

Unilateral Decreased Vision in a Middle-Aged Man

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CASE PRESENTATION

A 52-year-old man presented with decreased vision in his right eye from 1 month ago. He had no history of systemic disorders or drug consumption. Best corrected visual acuity (BCVA) was 1/10 in his right eye and 10/10 in the left eye at presentation. Anterior segment examination and intraocular pressure were within normal limits. Based on fundus examination, optical coherence tomography (OCT) (Figures 1 and 2) and fluorescein angiography (FA) findings (Figures 3 and 4), he received 1.25 mg intravitreal bevacizumab (IVB) in his right eye. After 6 weeks, he was not satisfied with the result and no improvement in BCVA was noted. OCT images (Figures 5 and 6), FA and indocyanine green angiography (ICGA, Figures 7 and 8) taken 6 weeks after injection are presented.

- What are the differential diagnoses?

- What do you suggest for the management of this patient?

Alireza Lashay, MD

This 52-year-old man has experienced decreased visual acuity in his right eye for more than 4 weeks. He received one 1.25 mg IVB injection without any improvement in BCVA after 6 weeks. Fundus autofluorescence (FAF) images of the right eye shows a large geographic hyperfluorescent area with a mottled hypofluorescent center which corresponds to multiple pinpoint foci of hyperfluorescence extending from the disc margin to at least 2 disc diameters temporal to the macula based on FA. At the center of this geographic area a serous pigment epithelial detachment (PED) is formed which demonstrates gradual gain in fluorescence in a well delineated circular pattern. This finding is consistent with OCT images of

