

Relentless placoid chorioretinitis – A case report

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SUMMARY

Introduction Relentless placoid chorioretinitis is an entity which belongs to the group of an atypical intermediate form of primary inflammatory choriocapillaropathies, resembling both acute posterior multifocal placoid pigment epitheliopathy and serpiginous choroiditis, but the retinal distribution and clinical course are not the same. Because of this similarity this entity was termed "AMPPiginous". This entity was first described by Jones et al. in 2000. The aim of our case report is to present a very specific case where the clinical course was progressive, with loss of vision in the affected eye.

Case Outline A 31-year-old man, with no previous ophthalmic diseases, was hospitalized at the Clinic of Ophthalmology, Clinical Center Kragujevac, because of a reduction of vision in the right eye, and scotoma and metamorphopsia in the left eye. The clinical course of retinal lesions in the left eye resembled the changes observed in acute posterior multifocal placoid pigment epitheliopathy, and the right eye changes were between acute posterior multifocal placoid pigment epitheliopathy and serpiginous choroiditis. The diagnosis of relentless placoid chorioretinitis was confirmed after clinical, laboratory, immunological, virological, and angiography examinations.

Conclusion The progressive clinical course of the disease, complemented by multimodal imaging and extensive laboratory diagnostics, has led us to the diagnosis of relentless placoid chorioretinitis. The combined anti-inflammatory and immunomodulatory therapy led to the stabilization of visual acuity of the left eye as opposed to the right, where there has been no recovery.

Keywords: anti-inflammatory therapy; immunomodulatory therapy; multimodal imaging; primary inflammatory choriocapillaropathies

INTRODUCTION

Relentless placoid chorioretinitis (RPC) is a relatively new and rare entity, which belongs to the atypical intermediate form of primary inflammatory choriocapillaropathies (PICCP), which was first described in 2000 by Jones et al. [1]. RPC is an entity in which multiple inflammatory, deep white-creamy lesions resembling those seen in acute posterior multifocal placoid pigment epitheliopathy (APMPPE) and serpiginous choroiditis (SC) develop. Unlike APMPPE, the lesions in RPC continue to expand in size and number with a relentless course over many months. Unlike SC, the lesions of RPC are multifocal and eventually involve all areas of the retina, including the anterior periphery prior to involvement of the posterior pole and macula. Therefore, RPC belongs to the group of an atypical intermediate form of PICCP [2]. PICCP is a heterogeneous group of diseases with a common pathogenetic mechanism – choriocapillaris non-perfusion with subsequent retinal ischemia above, which depends on oxygen and nutrients from choriocapillaris. Clinical differences in PICCP are explained by the level and severity of inflammatory insults at the level of choriocapillaris circulation [2]. Etiology is undefined; some suggest an autoimmune/inflammatory cause triggered by an exogenous agent [3]. Biochemi-

cal and serological examinations complemented by multimodal imaging and functional parameters facilitate diagnosis and management [4]. Multimodal imaging includes fluorescein angiography (FA), indocyanine green angiography (ICGA), fundus autofluorescence (FAF), and optical coherence tomography (OCT). FA provides information regarding the localization and extent of posterior segment inflammatory diseases [4]. ICGA is dominant to diagnose diseases of choriocapillaris, as well as for monitoring course of disease and effect of therapy [5]. OCT could help in the differential diagnosis of RPC, because RPC may rarely lead to severe macular atrophy [6]. Therapy should be adjusted to the course of inflammatory diseases because we know the structures that are primarily affected. The treatment includes hospitalization, patient monitoring, with anti-inflammatory [7], immunomodulatory [8–12], and anti-angiogenic therapy [13]. Most patients treated with prolonged systemic corticosteroid therapy recover their prior vision. However, maintenance of remission is possible if immunosuppressive therapy is involved in the early stages of the disease [2]. Also, vision can decrease significantly when fovea is involved [14]. Complications of RPC could be choroidal neovascularization, subretinal fibrosis, and epiretinal membrane formation [15].

