduction

Leukemias are a group of haematopoietic stem cells malignancies involving abnormal increase of white blood cells. We can classify them depending on the line of proliferation into lymphoid or myeloid leukemia and between acute and chronic type. Ocular involvement in leukemias has been reported in a large percentage of cases, between 9 and 90% depending on the study. Any ocular structure might be involved and the infiltration of the eye is the third most frequent extramedullar location after meningeal and testicular involvement. Although findings diverge from retinal haemorrhages and infiltrates to optic nerve or orbital infiltration, exudative retinal detachment is a very rare ocular finding in leukemia and has only been described in a few cases.

In this paper we report a case of acute lymphocytic leukemia presenting with bilateral exudative retinal detachment simulating probable acute Vogt-Koyanagi-Harada (VKH) disease.

Case Report

A 61-year-old man presented with gradual visual loss in both eyes over 1 week. The decline in left eye visual acuity had begun accompanied by flashes. He had no medical history of previous ocular trauma or surgery, and the clinical interview did not reveal any headache or tinnitus symptoms. He suffered from arterial hypertension and chronic rhinitis, and had recently been taking Cetirizine and inhaled corticosteroids for this reason. His best corrected visual acuities were 20/50 and 20/40 in the right and left eyes, respectively. There was no anterior chamber reaction or vitreous cells in either eye. Intraocular pressure was 12 mmHg both eyes. Fundus examination and Optical coherence tomography (OCT) showed bilateral exudative retinal detachment in both eyes, and 2 cotton wool spots in the right eye (Figs. 1 & 2).

Fluorescence angiography (FA) showed multiple hyperfluorescent pinpoint leakage in posterior pole and around optic disc without disc leakage, and diffuse hyperfluorescence in the area of cotton wool spots in the right eye (Fig. 3).

Although clinical diagnosis of bilateral central serous chorioretinopathy was first considered, incomplete Vogt-Koyanagi-Harada disease was finally assumed. The patient received an intravenous 500 mg methylprednisolone pulse and a complete blood analysis was sought. The following day the blood analysis results were received and showed decreased platelet counts (10,000/ μ L), normal white blood cell counts (15,000/ μ L) with an abnormally increased percentage of blast cells (69%) presenting with cup-like nuclear invagination. Hence, the patient was admitted in the hematology department where a bone marrow biopsy revealed presence of 91% of blast cells, some of them with their nucleus indented, and confirmed the diagnosis of acute B cell lymphoblastic leukemia. The patient received induction chemotherapy consisting of vincristine, prednisone, daunorubicin, asparaginase and rituximab. One week later, his best corrected visual acuities were 20/63 and 20/50, serous retinal detachments were nearly absorbed (Fig. 4), but newly developed cotton wool spots near the optic disc appeared. Despite blood cell counts improved after chemotherapy, the patient died of hepatic veno-occlusive disease 2 months after starting treatment which was attributed to asparaginase.

Discussion

In leukemic patients, visual loss was first described by Zimmerman in 1964 (1).

Beyond that, there are few reports described in the literature of a leukemic patient debuting with bilateral exudative retinal detachment similar to our case (1–12).

The typical ocular manifestations of leukemia are most commonly due to direct tissue infiltration with leukemic cells, but other hematologic mechanisms such as anemia and hyperviscosity may play a role. The mechanism behind serous retinal detachments associated with leukemia is less well understood. Proposed mechanisms include abnormal choroidal perfusion and/or damage to the outer blood-retinal barrier, along with neoplastic choroidal infiltration, localized choroidal hypoxia, or alterations of local oncotic and/or hydrostatic forces. Additionally, choroidal hypoxia may lead to dysfunction of retinal pigment epithelium with subsequent accumulation of subretinal fluid. This may appear on fluorescein angiography as pinpoint areas of hyperfluorescence in the early phase (13). Our patient presented Funduscopy, OCT and FA findings that resembled a VKH syndrome. However, assessing the clinical examinations images retrospectively some features were not typical of VKH. Firstly, prodromal symptoms of headache and tinnitus and anterior uveitis were absent in our case, which are frequently seen in VKH disease (14). Additionally, FAG did not show hyperfluorescence of optic disks, a typical finding of VKH. Cotton wool spots were observed in our patient, an abnormality that has not been described in VKH. Moreover OCT did not reveal bacillary detachments, usually observed in VKH disease (15).

Masquerade syndromes are ocular clinical pictures that resemble uveitic diseases. A typical example is vitreoretinal lymphoma, which may be associated to central nervous system lymphoma in a high

proportion of patients (16–17).