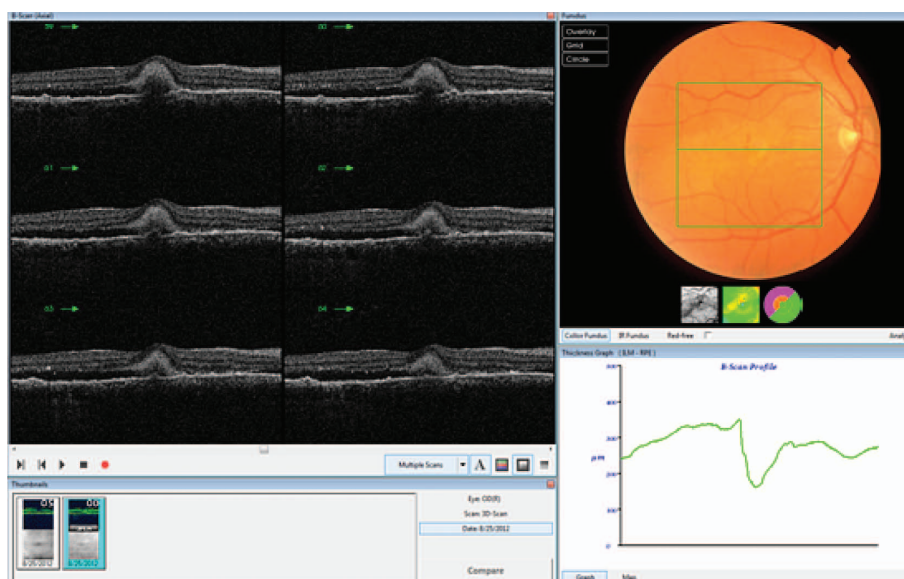


Figure 1. Optical coherence tomogram of the right eye at presentation

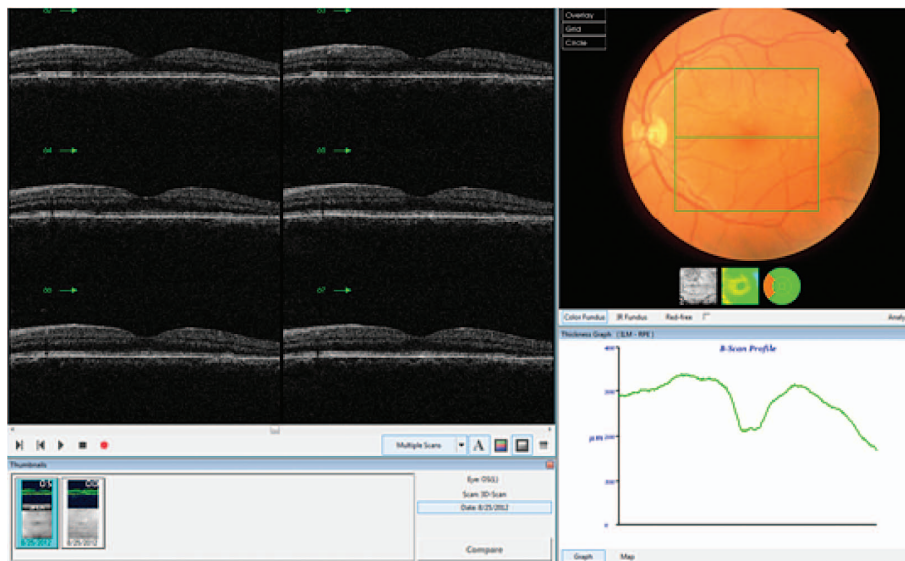


Figure 2. Optical coherence tomogram of the left eye at presentation

the right eye.

Although, OCT shows a mainly sero-fibrinous detachment of the neurosensory retina, interruption of retinal pigment epithelium (RPE) is noted in most scans. The same findings are visible in the fellow eye with the same imaging modality. After 6 weeks, this sero-fibrinous

detachment of the retina is still present and violation of RPE integrity is also visible. At the same time, in simultaneous FA/ICGA, hyperfluorescence of multiple foci in the central lesion has increased both in severity and magnitude. All of these findings are in favor of chronic multicentric central serous

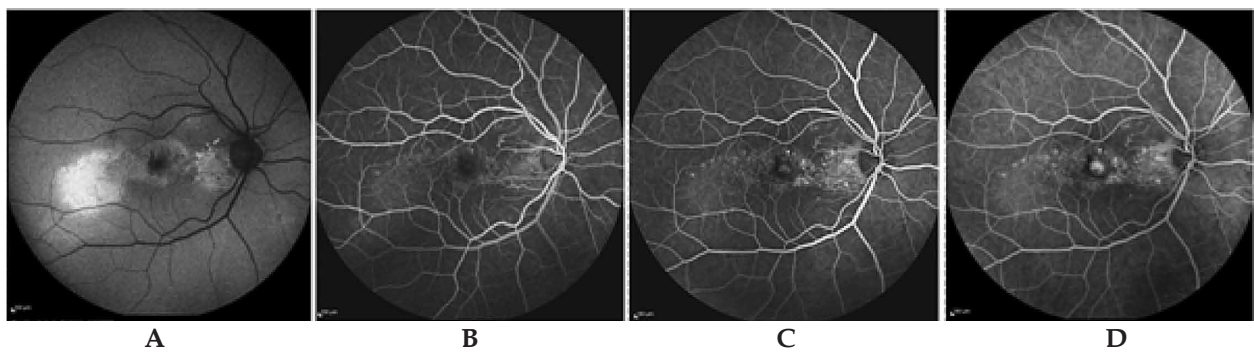


Figure 3. Autofluorescence (A) and fluorescein angiography (B-D) of the right eye at presentation

