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SHORT COMMUNICATION

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Adult-onset foveomacular vitelliform dystrophy and indocyanine green videoangiography

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Abstract ● Background: Adult-onset foveomacular vitelliform dystrophy (AOFVD) represents a heterogeneous group of disorders with different clinical, angiographic, and histopathological features. The most common form is characterized by a yellow, round to oval subretinal macular lesion with or without central pigmented spot. ● Methods: Eight patients affected by typical AOFVD underwent fluorescein angiography and indocyanine green videoangiography (ICGV). ● Results: Fluorescein angiography showed a central hypofluorescent spot surrounded by an irregular hyperfluorescent ring. ICGV demonstrated a foveal nonfluorescent spot, visible during the entire examination, and a hyperfluorescent area surrounding the central spot, which became evident soon after the beginning of the examination.
● Conclusions: In light of previous histopathological studies, the central nonfluorescent spot may be interpreted as a masking effect of a pigment clump, whereas the hyperfluorescent area may represent dye pooling or staining of the subretinal pigment epithelial material.

Introduction

Adult-onset foveomacular vitelliform dystrophy (AOFVD) represents a heterogeneous group of disorders with different clinical, angiographic, and histopathological features. The most common manifestation of AOFVD is characterized by onset in adulthood, a yellow, round to oval subretinal macular lesion, often with a central pigmented spot, mild loss of visual acuity and normal or mildly abnormal electrooculogram [1–5].

Our study reports on the indocyanine green videoangiography (ICGV) pattern of typical AOFVD.

Materials and methods

Eight patients affected by AOFVD in both eyes were investigated. There were five males and three females with a mean age of 58±15.32 years (median 54.5 years, range 40–80 years). Each patient provided informed consent and underwent ophthalmologic examination including fluorescein angiography (FA) and ICGV.

For FA, 5 ml of a 20% fluorescein solution was injected into the cubital vein. ICGV was performed with 25 mg of ICG dissolved in 5 ml of saline and injected into the cubital vein, followed by 5 ml saline flush. ICGV was performed using the IMAGEnet system (Topcon) as described elsewhere [6].

Results

The mean visual acuity was 0.82±0.13 (median 0.85, range 0.6–1).

Biomicroscopical evaluation showed a symmetric, yellow, round to oval subretinal lesion, one-third to one disc diameter in size, with mildly irregular contours. The yellow material faded gradually from the center of the lesion towards its periphery. In six patients there was evidence of a central pigmented spot in both eyes, whereas in two patients no such spot was visible (Fig. 1). In two eyes several smaller yellow flecks occurred close to the central lesion. Further, in three eyes paracentral drusen were also evident (Fig. 1). There was no evidence of chorioretinal atrophy, chorioretinal inflam-