Our patient presented with signs of VKH syndrome: exudative bilateral retinal detachments and pint point leakage in FAG. However, as mentioned above, other signs were lacking.

When managing atypical cases of uveitis masquerade syndromes have to be considered. A proper diagnosis by the ophthalmologist is crucial in order to initiate a prompt therapy for these frequently fatal diseases.

Abbreviations

VKH: Vogt-Koyanagi-Harada

OCT: Optical Coherence Tomography

FA: Fluorescence angiography

Declarations

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Author information

Affiliations

Ana Arce

- Ophthalmology department, Hospital Galdakao, University of the Basque Country, Spain.

Naiara Relaño

- Ophthalmology department, Hospital Universitari Sant Joan de Reus, Spain.

Alex Fonollosa Calduch:

- Begiker-Ophthalmology Research Group, Ophthalmology Department, BioCruces Health Research Institute, Cruces Hospital, University of the Basque Country, Spain.
- Instituto Oftalmológico Bilbao, Spain.

Joseba Artaraz Beobide:

- Begiker-Ophthalmology Research Group, Ophthalmology Department, BioCruces Health Research Institute, Cruces Hospital, University of the Basque Country, Spain.

Contributions

Naiara Relaño, Joseba Artaraz and Alex Fonollosa examined the patient and interpreted imaging. Alex Fonollosa drafted the manuscript. All authors read and approved the final version of the manuscript.

Corresponding author

Correspondence to Alex Fonollosa.

Ethics declarations

Ethics approval

The study adhered to the tenets of Declaration of Helsinki. Case reports involving a single clinical case do not require ethics approval by the Ottawa Health Science Network Research Ethics Board.

Consent for publication

Consent for publication was obtained from the subject.

Competing interests

The authors declare no competing interests.

Availability of data and materials

Not applicable.

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Figures



Figure 1

Fundus examination shows bilateral exudative retinal detachment in both eyes and two cotton wool spots in the right eye (green arrows).



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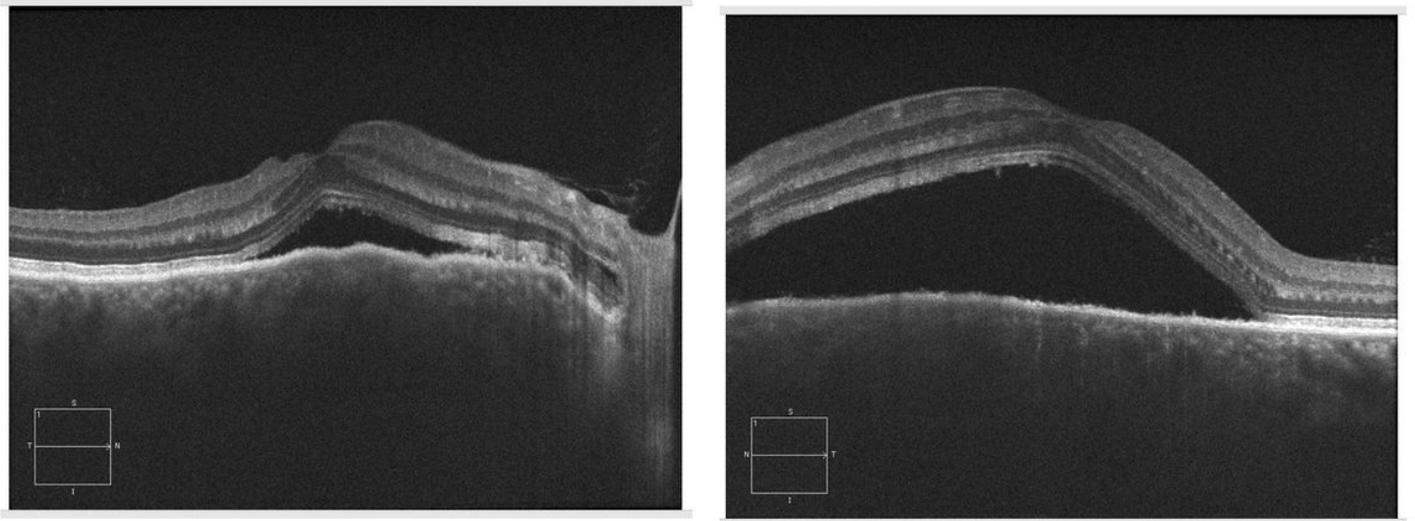


Figure 2

OCT imaging in both eyes showing exudative retinal detachment. The fluid appears greater in the left eye compared to right eye.