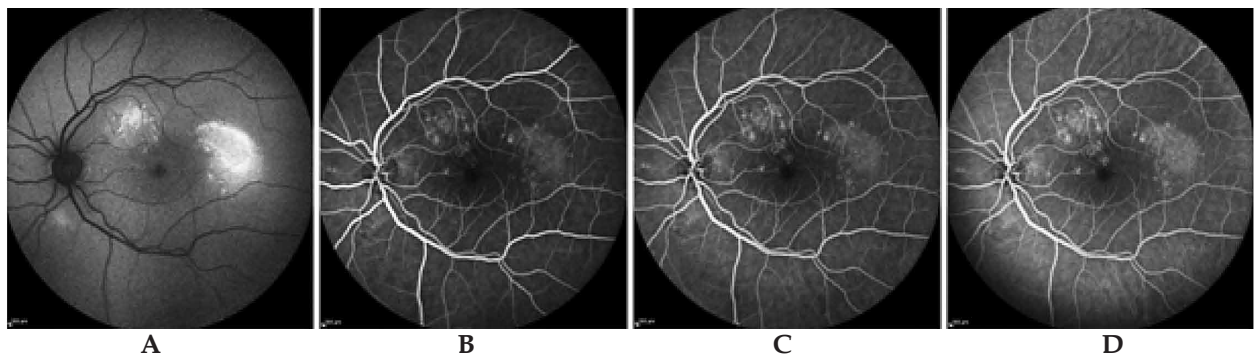


Figure 4. Autofluorescence (A) and fluorescein angiography (B-D) of the left eye at presentation

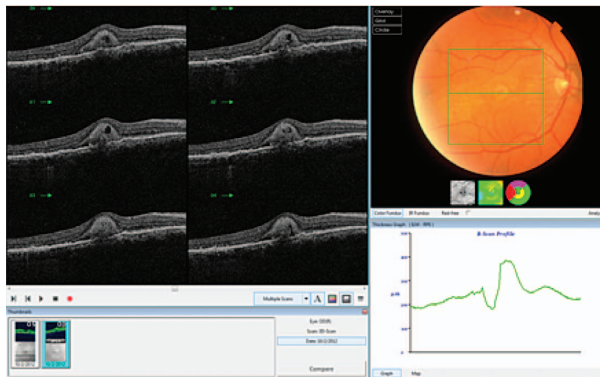


Figure 5. Optical coherence tomogram of the right eye 1.5 months after a single intravitreal injection of bevacizumab

chorioretinopathy (CSC), except that ICGA images disclose not only hyperpermeability of large choroidal vessels, but also discrete polyp-like hyperfluorescent spots which mostly washed out in late frames in both eyes.

These ICG findings raise the possibility of polypoidal choroidal vasculopathy (PCV), particularly considering the age of the patient and the deteriorating course despite treatment.

Review of the literature and our own experience support this idea that some CSC patients were originally PCV cases, and at times, CSC ultimately transforms to classic PCV. Other less possible differential diagnoses are Vogt-Koyanagi-Harada (VKH) syndrome, multiple evanescent white dot syndrome (MEWDS), acute posterior multifocal placoid pigment epitheliopathy (APMPPE), serpiginous chorioiditis and other inflammatory chorioretinal disorders.

Considering the treatment strategy and regardless of which of the first two diagnoses is more likely, photodynamic therapy (PDT) is the treatment of choice, although its application is somewhat different. In CSC, low fluence or half dose PDT is advised while in PCV, full dose PDT in conjunction with intravitreal injection of an anti-vascular endothelial growth factor (anti-VEGF) agent such as bevacizumab is recommended.

To summarize the case, this patient presented with signs and symptoms of chronic bilateral multicentric CSC before ICGA was performed but during follow-up and with the aid of ICGA, evidence of PCV prevailed. Since the center of the macula is not involved in his left eye and visual acuity is intact, the patient may be a candidate for full dose PDT along with IVB for the right eye however any decision for the left eye depends on the patient's concept of the disease. Discussing all aspects of treatment modalities and their possible complications, particularly for his better left eye, is mandatory. Unless, the patient is willing to receive treatments such as PDT, IVB, or even extramacular conventional laser therapy, close observation could be preferred.