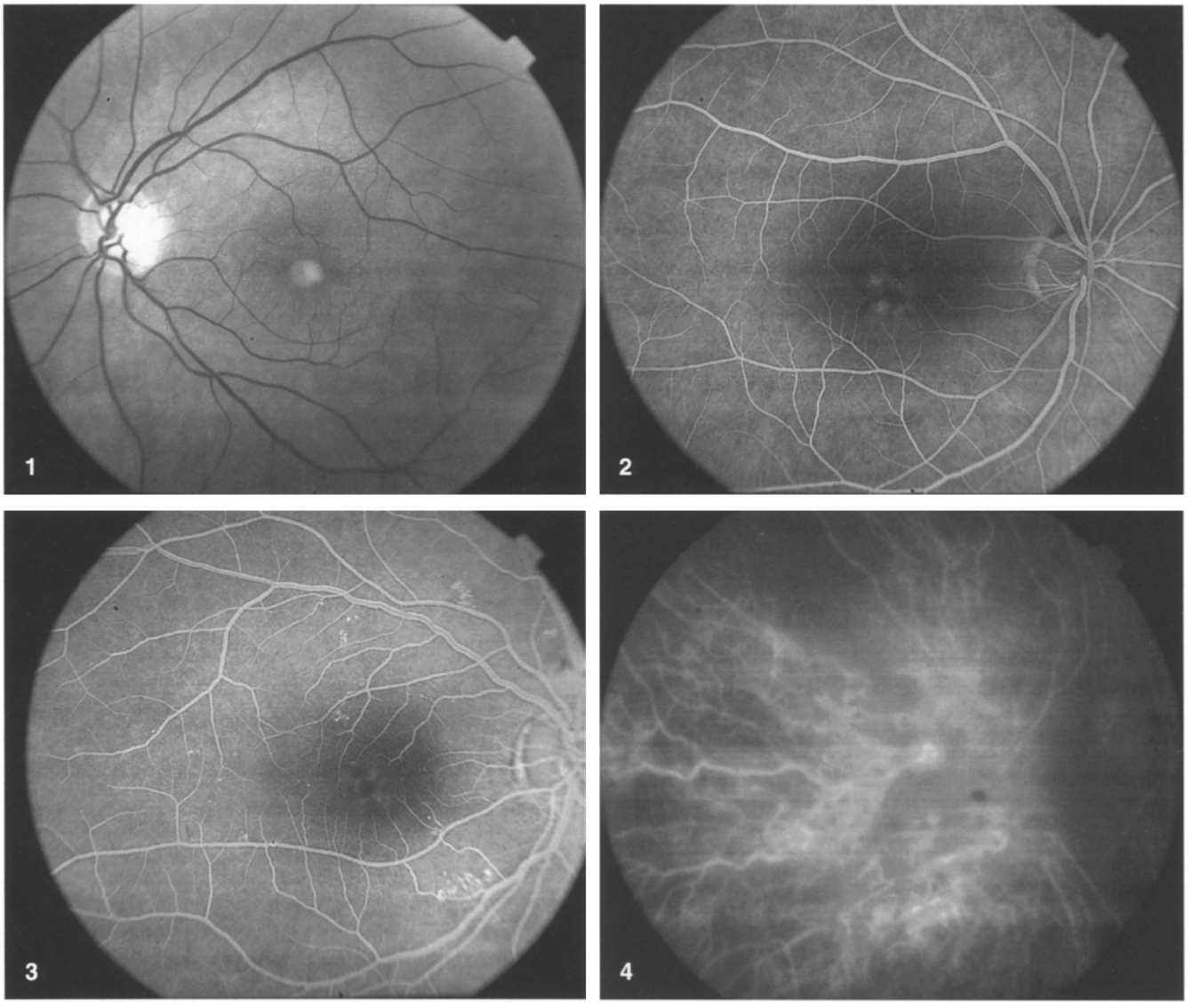


Fig. 1 Case 1. Red-free frame showing the typical central lesion without foveal pigmented spot, associated in this case with paracentral drusen

Fig. 2 Case 2. Fluorescein angiography: central nonfluorescent spot and irregular hyperfluorescent ring

Fig. 3 Case 3. Fluorescein angiography: the typical features and several drusen at the posterior pole

Fig. 4 Case 2. ICGV frame (13 s) showing the central nonfluorescent spot, visible since the first frame

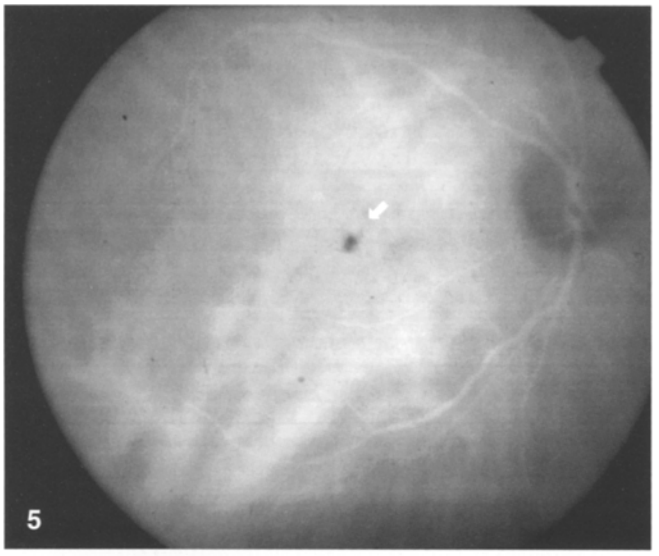
mation, laser scars, fibrosis, neurosensory fluid, or hemorrhage.