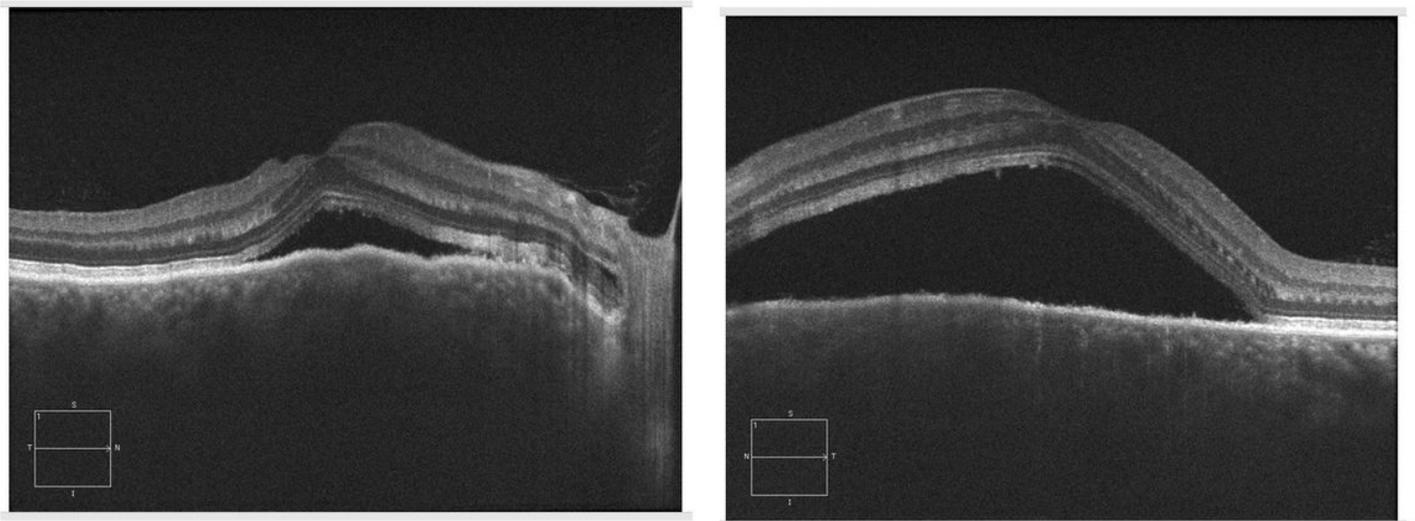


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OCT imaging in both eyes showing exudative retinal detachment. The fluid appears greater in the left eye compared to right eye.

