CASE REPORT

Idiopathic polypoidal choroidal vasculopathy

Sonja Cekić*, Dijana Risimić†, Ivan Jovanović‡, Jasmina Djordjević Jocić*

*Clinical Centre Niš, Eye Clinic, Niš, Serbia; †Clinical Centre of Serbia, Clinic for Eye Diseases, Belgrade, Serbia; ‡Medical Faculty, University of Niš, Department for Anatomy, Niš, Serbia

Abstract

Background. Idiopathic polypoidal choroidal vasculopathy (IPCV) is uncommon condition. It is considered to be a variant of neovascular age-related macular degeneration, but it can be also found in younger patients. Case report. We presented a case of otherwise healthy, 36-year-old women, with sudden unilateral visual impairment in the left eye and metamorphosia. Slit lamp biomicroscopy examination of the eye anterior segment was normal. Intraocular pressure determined by aplanation tonometry was 16 mmHg in both eyes. Indirect slit lamp biomicroscopy examination showed signs of serosanquinous detachments of the retinal pigment epithelium. Fluorescein angiography showed a subretinal vessel network through the pigment epithelial atrophy with hyperfluorescence in superior part of serohemorrhagic pigment epithelial detachment and the inferior hypofluorescence, caused by hemorrhage. Optical coherence tomography proved detachment of the retinal pigment epithelium.

Conclusion. In patients with IPCV a mild, natural course with spontaneous resorption of exudations and hemorrhage and improvement in visual acuity can be observed. There is no approved treatment at present.

Key words: choroid diseases; macular degeneration; tomography, optical coherence; fluorescein angiography; diagnosis; diagnosis, differential; choroid hemorrhage; prognosis.


Ključne reči: sudovnača, bolesti; makula, degeneracija; optička koherentna tomografija; fluoresceminska angiografija; dijagnoza; dijagnoza, diferencijalna; sudovnača, krvenjenje; prognoza.
The precise definition of the condition – whether it constitutes a subset of AMD, is associated with it, or represents a completely different entity – is currently a matter of debate.

Some novel studies evaluate the differences in the optical coherence tomographically (OCT) determined macular morphology between the eyes with IPCV and those with age-related macular degeneration (AMD). Eyes with IPCV are characterized by a higher incidence of serous retinal detachment (SRDs), greater SRD height, and less intraretinal edema than eyes with exudative AMD\textsuperscript{10,11}.

The aim of our study was to present a case of young, otherwise healthy woman, with sudden, unilateral visual impairment in the left eye and metamorphopsia.

Case report

We presented a case of 35-years-old, otherwise healthy woman with sudden visual loss in the left eye and metamorphopsia in the same eye. Visual acuity, with a Snellen chart in the right eye was 1.0 and the left eye 0.3.

Slit lamp biomicroscopy examination of anterior segment of the eye was normal. Intraocular pressure, determined by aplanation tonometry, was 16 mmHg in both eyes. Indirect slit lamp biomicroscopy examination, by Volk Super Field NC Lenses, showed signs of serosanguineous detachments of the retinal pigment epithelium (Figure 1).

Fluorescein angiography (FA), with a fundus camera Zeiss Visucam Lite, showed a subretinal vessel network through the pigment epithelial atrophy with hyperfluorescence in superior part of serohemorrhagic pigment epithelial detachment and the inferior hypofluorescence, caused by the hemorrhage (Figure 1).

Optical coherence tomography showed double reflective layers that consisted of RPE and another highly reflective layer beneath the RPE ("double-layer sign") in the area of the branching network vessels (Figure 2a) (Topcon 3D OCT-1000).

After 3 months, visual acuity in the left eye was 0.5 by Snellen chart. Indirect slit lamp biomicroscopy examination, by Volk Super Field NC Lenses, showed signs of small, round, orange subretinal elevation that accords to serous detachments of the retinal pigment epithelium (Figure 3).

Optical coherence tomography was without any significant changes (Figure 2b).