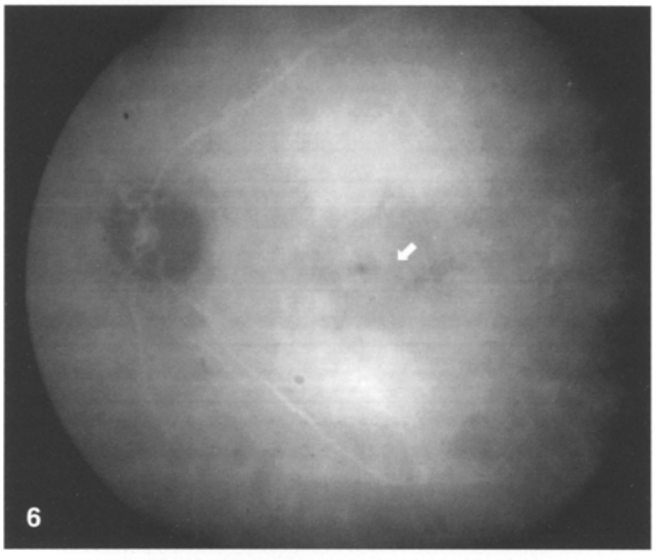
FA demonstrated a central nonfluorescent spot surrounded by an irregular ring of hyperfluorescence (Figs. 2, 3). The yellow flecks biomicroscopically visible near the central lesion in two eyes appeared hypofluorescent on FA.

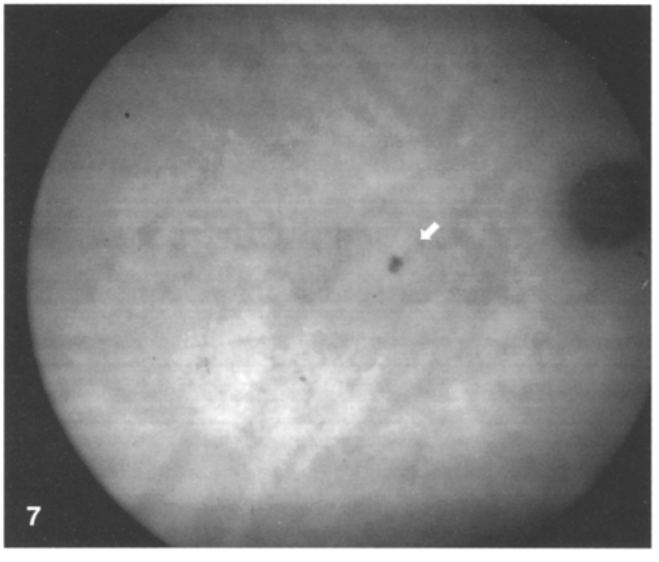
ICGV revealed no abnormality in choroidal perfusion, but a central nonfluorescent spot became evident in the first few frames (Fig. 4). This central dark spot had a round to oval shape and appeared able to mask the underlying choroidal vessels. It appeared unmodified during the whole examination.

At a time varying between 8 and 15 min a delimited, hyperfluorescent, irregularly round area appeared around the central nonfluorescent spot (Fig. 5). Such hyperfluorescence increased up to 20–30 min, maintaining the same shape and size (Fig. 6). In the late phases (30–40 min and more), during progressive decrease of the choroidal fluorescence due to choroidal outflow, the central hyperfluorescence remained visible (Fig. 7). Moreover, the yellow flecks biomicroscopically located close to the central lesion appeared nonfluorescent.

The region mapping function of IMAGEnet showed a correspondence in location between the central pigment-







ed spot visible in the red-free frame and the nonfluorescent spot detected by ICGV. Nevertheless, the extension of the latter appeared greater than that of the former. Moreover, the region mapping demonstrated a correspondence in location between the central yellow lesion visible in the red-free frame and the hyperfluorescent area detected by ICGV. The hyperfluorescent area appeared somewhat larger.