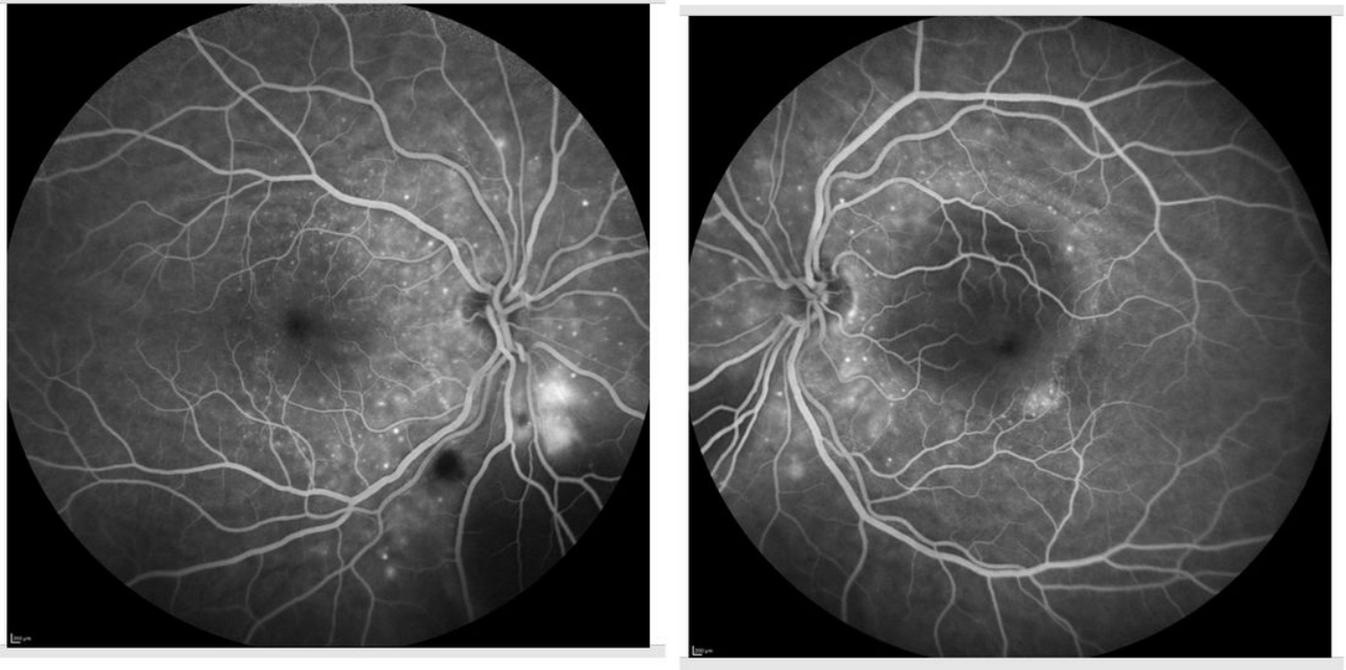


Figure 3

Fluorescein angiography shows multiple hyperfluorescent pinpoint leakage in posterior pole and around optic disc without disc leakage, and diffuse hyperfluorescence in the area of cotton wool spots in the right eye.