Ahmad M Mansour, MD

The presence of subretinal exudation in a patient with neurosensory detachment of the macula frequently suggests a diagnosis of choroidal neovascularisation (CNV). Twenty-five years ago, I would have read the case as

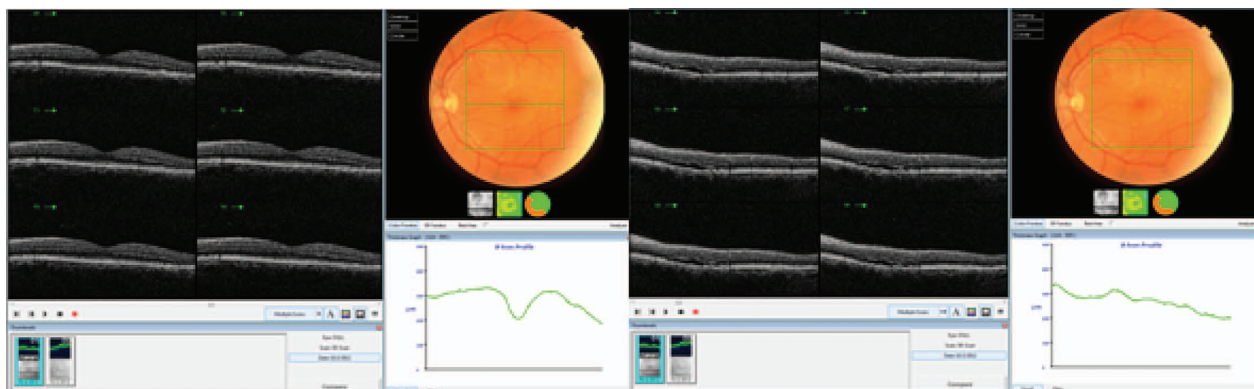


Figure 6. Optical coherence tomogram of the left eye at two scan levels, 1.5 months after intravitreal injection of bevacizumab in the right eye

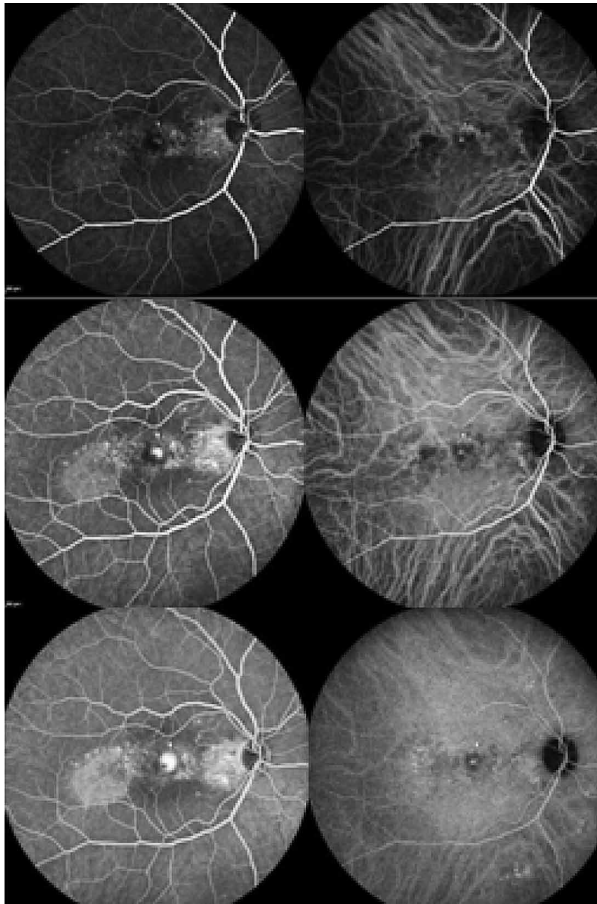


Figure 7. Fluorescein angiography and indocyanine green angiography of the right eye 1.5 month after a single intravitreal injection of bevacizumab.