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CASE REPORT

A 31-year-old male was hospitalized complaining of decreased vision in his right eye for the previous two days. The patient denied any previous ophthalmic diseases, febrile or flu-like episodes. On initial examination, visual acuity was 0.05 on the right eye and 1.0 (Snellen charts) on the left. Color vision (Ishihara plates) on the left was preserved. Intraocular pressure in both eyes was 16 mmHg. The anterior segments were quiet, and there were no vitreous cells. Fundus examination (Visucam, Carl Zeiss AG, Oberkochen, Germany) showed lesions characterized as multiple creamy-white at the level of choriocapillaris, with an inflamed retina above in the mid periphery, posterior pole and prae-equatorial, but in the right eye the inflammation was a damaged macula. On the right eye, the perimetry (central 30-2 and macular threshold, Humphrey Systems Instruments, Carl Zeiss AG) showed an absolute absence of sensitivity and, on the left, objective, multifocal scotoma decreased sensitivity. A fluorescein angiogram (Visucam FA, Carl Zeiss AG) showed an area of subretinal placoid lesions characterized in the early stages by visible hypofluorescence, which indicates choriocapillaris non-perfusion, and in the late stage of progress show hyperfluorescence, depending on the severity of the ischemic process (Figure 1) (unlike APMPE, active lesions do not show blocked fluorescence). The previously described changes were observed in both eyes in the region of the equatorial zone; in the right, the changes had much more extensive involvement of the macula. Visual evoked potentials showed the left eye had a proper cortical response, while cortical response was not generated in the right eye. Optical coherence tomography (Cirrus OCT, Carl Zeiss Meditec AG, Jena, Germany) of the right eye presented macular atrophy [central macular thickness (CMT) was 142 μm], and left eye thickness of the retinal pigment epithelium and mild retinal pigment epithelium detachment in the macula (CMT was 282 μm). Biochemical, antinuclear factor, rheumatoid factor, angiotensin-converting enzyme, serum lysozyme levels, and lupus anticoagulant were all normal. Serological tests on syphilis, human immunodeficiency virus, varicella zoster virus, herpes simplex virus, mycoplasma pneumoniae, cytomegalovirus, toxoplasmosis, rubella, *Borrelia burgdorferi* were within reference ranges. Chest X-rays were normal and tuberculin skin test (Mantoux test) was negative. After the previous examination, the patient was diagnosed with RPC in the right eye, and with APMPE in the left. The patient was prescribed parenteral corticosteroid therapy (1 mg/kg/day). Only three days after the hospitalization, the disease progression was as follows: visual acuity amaurosis on the right, and best corrected visual acuity on the left eye was 0.8. Hence, we administered cyclosporine (5 mg/kg) as well, after which best corrected visual acuity on the left eye was 1.0, but on the opposite eye the amaurosis remained because the macula and mid-periphery of the retina was already damaged. Characteristic local findings as well as progressive, almost aggressive course of the disease, suggest a rare clinical syndrome – a relentless placoid chorioretinitis. We monitored the patient



Figure 1. Image shows multiple areas of hyperfluorescence in the mid-periphery and posterior pole, capturing the macula, only three days after hospitalization

monthly during the first six months, and since there was no progression of the disease on the opposite eye, we reduced cyclosporine to a dose of 2 mg/kg after six months. Regular two-month control check-ups were performed for the next six months. Local funduscopic findings and findings obtained by the fundus camera remained unchanged. The patient had no side effects to the drug, but because of the stable fundus, we discontinued cyclosporine administration. After 18 months from the disease onset, funduscopic findings of both eyes remained unchanged.

