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SHORT COMMUNICATION

Case report

Vascularized pigment epithelial detachment in adult-onset foveomacular vitelliform dystrophy

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PURPOSE. *To report a case showing adult-onset foveomacular vitelliform dystrophy (AOFVD), associated with vascularized pigment epithelial detachment.*
CASE REPORT. *A 72-year-old female affected by AOFVD complained with blurred vision and metamorphopsia in her right eye, seven months after a routinary clinical examination. Visual acuity in right eye dropped from 0.6 to 0.3, and biomicroscopic fundus examination revealed a serous pigment epithelial detachment arising from the temporal margin of the pseudovitelliform lesion. Fluorescein angiography showed an uneven filling of the pigment epithelial detachment, suggesting the presence of a subfoveal choroidal neovascularisation, which was confirmed by indocyanine green angiography.*
DISCUSSION. *The association between AOFVD and vascularized pigment epithelial detachment, supports the hypothesis that AOFVD may be a different subgroup of age-related macular degeneration with specific genetic predisposition. (Eur J Ophthalmol 2000; 10: 266-9)*

KEY WORDS. *Adult-onset foveomacular vitelliform dystrophy, Vascularized pigment epithelium detachment, Age-related macular degeneration*

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Visual acuity in adult-onset foveomacular vitelliform dystrophy (AOFVD) can deteriorate for several reasons, including the natural course of the disease (1-4), atrophy (1, 3-5), or choroidal neovascularisation (1-6). We report a case showing AOFVD associated with vascularized pigment epithelial detachment.

Case report

A 72-year-old woman was referred to our center for clinical evaluation. General history revealed hypertension. Visual acuity was 0.6 in both eyes. Anterior segment showed a mild corticonuclear cataract, and intraocular pressure was 13 mmHg in both eyes. Biomiscroscopic fundus examination showed a subfoveal round yellow lesion, with slightly irregular contours, measuring one disc diameter, in the left eye. The right eye had the same lesion, associated with retinal pigment epithelium atrophy and pigment clumping. The macular region showed drusen in both eyes (Fig. 1 and 2). Fluorescein angiography revealed a central nonfluorescent round area surrounded by a hyperfluorescent ring, which stained in late phases in the left eye, and a combination of hypofluorescence and hyperfluorescence due respectively to a masking effect and a window defect secondary to the pigmentary changes in the right eye (Fig. 3 and 4). Indocyanine green angiography showed a central dark spot surrounded by increasing hyperfluorescence, corresponding to the biomicroscopically visible subretinal yellow lesion, which was detectable more clearly during the late phases (Fig. 5 and 6). Electrooculographic findings were subnormal (150 in both eyes), and the electroretinogram was normal. The patient's clinical and angiographic features were consistent with the diagnosis of AOFVD (1-7). Examination of her relatives was negative.

Seven months later the patient asked for a fur-

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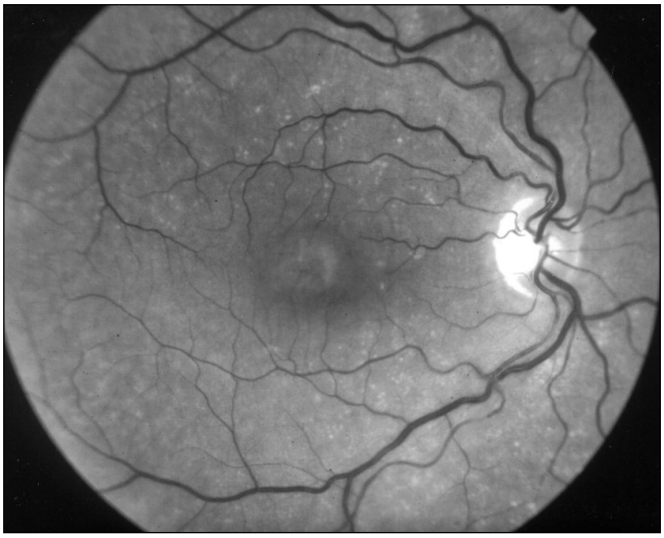