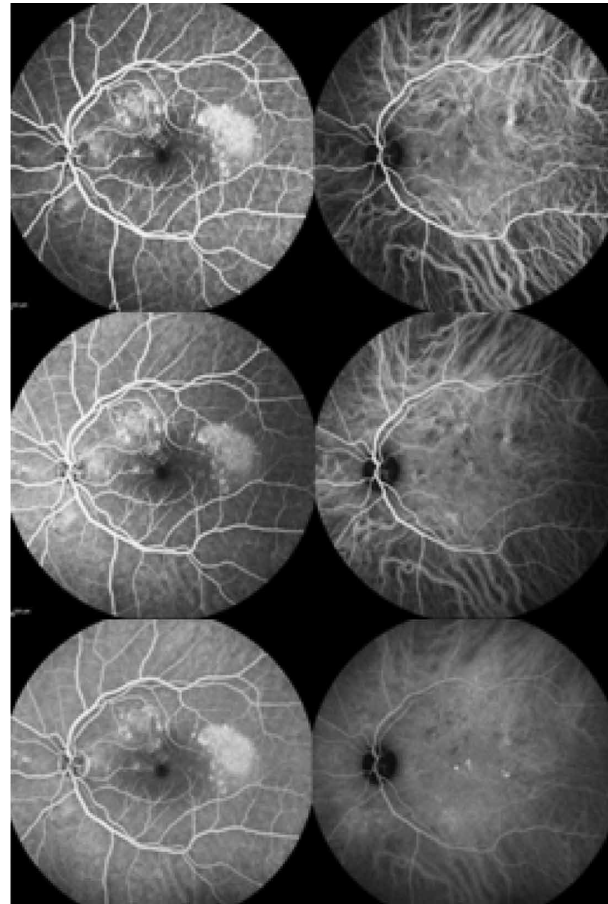


Figure 8. Fluorescein angiography and indocyanine green angiography of the left eye 1.5 month after intravitreal injection of bevacizumab in the right eye.

follows: the right eye shows subretinal fluid and a solid dome-shaped elevation of the neurosensory retina under the fovea with breaks in Bruch's membrane, corresponding loss of autofluorescence and late hyperfluorescence compatible with subretinal lipid or sub-RPE fibrovascular proliferation and hence the need for intravitreal bevacizumab.

However, advances in imaging by OCT and FAF have changed the way we look at different retinal diseases which may have similar appearance. This patient has serous elevation of the retina with oval subfoveal deposits in the right eye. Differential diagnoses include adult vitelliform macular dystrophy (VMD), Best disease, confluent drusen, and CSC. Adult VMD generally debuts between the third and fifth decades of life and causes slight visual deterioration. Fundus appearance can match the

current case and is frequently associated with drusen. In addition, it exhibits autofluorescence compatible with lipofuscin of vitelliform deposits, OCT hyper-refringence, and marked hypofluorescence in both early and late stages of FA. Generally, adult VMD is bilateral although unilateral cases have been described. This case had hypofluorescence which is unusual in the early stages of vitelliform deposition. Also the hyperfluorescent spot in the current case is not seen in vitelliform lesions unless CNV occurs which is rare.

Atypical VMD has been misdiagnosed as chronic CSC in few cases with serous macular detachment similar to the current case. Moreover, some authors have labelled two diagnoses, CSC and foveomacular vitelliform dystrophy (FMD), in patients with FMD who demonstrated a CSC-like appearance. But in my opinion this

condition is actually a variant of FMD.

Age of presentation in this case is not in favor of Best dystrophy. VMD is a rare disease with autosomal dominant inheritance. It can occur at any age, but in patients aged 3 to 15 years, it is known as Best VMD and in patients aged 30 to 50 years, it is considered to be adult-onset VMD with some differences in clinical manifestations. It was recently discovered however that both entities are due to abnormalities in the Best1 gene with different phenotypes and the primary cause is abnormalities in bestrophin function.

In the current case, we are essentially dealing with CSC; leakage points on FA correspond to hyperfluorescence on ICGA and areas of hyperautofluorescence correspond to tracts of CSC. Subfoveal deposits often noted in CSC resemble fibrin and may even mimic vitelliform lesions, and several reports have delineated the entity of dense subretinal deposits in CSC.

In summary, my presumptive diagnosis for this case is CSC with vitelliform-like subretinal deposits. I would suggest avoiding smoking, exercise, decreasing stress and consuming a diet rich in fruit, green vegetables and fish. Despite some studies in the literature, CSC has no treatment except decreasing the level of stress.