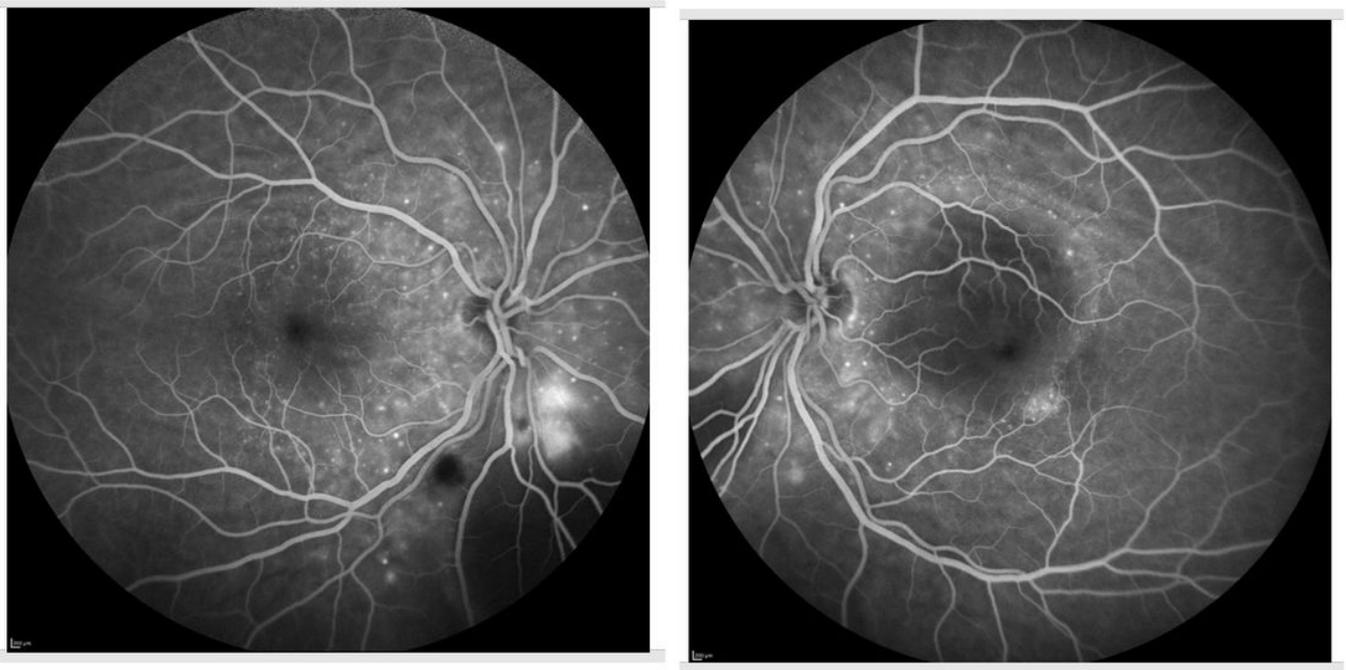


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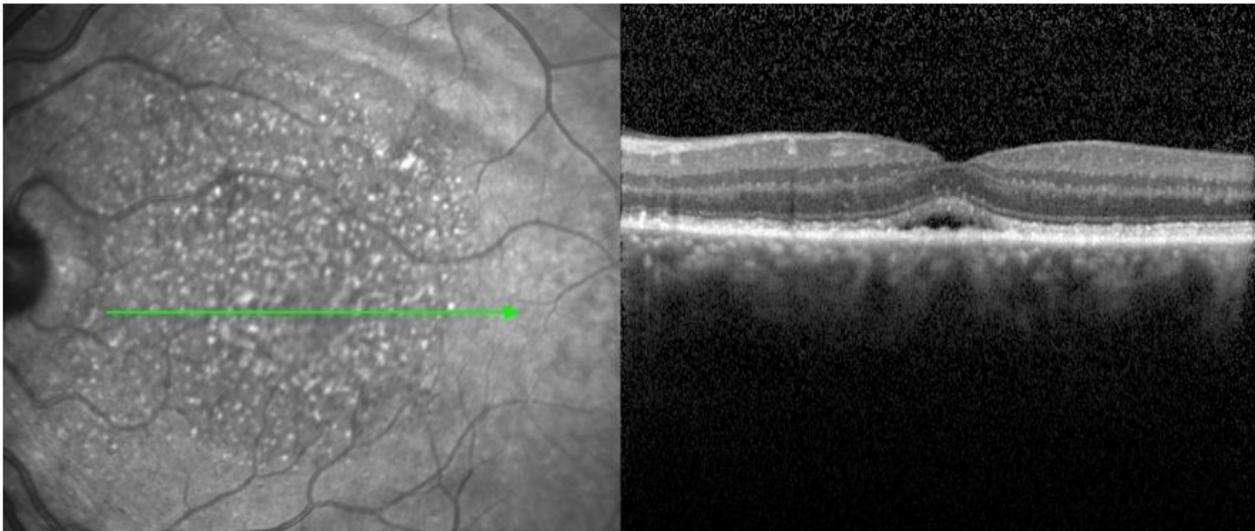
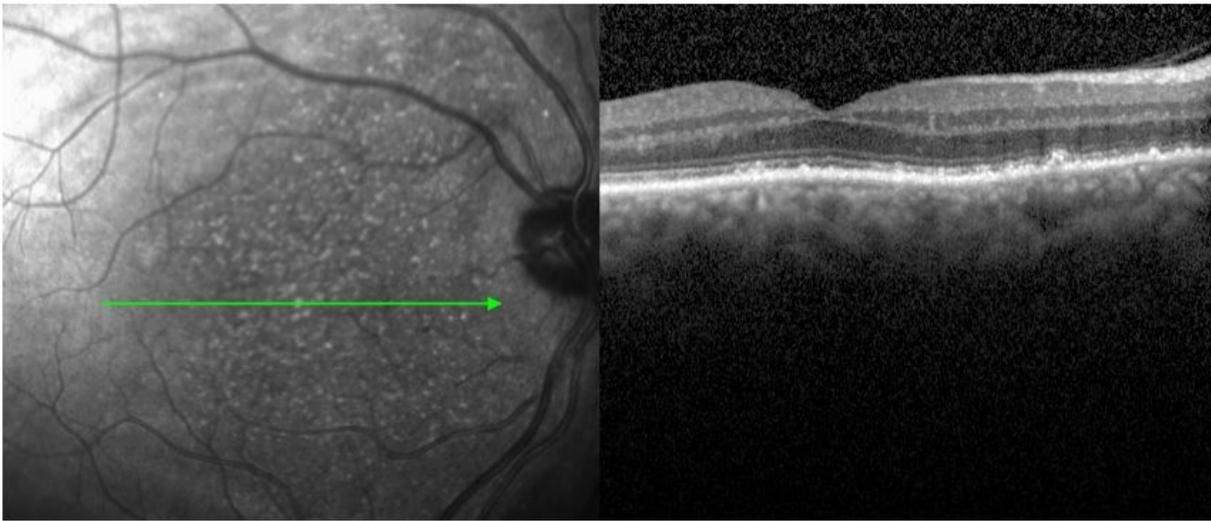


Figure 4

OCT horizontal transfoveal B-scans of the right and left eye at last follow up visit. Complete resolution of subretinal fluid in the right eye and remaining trace of subretinal fluid in the left eye is demonstrated.