Fig. 1 - *Right eye; red-free frame; AOFVD with central lesion and retinal pigment epithelium changes.*

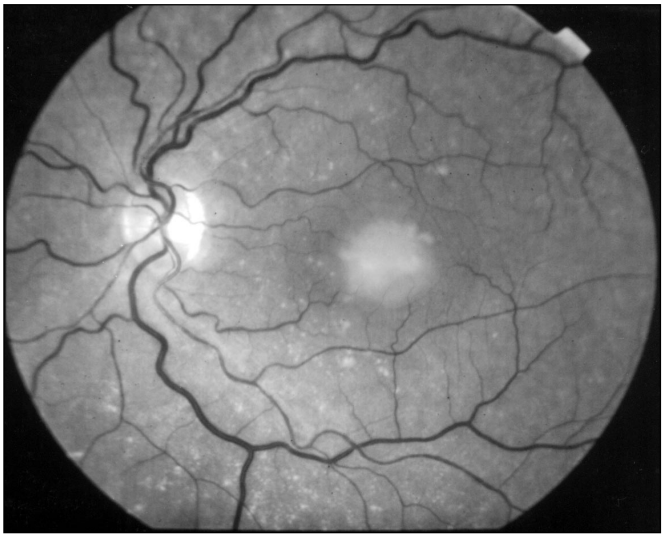

Fig. 2 - *Left eye; red-free frame; typical AOFVD with subfoveal round lesion, with slightly irregular contours.*

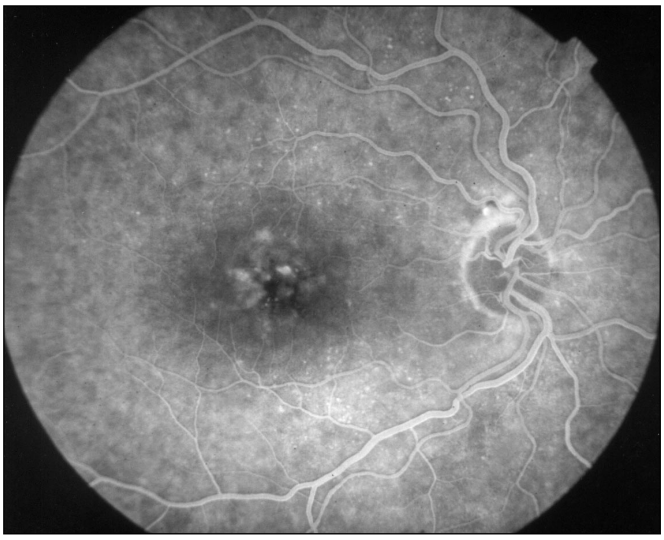

Fig. 3 - *Right eye; fluorescein angiography (3 minutes) shows retinal pigment epithelium atrophy and pigment clumping.*

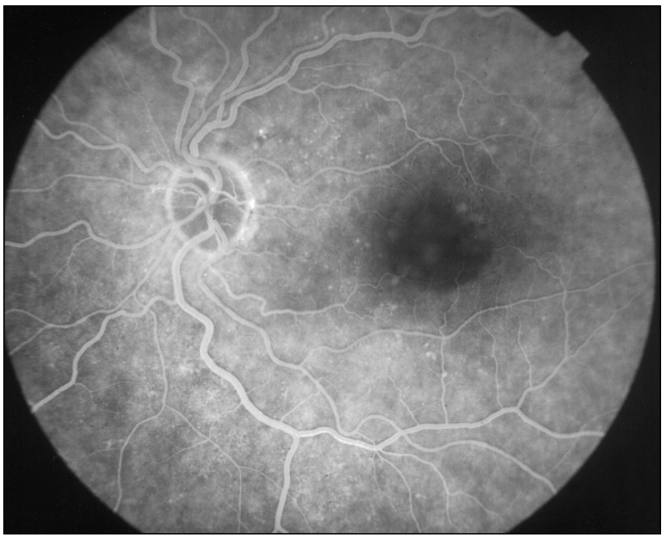

Fig. 4 - *Left eye; fluroescein angiography (4 minutes) showing slight staining of the central lesion.*