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Touka Banaee, MD

This case shows development of subfoveal CNV in the right eye superimposed on a background of bilateral chorioretinal disease. The first OCT of the right eye shows abnormal subfoveal deposits between the RPE and neurosensory retina along with subretinal fluid temporal to the fovea. There are some irregularities of the RPE line both temporal and nasal to the foveal center. FA images of the right eye are notable for the presence of patches of hyperautofluorescence studded with hypofluorescent dots, some of which seem to be due to intra/sub-retinal hemorrhage. The hyperautofluorescent areas correspond to areas of subretinal fluid on OCT. The first FA images of the right eye demonstrate the presence of classic subfoveal CNV along with a band of hyperfluorescence spanning the fovea composed of hyperfluorescent dots with minimal leakage.

OCT in the left eye is essentially normal except for the presence of mild RPE irregularities temporal to the fovea. There are hyperautofluorescent areas superior and temporal to the fovea on FAF images which show irregular hyperfluorescence with no leakage on FA. These areas can be presumed to have subretinal fluid, but no OCT scans of the affected areas are provided.

Differential diagnosis at this stage includes chronic CSC, or occult CNV of both eyes complicated by classic subfoveal CNV in the right eye. I am mostly in favour of the second diagnosis due to the presence of RPE irregularities on OCT which is not seen in CSC. However the pattern of hyper-fluorescent areas especially in the FA of left eye is compatible with a diagnosis of chronic CSC.

OCT and FA images one month after IVB injection in the right eye are essentially unchanged except for documentation of subretinal fluid superior to the left fovea corresponding to the site of hyper-autofluorescence on FAF. ICGA of both eyes show choroidal vessel wall

irregularities, small polyps, and multifocal choroidal hyperpermeability in later stages of ICGA. In the right eye, there may be a branching vascular network (BVN) of PCV superonasal to the foveal center, but in the left eye, no BVNs is noted. A small patch of choroidal hypoperfusion also exists temporal to the foveal center on ICGA of the right eye.

The presence of RPE irregularities on OCT, and the small polyps, choroidal vessel wall irregularities and multifocal choroidal hyperpermeability in ICGA in this case are compatible with a diagnosis of PCV. The pattern of hyperfluorescence in FA, and the choroidal hyperpermeability and hypoperfusion patches on ICGA are compatible with a diagnosis of CSC.

As has previously been stated, some cases of CSC turn into PCV later in life.¹ CSC and PCV seem to have a common pathophysiology originating in the choroidal vessels manifesting as choroidal vascular hyperpermeability which is a characteristic feature of CSC² and seen in up to approximately 60% of cases with PCV.³ These cases also have a thick choroid.⁴ Performing enhanced depth imaging (EDI) OCT in this case may be helpful in this regard.

Park et al⁵, recently reported 13 cases of PCV with signs of chronic CSC. All of their cases had unilateral PCV, and 84.6% had signs of chronic CSC in their fellow eyes. They also reported that there were no prominent BVN in any of their cases and that all polyps were outside the boundary of the atrophic retina. These authors reported good response to treatment in their series.

Recently, Koizumi et al⁴ reported that cases with PCV and multifocal choroidal hyperpermeability who are more likely to have bilateral disease, have a history of CSC and are less responsive to anti-VEGF treatment.

Symptoms of the present case originate from development of classic subfoveal CNV and not the PCV itself. This complication can be considered as a sequel of chronic CSC. It seems that continuation of treatment with anti-VEGF agents will have a good effect and will control the classic CNV. If the response is suboptimal, then combination therapy with PDT and anti-VEGFs as suggested for PCV can be considered.

In my opinion, this is an interesting case which demonstrates features of both PCV and CSC, and is probably a case caught in the transition phase from CSC to PCV, especially in the left eye where polyps are not very prominent and distinct. What has brought this case to attention has been the development of subfoveal type 2 CNV in the right eye which can be considered a complication of chronic CSC.

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Conflicts of Interest

None.

Consultants

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