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SHORT COMMUNICATION

Intravitreal bevacizumab for adult-onset vitelliform dystrophy: A case report

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PURPOSE. *Adult-onset foveomacular vitelliform dystrophy (AFVD) is often misdiagnosed as occult choroidal neovascularization (CNV) secondary to age-related macular degeneration (AMD). The authors report the anatomic and functional outcome of intravitreal bevacizumab in a case of AFVD associated with a suspected occult CNV.*
METHODS. *Prospective, interventional, single case report. One female patient with decreased visual acuity (VA) and metamorphopsia secondary to AFVD received one single intravitreal injection of bevacizumab 1.25 mg.*
RESULTS. *The patient reported unchanged VA and decreased metamorphopsia 6 weeks after the injection. Fluorescein angiography (FA) and optical coherence tomography (OCT) showed progressive decrease of subretinal fluid until complete disappearance. VA, OCT, and FA remained unchanged during 10 months follow-up.*
CONCLUSIONS. *Intravitreal bevacizumab showed a morphologic improvement and stable VA in a patient with AFVD. Further case series are required to confirm this observation. (Eur J Ophthalmol 2007; 17: 983-6)*

KEY WORDS. *Adult-onset foveomacular vitelliform dystrophy, Age-related macular degeneration, Bevacizumab, Choroidal neovascularization, Retinal pigment epithelium dystrophy*

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INTRODUCTION

Adult-onset foveomacular vitelliform dystrophy (AFVD) is a degenerative condition affecting retinal pigment epithelium (RPE) that is often misdiagnosed as occult choroidal neovascularization (CNV) secondary to age-related macular degeneration (AMD) (1). Fluorescein angiography (FA) typically shows a hypofluorescent area corresponding to the vitelliform lesion surrounded by a hyperfluorescent ring, with late frames usually showing increased perilesional fluorescence which is possibly caused by staining of the subretinal fluid or fluorescein pooling. Optical coherence tomography (OCT) shows vitelliform lesions located in the RPE layer or between the RPE and photoreceptor layer. CNV may appear as a complication of AFVD, usually causing decrease of BCVA in 15% of the cases and presence of subretinal fluid in OCT (2).

Case report

A 72-year-old woman was referred to our center to be treated for neovascular AMD by photodynamic therapy (PDT). Her clinical history was positive for high blood pressure, hypercholesterolemia, hepatic angioma, and vitiligo. Best-corrected visual acuity (BCVA) was 20/80 in her right eye and 20/60 in her left eye. She reported decreased visual acuity with metamorphopsia in her right eye during the past month. Fundus examination revealed

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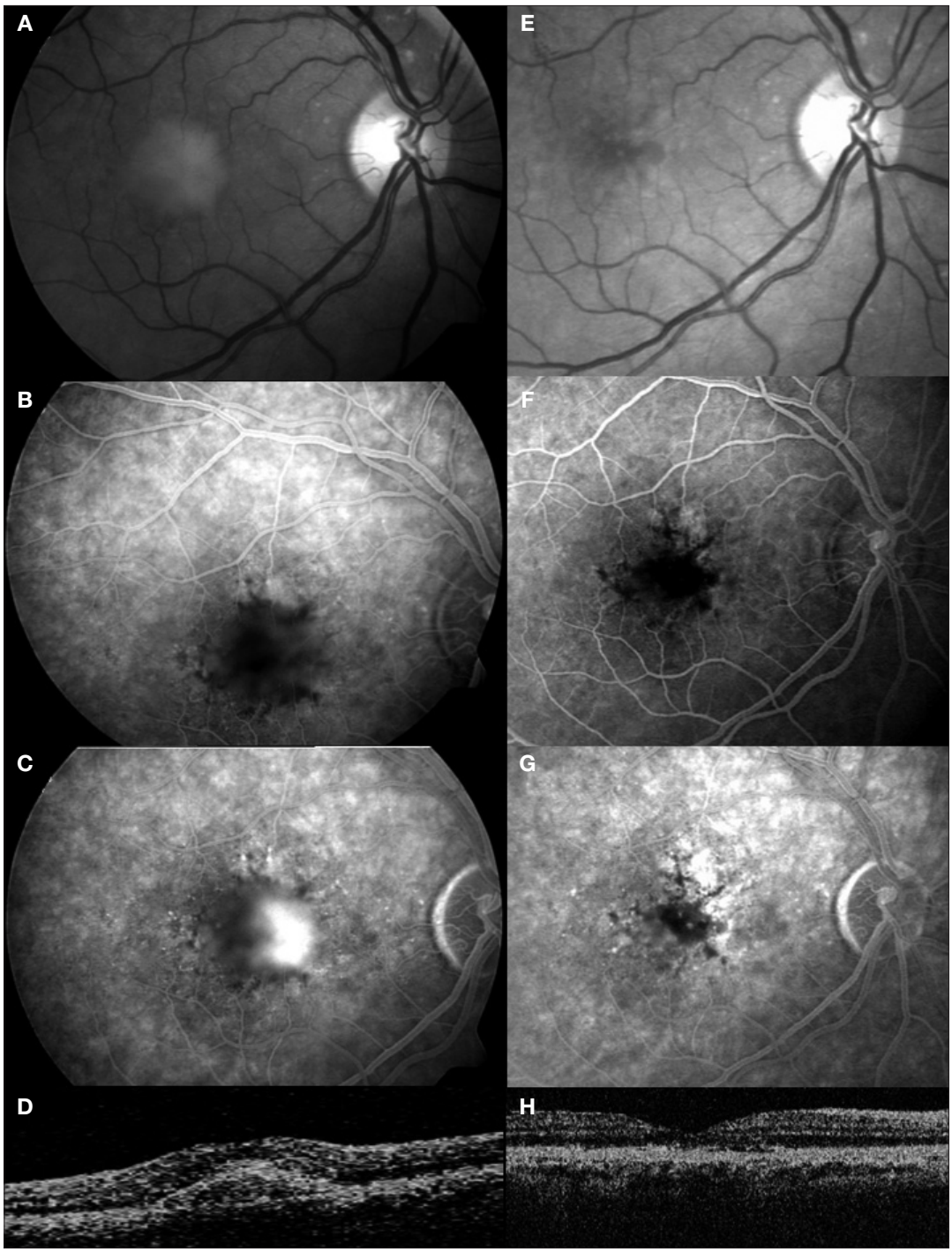

