Perifoveal exudative vascular anomalous complex

Complexe vasculaire exsudatif perifovéal anormal

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Type 3 neovascularization

Summary
Purpose.— To report the angiographic and optical coherence tomography (OCT) features of isolated “perifoveal exudative vascular anomalous complex (PEVAC)”, a peculiar clinical entity.
Methods.— A complete ophthalmologic examination was performed in two patients (a 82-year old woman [case 1]; a 52-year old man [case 2]) that were referred to our department for unilateral blurred vision.
Results.— In both cases, fundus examination of the right eye showed a perifoveal isolated large aneurismal change, accompanied by small hemorrhages, intraretinal exudation, and small hard exudates accumulation. Both FA and ICGA revealed the absence of any other retinal or choroidal vascular abnormality associated. OCT showed a round hyperreflective lesion in correspondence of the perifoveal vascular anomalous complex, surrounded by intraretinal cystic spaces. In case 2, the lesion remained unchanged despite 3 monthly intravitreal injections of ranibizumab.
Conclusion.— PEVAC may develop in absence of capillary ischemia or inflammation, probably due to progressive retinal endothelial cell degeneration. This could explain the unresponsiveness to anti-VEGF treatments.

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**Introduction**

Perifoveal retinal vascular abnormalities are usually secondary to retinal vascular occlusive or inflammatory disease. Atypical perifoveal retinal vascular abnormalities have been reported also in eyes with drusen and retinal pigment epithelium detachment (PED) [1]. This peculiar aspect of age-related macular degeneration (AMD) was originally labeled as "deep retinal vascular anomalous complex" [2], and than as "retinal angiomatous proliferation" (RAP) [3]. However, perifoveal retinal vascular abnormalities may also develop without a known cause, as it happens for idiopathic macular telangiectasia.

We report the angiographic and optical coherence tomography (OCT) features in two otherwise healthy patients showing isolated perifoveal retinal vascular abnormalities, and we suggest the term "perifoveal exudative vascular anomalous complex (PEVAC)" to characterize this peculiar clinical entity, that does not fit into any other previously described macular disease.

**Report of cases**

**Case 1**

A 82-year old woman was referred to our department in 2004 for blurred vision in her right eye (RE). The patient signed a comprehensive consent form according to good clinical practice guidelines, before proceeding with any examinations and treatments. There was no history of diabetes, hypertension, inflammatory diseases, or blood dyscrasias. Best-corrected visual acuity (BCVA) was 20/80 in RE and 20/20 in the left eye (LE). Fundus examination showed, in RE, a perifoveal isolated large aneurismal aspect, accompanied by small hemorrhages, intraretinal exudation, and hard exudates accumulation (Fig. 1A); a discrete pigment mottling was detected in the LE macula. Both fluorescein angiography (FA) (Fig. 1B) and indocyanine green angiography (ICGA) (Fig. 1C) revealed the absence of any other retinal or choroidal vascular abnormality associated. Time-domain OCT (OCT; OCT3 Stratus, Carl Zeiss, Dublin, CA) showed a round hyperreflective lesion in correspondence of the perifoveal vascular anomalous complex, surrounded by intraretinal cystic spaces (Fig. 1D). No any sign of choroidal neovascularization (CNV) was detected on OCT.

Considering the absence of CNV, and the close proximity to the foveal center, we decided not to treat the patient neither by photodynamic therapy (PDT), nor by laser photocoagulation. When she presented 3 months later for her scheduled follow-up visit, we found overall unchanged findings. Unfortunately, the patient was lost to follow-up.