ther clinical examination complaining of blurred vision and metamorphopsia in the right eye. Visual acuity was 0.3 in the right eye and 0.6 in the left. Biomicroscopic fundus examination of the right eye revealed a serous pigment epithelial detachment arising from the temporal margin of the pseudovitelliform lesion. Fluorescein angiography showed uneven filling of the detachment, suggesting subfoveal choroidal neovascularisation (Fig. 7). Indocyanine green angiography confirmed the subfoveal choroidal neovascularisation, highlighting the relationship with the subretinal pigment epithelial material (Fig. 8). The patient was not given any treatment.

Three months later, visual acuity was 0.1 in the righ eye and 0.4 in the left. Biomicroscopic fundus examination revealed flattening of the vascularized pigment epithelial detachment with fibrotic evolution in the right eye (Fig. 9), and partial reabsorption of pseudovitelliform material associated with pigmentary changes in the left eye. Fluores-

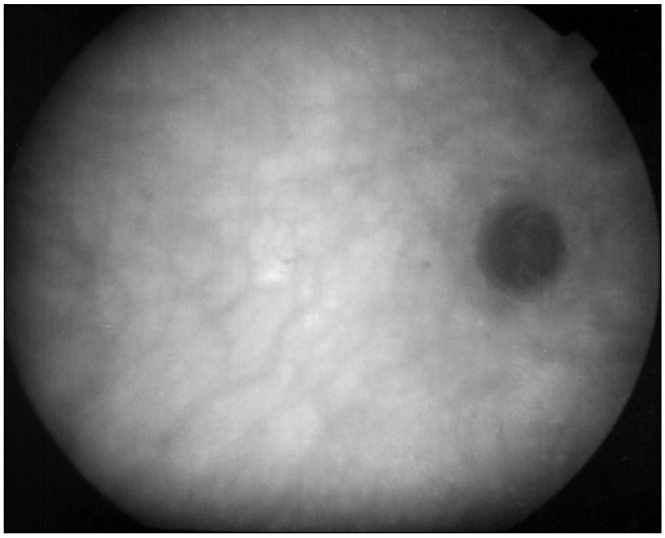

Fig. 5 - *Right eye; indocyanine green angiography (40 minutes). Central hyperfluorescence probably due to dye binding to pseudovitelliform material.*

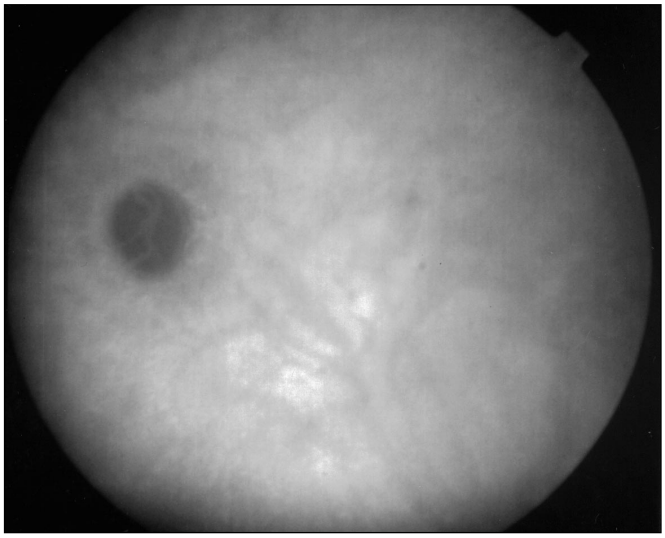

Fig. 6 - *Left eye; indocyanine green angiography (40 minutes), showing a more extensive central hyperfluorescence corresponding to the AOFVD.*