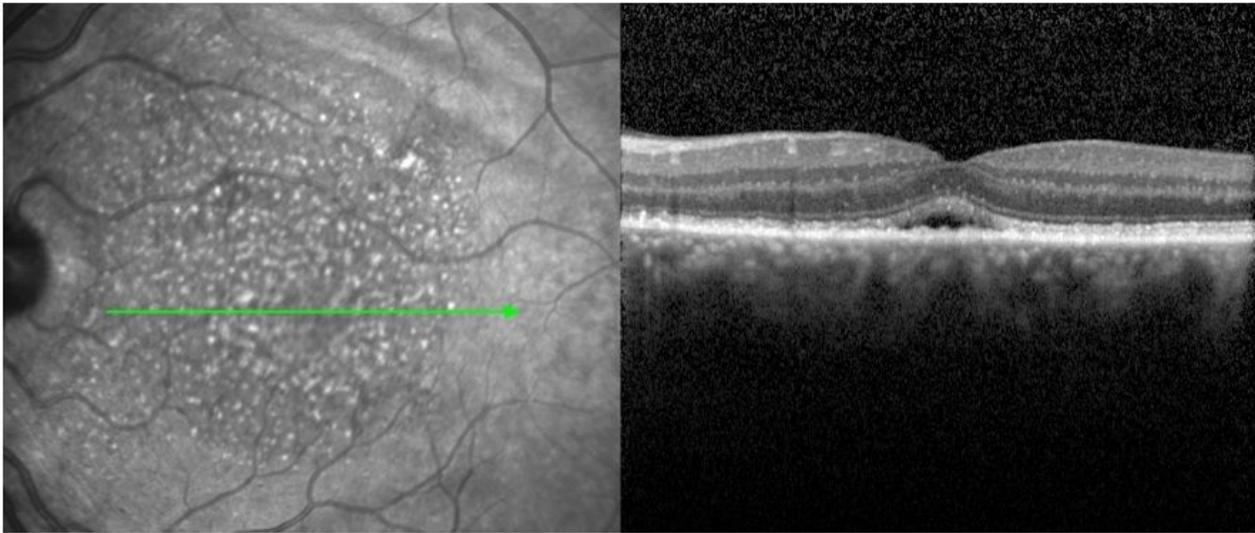
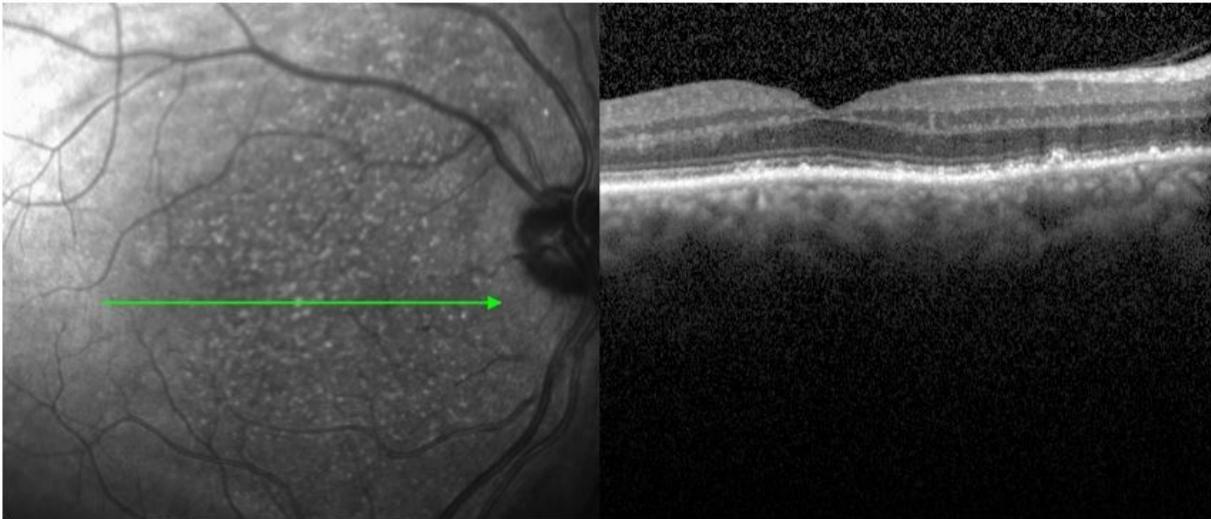


Figure 4

OCT horizontal transfoveal B-scans of the right and left eye at last follow up visit. Complete resolution of subretinal fluid in the right eye and remaining trace of subretinal fluid in the left eye is demonstrated.