Fig. 1 - (A-C) *Red-free retinographies and fluorescein angiography (FA) show central vitelliform lesions with progressive hyperfluorescence.* **(D)** *Optical coherence tomography (OCT) reveals a subretinal spindle-shaped deposit with subretinal fluid.* **(E-G)** *Red-free retinographies and FA show disappearance of vitelliform lesion and fluorescein leakage 6 weeks after treatment.* **(H)** *OCT 1 month after bevacizumab injection shows disappearance of the subretinal lesion as well as complete anatomic recovery of the fovea.*

central vitelliform lesions in both eyes (Fig. 1A). FA showed right eye early hypofluorescent macular lesions with progressive hyperfluorescence that was suspicious for an occult underlying CNV (Fig. 1, B and C). OCT disclosed a subretinal spindle shaped deposit with reinforcement of the RPE and subretinal fluid surrounding the lesion in her right eye (Fig. 1D). Six weeks later, BCVA, FA, and OCT remained unchanged, whereas the patient described increased metamorphopsia in her right eye.

Therapeutic options were discussed with the patient and

one 1.25 mg bevacizumab intravitreal injection was performed after obtaining written informed consent and authorization for compassionate use.
Treatment and data gathering were approved by the local ethics committee and performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

RESULTS

Six weeks after the injection, BCVA in the right eye was 20/80 and the patient described decreased metamorphopsia. Fundus examination showed disappearance of subretinal vitelliform deposits (Fig. 1E) and FA showed a persistent hypofluorescent area surrounded by mottled RPE atrophy (Fig. 1, F and G). OCT showed progressive decrease of subretinal deposit until disappearance (Fig. 1H).
Ten months after the injection, BCVA was 20/80 in the right eye and metamorphopsia was no longer present. No subretinal deposits were observed by OCT and no leakage appeared in FA. BCVA in the left eye had dropped to 20/100 with central scotoma and metamorphopsia. Left eye FA showed increased hyperfluorescence suggesting CNV and OCT revealed progression of subretinal deposits with presence of subretinal fluid.

DISCUSSION

AFVD has been reported to represent a subgroup of AMD (2) as is shown by its association with RPE detachment and CNV. This condition usually ends as atrophic areas of RPE with macular thinning and visual deterioration (3, 4). RPE atrophy is probably the most important factor limiting improvement of BCVA.
AFVD has been treated in cases with or without CNV, intentionally or by mistake, by conventional PDT without success, occasionally inducing further visual loss (1).
In the past years, antiangiogenic therapy has been used for conditions associated with CNV and followed by good visual and anatomic results (5). However, repeated injections of antiangiogenic drugs have been necessary to achieve final closure of new vessels.
The case reported represents disappearance of subretinal deposits and fluid associated with AFVD after one injection of an antiangiogenic agent, reducing metamorphopsia while leaving BCVA unchanged.
Proper identification of CNV associated with AFVD is not always easy as has been demonstrated by mistaken treatment of AFVD by PDT (1). The presence of subretinal fluid and late FA stain in the case reported might be considered signs of associated occult CNV. However, the absence of subretinal hemorrhage and lipids may support the contrary, this being a case of pure AFVD.
Preservation of visual acuity and disappearance of fluorescein leakage in the case reported seem to have been induced by the treatment, especially considering the events in the untreated fellow eye, where BCVA decreased and subretinal fluid appeared during the following months. However, the question remains open whether disappearance of subretinal fluid can be attributed to closure of the suspected occult CNV, or to the effect of the drug on the increased vessel permeability associated with AFVD, which seems to be responsible for the presence of this subretinal fluid. Intravitreal bevacizumab might be a useful therapy in both cases, reducing vascular hyperpermeability.
Disappearance of the vitelliform deposits may or not be related with the intravitreal injection of the antiangiogenic drug, while mottled atrophy of the RPE is probably related to the natural course of the condition, as has been previously described (4).
Further case series are required to confirm the real advantages of this therapy on subretinal fluid, associated or not with occult CNV, in AFVD.

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