Sudden Macular Distortion in a Middle-aged Woman

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CASE PRESENTATION

A 40-year-old woman presented with decreased vision and metamorphopsia in her left eye since 3 weeks ago. She had no history of systemic disorders and drug history was unremarkable. Best corrected visual acuity (BCVA) was 10/10 and 8/10 in her right and left eyes, respectively and afferent pupillary defect was negative. Anterior segment examination and intraocular pressure were normal. On fundus examination, a small area of subretinal fluid accumulation was noted in the foveal region of the left eye which was confirmed by optical coherence tomography (OCT).

Figures 1 and 2 respectively represent the fluorescein angiography (FA) and OCT images at presentation. A single injection of intravitreal bevacizumab (IVB, 1.25mg) was administered and two weeks afterwards, visual acuity increased to 10/10 and metamorphopsia significantly improved. OCT images (2 and 4 weeks after the IVB injection) together with FA (4 weeks after the injection) and indocyanine green angiography results (5 weeks after the injection) are demonstrated in figures 3 to 6.

What are your differential diagnoses and suggestions for the management of this patient?

Figure 1. Fluorescein angiography (FA) at presentation (left image, right eye; right image, left eye).

Figure 2. Optical coherence tomography (OCT) images at presentation.
Based on the clinical picture, FA and OCT images, this lady seems to be affected with idiopathic juxtafoveal choroidal neovascularization (CNV) in her left eye which partially regressed two weeks after IVB injection, as evidenced by visual improvement and OCT imaging. However, follow-up FA is not available to evaluate leakage activity of the lesion.

Four weeks after the injection, the lesion is still active as shown by leakage on FA and ICG. OCT shows minimal subretinal fluid at this stage, but this does not rule out exudative
activity of the lesion.

Considering the good level of visual acuity, juxtafoveal location of the CNV and its obvious activity, I would recommend further IVB injections at this stage (5 weeks following the first injection) and one month thereafter. Four weeks after the third injection, I would repeat FA and OCT and later decide about lesion activity which would be the basis for future treatment recommendations. I believe that the benefits of intravitreal anti-vascular endothelial growth factor (VEGF) treatment for this young patient far outweigh possible risks associated with such therapy.

Reza Karkhaneh, MD

The patient described herein is a case of intraretinal neovascularization. According to baseline OCT and FA images, it seems that the new vessels have originated from deep retinal capillaries between the inner nuclear layer and the outer plexiform layer (OPL), and progressed toward the retinal pigment epithelium (RPE) to form intraretinal neovascularization and retinal–retinal anastomosis.

Baseline FA images reveal a focal area of intraretinal staining and leakage corresponding to intraretinal neovascularization and edema (hyperfluorescence). The baseline OCT image shows a highly reflective area between the OPL and RPE corresponding to macular edema and intraretinal neovascularization, disintegrity of the outer retina, inner segment outer segment (IS/OS) junction and cone outer segment tip (COST) line. Early-phase ICG angiography performed 5 weeks after IVB injection reveals a focal area of intense hyperfluorescence or “hot spot” corresponding to intraretinal neovascularization and retinal–retinal or even retino-choroidal anastomosis.

Differential diagnoses in this case may include parafoveal telangiectasia, macular branch retinal vein occlusion (BRVO), CNV secondary to central serous retinopathy (CSR) and idiopathic CNV.

Patients with parafoveal telangiectasia are usually male and intraretinal exudates are a prominent feature in this entity; these features are not present this case. FA and ICG findings are also not consistent with BRVO. According to OCT findings, the lesion is located in the outer retina but the RPE seems intact without invasion of neovascularization into the subretinal space, therefore OCT findings are not consistent with CNV.

Anti-VEGF therapy is a promising treatment modality for eyes with intraretinal neovascularization. After receiving a single IVB injection, the patient experienced good visual recovery while FA and ICG angiography still demonstrated intraretinal leakage, and OCT on the other hand revealed residual macular edema. Considering the good response to a single IVB injection, additional monthly intravitreal injections over the following 2 months may effectively reduce intra-retinal leakage and edema. However, in cases resistant to this treatment, photodynamic therapy (PDT) guided by ICG in combination with IVB, may be considered as the next treatment option.