**Case 2**

A 52-year old man was referred to our department for blurred vision in RE. The patient signed a comprehensive consent form according to good clinical practice guidelines, before proceeding with any examinations and treatments. He had no history of diabetes, hypertension, inflammatory diseases, or blood dyscrasias. BCVA was 20/40 in RE and 20/20 in LE. Fundus examination showed, in RE, a perifoveal isolated large aneurismal change, accompanied by small hemorrhages, intraretinal exudation, and small hard exudates accumulation (Fig. 2A); LE macula was overall normal. Both FA (Fig. 2B) and ICGA (Fig. 2C) revealed the absence of any other retinal or choroidal vascular abnormality associated. Spectral-domain OCT (Spectralis HRA-OCT Heidelberg Engineering, Heidelberg, Germany) showed a round hyperreflective lesion in correspondence of the perifoveal
Figure 1. Fundus color photograph (A) of the right eye of case 1 show a perifoveal isolated large aneurismal change, accompanied by small hemorrhages, intraretinal exudation, and small hard exudates accumulation. Both fluorescein angiography (B; B1, enlarged view; B2, peripheral frame) and indocyanine green angiography (C) reveal the perifoveal exudative vascular anomalous complex and the absence of any other retinal or choroidal vascular abnormality associated. Optical coherence tomography scan (D) shows a round hyperreflective lesion in correspondence of the perifoveal vascular anomalous complex, surrounded by intraretinal cystic spaces.

Due to the exudative nature of the isolated perifoveal complex, we decided to submit the patients to 3 monthly intravitreal injections of ranibizumab (0.05 ml/0.5 mg). One month after the third intravitreal injections, fundus examination, FA, ICGA and OCT showed the persistence of the perifoveal vascular anomalous complex, without still any other retinal or choroidal vascular abnormality associated.

Figure 2. Fluorescein angiography (A; B, enlarged view) and indocyanine green angiography (C, early phase; D, late phase) frames of the right eye of case 2 reveal the perifoveal exudative vascular anomalous complex and the absence of any other retinal or choroidal vascular abnormality associated. Spectral-domain optical coherence tomography scans (C and D) show a round hyperreflective lesion in correspondence of the perifoveal vascular anomalous complex, surrounded by intraretinal cystic spaces.
The exudative retinal changes appeared also unchanged, despite the 3 intravitreal injections of ranibizumab.

Comment

The main causative factors for perifoveal retinal vascular abnormalities are diabetic retinopathy, hypertension, venous occlusion, inflammatory diseases, and blood dyscrasias. Type 1 macular telangiectasia, also known as aneurysmal telangiectasia [4], is an idiopathic macular telangiectasia that usually affects young patients, and is characterized by multiple capillary, venular, and arteriolar aneurysms. A profound exudation is constantly observed, and some patient shows minimal, patchy nonperfusion or capillary ischemia and lipid deposition.

Recently, Bourhis et al. [5] reported on the ICGA and OCT features of capillary and/or venous macroaneurysms secondary to retinal vein occlusion and diabetic retinopathy. In their series, ICGA provided a better delineation of macroaneurysms than FA, and OCT identified macroaneurysms under the form of a vascular structure with a reflective wall surrounding a lumen containing variably reflective material. Similar behaviors on both ICGA and OCT was detected in our two cases affected with PEVAC. However, while in the cases reported by Bourhis et al. [5], macroaneurysms were multiple and several macroaneurysms were seen to be closely located, in our cases, PEVAC was seen as an isolated (single and not associated with microaneurysms) perifoveal large aneurysmal change. Moreover, the two cases reported here were both otherwise healthy patients that did not show evidence of arterial hypertension, diabetes or any other vasculopathy. Interestingly, they differed in genders (Case 1 was a female patient, and case 2 was a male patient), and ages of onset of ocular symptoms (82 years vs. 52 years, for case 1 and case 2, respectively). The presence of an isolated PEVAC, in absence of retinal vascular occlusive or inflammatory disease, was the consistent feature showed by these two patients. Of note, PEVAC was far different from Type 1 idiopathic macular telangiectasia, and the two patients did not show any sign of AMD, that would suggest the diagnosis of stage 1 RAP [3]. Moreover, differently from exudative microangiopathies secondary to ischemia or inflammation, or type 1 idiopathic macular telangiectasia [6], PEVAC seems to be unresponsive to anti-VEGF treatments. We propose that, in absence of capillary ischemia or inflammation, progressive retinal endothelial cell degeneration may be the triggering, vasogenic cellular mechanism for PEVAC; this could explain the unresponsiveness to anti-VEGF treatments.

The description of more cases of PEVAC, as well as studies on the ‘natural history’ of these lesions, would help in understanding the physiopathology of such an unusual maculopathy.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References