After 4 months a new hemorrhage occurred again with vision acuity to 0.1 by Snellen’s chart. We performed laser photocoagulation in the affected area, number of spots 29, power 70 and duration 1.2 µm (Figure 4).

Fig. 1 – Fundus photo and fluorescein angiography (FA, fundus camera Zeiss Visucam Lite)

a) Native fundus photo of the right eye; b) Native fundus photo of the left eye; c–f) FA of the left eye
Fig. 2 – Optical coherence tomography

Fig. 3 – Fundus photo and fluorescein angiography after three months

Fig. 4 – Fundus photo after laser coagulation

Visual acuity on the next visit after 2 weeks was 0.7 uncorrected visual acuity (UCVA) by a Snellen `chart.

Discussion

Idiopathic polypoidal choroidal vasculopathy, also known as posterior uveal bleeding syndrome or multiple serosanguineous retinal pigment (RPE) detachment syndrome, is a rare condition. It encompasses approximately 8% of new patients diagnosed with neovascular AMD and is found more commonly in patients of African and Asian origin 12. Its appearance in young patients is rare, especially in Caucasians as in our case.

In the pathogenesis an abnormality of the inner choroidal vessels consisting of a dilated network and multiple terminal aneurismal protruberence in a polypoidal configuration that have a predilection for the macula, as in presented case, and less frequently the peripapillary area, is a key factor for all further clinical changes 10, 13.

Others presumed that retinal microangiopathy may occur in a chronic macular detachment secondary to polypoidal choroidal neovascularization. The development of these secondary retinal changes is not clearly understood; however, hypoxia from chronic detachment, a neurotoxic effect from a lipid deposition, or a biochemically induced microvascular abnormality from secretion of vasogenic mediators are possible mechanisms 14.

Loss of visual acuity originates from subfoveal exudations and hemorrhage. In contrast to pure classic or occult choroidal neovascularisation (CNV) secondary to AMD, spontaneous recovery can be observed in a significant proportion of cases 12. Also, eyes with IPCV are characterized by a higher incidence of serous retinal detachment (SRDs), greater SRD height, and less intraretinal edema than eyes with exudative AMD 11, 13.

In the presented patient for a period of almost 3 months of following, visual acuity from a 0.3 by Snellen´ chart came to 0.7 by Snellen´ chart. The recovery was spontaneous.

After 7 months of the first visit, a new hemorrhage occurred again with vision acuity of 0.1 by Snellen´s chart. We preformed laser photocoagulation in the affected area. The number of spots was 29, power 70 and duration 1.2 μm.

Visual acuity on the next visit after 2 weeks was 0.7 without correction by Snellen´ chart.

Optical coherence tomography showed double reflective layers that consisted of RPE and another highly reflective layer beneath the RPE ("double-layer sign") in the area of the branching network vessels (Figure 2a) (Topcon 3D OCT-1000).

In PCV, a double-layer sign is seen frequently in the area of the network vessels, particularly in eyes with a serous retinal detachment. The sign may reflect fluid accumulation between RPE and Bruch membrane resulting from the network of abnormal vessels leakage.

There is no approved treatment at present 15. The localisation of lesion determined terapeutical approach. IPCV lesions were found in the extrafoveal area (63%), subfoveal (29.7%), jyxta foveal (15.9%) and peripapillary (18%) area 9.

Patients with extramacular lesions without leakage into the fovea should be observed closely.

Laser coagulation of the extrafoveal polypidal lesions can be carried out if the leakage threatens the fovea.

Laser coagulation is a therapy that gave a good result in the presented case.

Photodynamic therapy (PDT), macular surgery and intravitreal injection of anti-VEGF substances can be attempted in subfoveal lesions or in progressive macular edema after laser coagulation16–18.

Conclusion

Subretinal hemorrhage after PDT can be a common complication in patients with PCV and may have a minor effect on visual outcome. However, postoperative hemorrhage is occasionally so massive to lead to vitreous hemorrhage and poor visual prognosis. When considering PDT for eyes with a large PCV lesion, ophthalmologists should be aware of the risk of serious hemorrhagic complications.

References


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