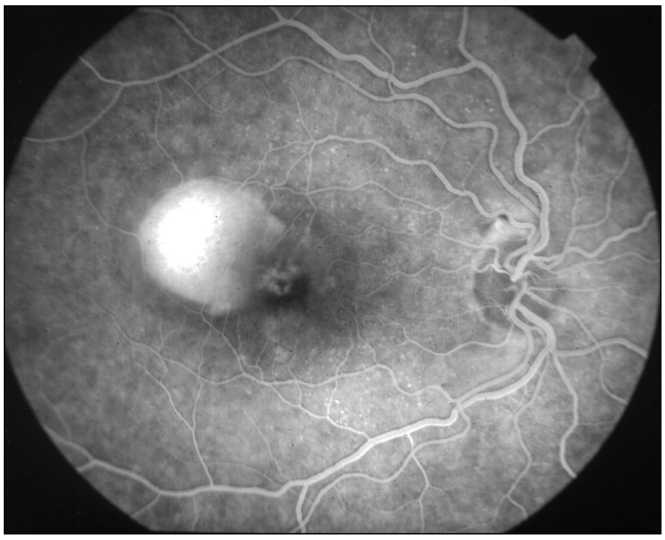

Fig. 7 - *Right eye; fluorescein angiography (6 minutes) showing the neovascular pigment epithelium detachment.*

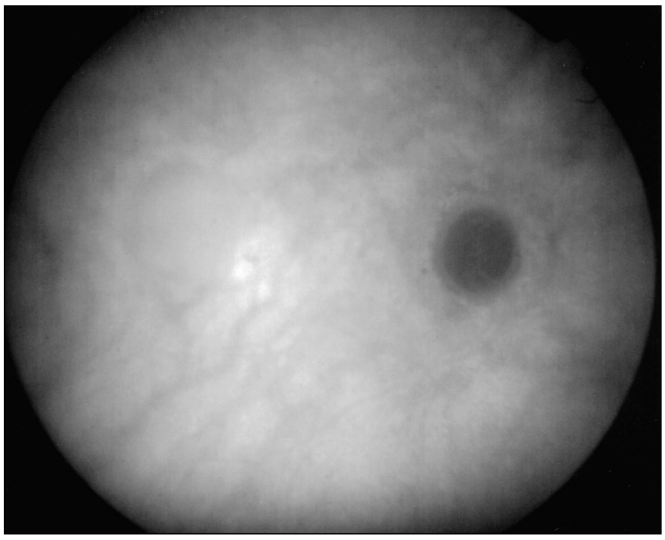

Fig. 8 - *Right eye; indocyanine green angiography (40 minutes) showing subfoveal choroidal neovascularisation.*

cein and indocyanine green angiography even more clearly showed the presence of subfoveal choroidal neovascularisation in the right eye (Fig. 10 and 11).

DISCUSSION

AOFVD comprises a heterogeneous group of disorders presenting different clinical, angiographic and histopathologic features. Choroidal neovascularisation may develop in from 5 to 15% of cases (1-6). AOFVD shares several points with age-related macular degeneration, inlcuding the patients' mean age, macular drusen, retinal pigment epithelium changes, progression towards atrophy or choroidal neovascularisation (1, 2, 4). AOFVD eyes presenting drusen are more likely to develop choroidal neovascularisation (4).