Morteza Entezari, MD

The patient is a 40-year-old woman with decreased vision and left sided metamorphopsia with neither systemic disorders nor history of drug consumption. On funduscopy, there was a small area of subretinal fluid in the left foveal region.

Based on the clinical presentation, fundus findings, localized RPE detachment on OCT and fluorescein leakage on FA, the most probable diagnosis is CNV which may have different etiologies. However, considering the patient’s age, her negative systemic disease and drug consumption history, normal anterior chambers and a normal appearing fellow eye, idiopathic CNV remains the most likely diagnosis; nevertheless, it would be better to perform an ICG angiography for a more precise diagnosis.

The left eye showed improvement after IVB injection; however 4 weeks after the injection, OCT shows subfoveal fluid, and leakage is evident on FA and ICG angiography which demonstrate an angiomatous appearance. After performing the ICG angiography my diagnosis is retinal angiomatosis proliferation (RAP).
As for the treatment of this patient, I would suggest a combination of photodynamic therapy and intravitreal anti-VEGF or triamcinolone acetonide.

Nasser Shoeibi, MD

The patient presented herein is a middle-aged lady with unilateral metamorphopsia and mild visual loss. The fundus photograph reveals an ill-defined yellowish subretinal lesion in the superotemporal parafoveal area. Oblique B scan OCT through the lesion, shows disruption of RPE/Bruch’s membrane, disruption of inner segment/outer segment junction, possible extension of CNV from the RPE into the subretinal space, few cystoid spaces in the outer retina, and relatively intact inner retina in the parafoveal area. Two areas of hyper-reflectivity in the inner retina with backscatter shadowing correspond to normal retinal vessels crossing the selected section. Corresponding FA reveals few points of leakage in the parafoveal area with corresponding leakage in the arteriovenous phase (82 seconds) which can be indicative of a CNV lesion. However, similar OCT findings can sometimes be found with RAP, but the age of the patient, absence of retinal hemorrhage and corresponding vessels on angiography make this diagnosis less likely. White dot syndromes should be considered as a differential diagnosis for similar retinal findings without vitritis in a young lady. Absence of multiple bilateral lesions and early hypofluorescence on ICG angiography make them a less probable diagnosis but with time, new lesions may appear changing the clinical picture.

In patients under the age of 50, CNV is often the result of pathologic myopia, inflammatory chorioretinopathies, angioid streaks or idiopathic causes. Given the patient’s presentation with unilateral metamorphopsia, lack of ocular inflammation (iritis or vitritis), absence of associated retinal or choroidal findings for specific diagnoses and angiographic evidence of an active choroidal neovascular membrane, a presumptive diagnosis of idiopathic CNV can be made.

Management of CNV is a challenge. Multiple therapeutic strategies can be used with varying success. Symptoms have been diminished after IVB injection in this patient which indicates a positive response to such treatment. B-scan OCTs after IVB, demonstrate restoration of normal retinal structure. However, one should remember that these horizontal B scan OCTs do not cross the lesion and cannot be relied on for lesion evaluation; therefore, we can only conclude that subfoveal fluid has resorbed.

Angiographies at 4 and 5 weeks after IVB, show a still active lesion (dye leakage on FA and a hot spot on ICG angiography) reflecting the necessity for further therapy. In my opinion, repeat intravitreal injections of bevacizumab at 6-week intervals for at least 3 sessions and then extending the treatment as needed would be effective. One can combine intravitreal angiostatic agents with intravitreal or subtenon corticosteroids. The lesion lies in the parafoveal area, therefore, in case of persistent CNV activity, a combination of IVB with laser or photodynamic therapy can be helpful. OCT and FA will be useful in assessing the response to therapy.

Consultants

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