Discussion

In 1974 Gass first described AOFVD in nine patients and reported the histopathological findings in one of them [1]. Further studies considered AOFVD as a heterogeneous group of disorders displaying variability in size, shape, pigmentary deposition, distribution of the primary and accessory lesions, fluorescein angiographic characteristics, choroidal neovascularization, neurosensory detachment, psychophysiology, and genetic mutation [2–5, 7–11].

The variability in appearance of AOFVD corresponded to different histopathological findings. Gass' report referred to a case with a central pigmented spot, corresponding histologically to a large clump of retinal pigment epithelium (RPE) cells in the foveal area with paracentral thinning of the RPE [1].

In 1985 Patrinely et al. described a second case biomicroscopically characterized by the absence of a central pigmented spot [8]. The histopathological features showed RPE atrophy associated with a surrounding zone of RPE hypertrophy and photoreceptor degeneration.

A third case was reported by Jaffe and Schatz in 1988 [9]. The clinical aspect resembled that of Gass' case with the presence of a central pigmented spot. The histological findings demonstrated a foveolar clump of RPE, central photoreceptor degeneration, and central retinal detachment.

Our study included eight patients biomicroscopically characterized by a yellow subretinal lesion with and without central pigmented spot in both eyes. FA in these patients showed the classical central hypofluorescence surrounded by an irregular hyperfluorescent ring. On

Fig. 5 Case 3. ICGV frame: at about 10 min appears a hyperfluorescent area (*arrow*) surrounding the central spot

Fig. 6 Case 1. ICGV frame: at about 23 min the hyperfluorescent lesion increases in intensity, maintaining the same shape and size (*arrow*). Even if not detected by biomicroscopical examination, the central nonfluorescent spot remains visible during the entire examination

Fig. 7 Case 3. ICGV late phase (39 min), showing the persistence of both the central nonfluorescent spot and the surrounding hyperfluorescent area (*arrow*) during the progressive decrease of the choroidal fluorescence

ICGV we noted two main features: the central nonfluorescent spot, which was evident from the first few frames up to the late phases, and a delimited hyperfluorescent area surrounding the central spot.

The central dark spot might be interpreted as the consequence of the RPE clump histologically described by Gass [1] and Jaffe and Schatz [9]. This dense pigment clump may mask choroidal fluorescence. In fact, there was no evidence of choroidal flow abnormality and the central spot covered the choroidal vessels. Moreover, region mapping revealed a correspondence between the central pigmented spot and the nonfluorescent spot in ICGV. The absence of difference in ICGV pattern between cases with and those without central pigmented spot on biomicroscopical examination might hint at the same histological pattern; the central pigment clump may be present in all cases, but sometimes covered by the central yellow material.

As regards the hyperfluorescent area surrounding the central spot, it became visible after only a few minutes. It increased in intensity but not in size, and did not change shape. It was detectable up to the late phases during the reduction of the background fluorescence due to dye outflow.

There are several different possible explanations for this irregular hyperfluorescent area surrounding the central nonfluorescent spot. The existence of a dense eosinophilic material between Bruch's membrane and RPE, as described in the three above-mentioned histopathological studies [1, 8, 9], results in the yellow material seen clinically [1, 9].

This hyperfluorescent area detected during ICGV could perhaps be explained by progressive dye pooling in the space between Bruch's membrane and RPE. FA did not demonstrate dye pooling, but it may have failed to do so because of a masking effect of the entire central lesion, consisting of pigment clump and yellow material.

Alternatively, the hyperfluorescent area may represent progressive staining of the subretinal pigment epithelial material. In particular, it could be hypothesized that the ICG molecule binds to the eosinophilic material described histologically.

Both three explanations could account on the one hand for the progressive increase in hyperfluorescence without modifications in size and shape and on the other for the correspondence between central yellow lesion and hyperfluorescent area on ICGV.

In conclusion, the ICGV pattern of typical AOFVD is represented by a foveal nonfluorescent spot, visible during the entire examination, and a hyperfluorescent area surrounding the central spot, which becomes evident after only a few minutes.

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