As far as we know, this is the first report of vas-

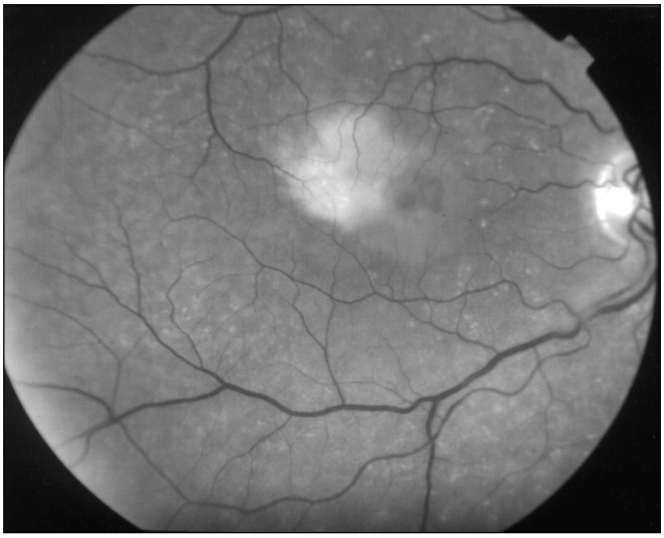

Fig. 9 - *Right eye; red-free frame. Flattening of the pigment epithelium detachment with fibrotic evolution.*

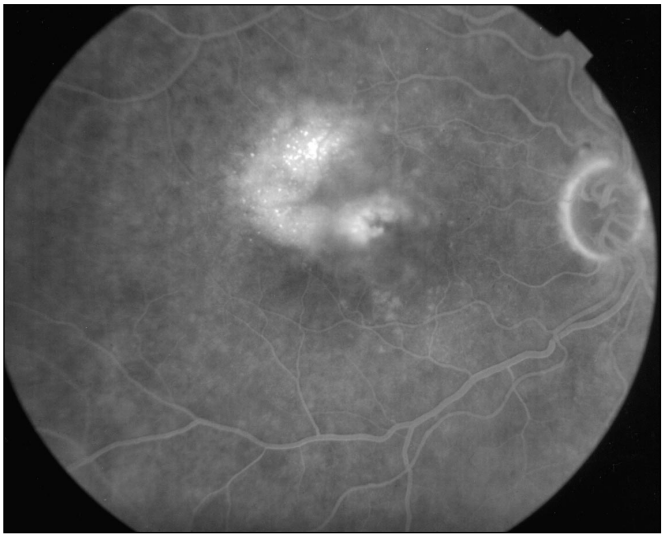

Fig. 10 - *Right eye; fluorescein angiography (5 minutes) showing irregular staining at the posterior pole.*

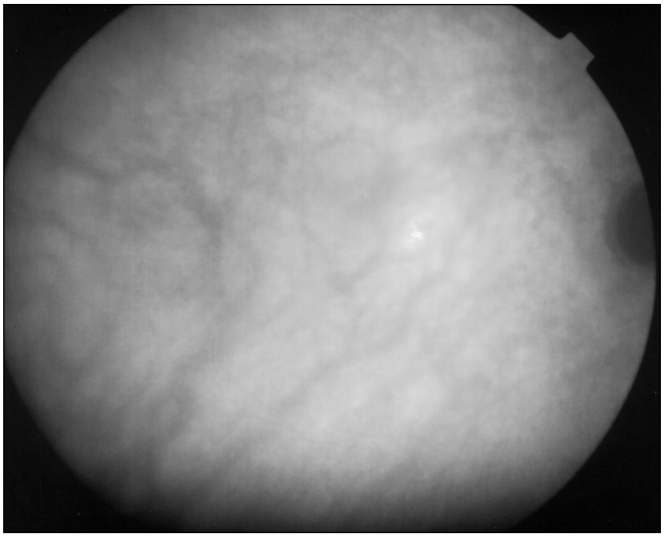

Fig. 11 - *Right eye; indocyanine green (40 minutes). Focal subfoveal choroidal neovascularisation, with almost complete disappearance of the shadow of pigment epithelium detachment.*

cularized pigment epithelial detachment associated with AOFVD. This type of detachment is a typical expression of age-related macular degeneration. The findings in the present case suggest a close relationship between AOFVD and age-related macular degeneration. Future studies will clarify whether two deseases share a common genetic basis.

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