DISCUSSION

PICCP are caused by an inflammation producing choriocapillaris non-perfusion and show the characteristic local retinal and subretinal lesions on fundus examination. The differential diagnosis includes some of the forms of PICCP and some systemic diseases. Unlike APMPE, the lesions in RPC continue to expand in size and number, while lesions in APMPE are reversible and without scars **sans** immunosuppressive therapy. In contrast to SC, the lesions of RPC are multifocal and involve all areas of the retina, including the anterior periphery prior to involvement of the posterior pole and macula [2]. The progressive outer retinal necrosis syndrome in the early stages of the disease is characterized by multifocal deep retinal lesions. However, lesions rapidly progress to total retinal necrosis with retinal detachment, and laboratory findings on herpes zoster virus was negative [16, 17]. Ocular ischemic syndrome (choroidal ischemia) is a rare condition associated with severe artery occlusive disease leading to ocular hypoperfusion. Choroidal hypoperfusion results with lesions resembling those in RPC in the posterior pole or the mid-periphery. However, prolonged ischemia results in retinal detachment and retinal atrophy [18]. Infectious ocular diseases like acute retinal necrosis [17], systemic diseases like systemic lupus erythematosus [19], AIDS [20], and non-Hodgkin's lymphoma [21] may have similar fundoscopic lesions, but all of them have vitritis and/or mild or severe anterior uveitis, as well as laboratory analyses results beyond reference values, which differentiates them

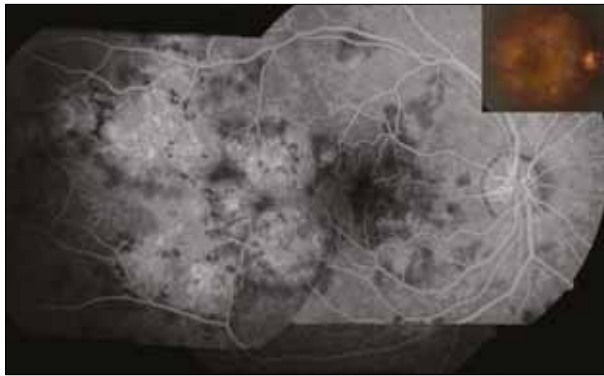


Figure 2. Fundus examination of the left eye presents multiple creamy-white lesions at the level of choriocapillaris

from RPC. Characteristic local findings which were confirmed with additional diagnostic tests, as well as a progressive, almost aggressive course of the disease indicate a clinical syndrome – a relentless placoid chorioretinitis. Application of corticosteroid therapy alone has not proved as effective as expected. Among patients where the disease progresses rapidly, as was the case with our patient, with some complications present, or if vision is threatened, the

addition of immunosuppressive therapy may lead to the stabilization of the disease. Combined, anti-inflammatory and immunosuppressive therapy should be administered for a sufficiently long period of time, as early discontinuation of such therapy can lead to the reactivation of the disease. But in the described case even the combined and long-term therapy of 12 months did not lead to an improvement because the macula was damaged, which led to permanent loss of vision on the right eye; however, we managed to stop the process in the left eye (Figure 2).

NOTE

The paper has been presented at the 1st Congress of Ophthalmologists of Republic of Srpska with International Participation, Bijeljina, 29th to 31st of May 2015, where it has been published in the form of an abstract (Jovanović S, Obradović Lj, Jovanović M, Jovanović Z. A case of ampiginous choriocapillaritis syndromes – Case report. Proceedings and Abstract Book of the 1st Congress of Ophthalmologists of the Republic of Srpska with International Participation. Bijeljina; p. 156–7.

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Рефракторни плакоидни хориоретинитис – приказ болесника

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КРАТАК САДРЖАЈ

Увод Рефракторни плакоидни хориоретинитис је ентитет који припада групи атипичних интермедијарних форми примарних инфламаторних хориокапиларопатија, подсећајући и на акутни задњи мултифокални плакоидни пигментни хориоретинитис и на серпигиозни хориоретинитис, али ретинална дистрибуција и клинички ток нису исти. Због такве сличности овај ентитет се може наћи у литератури и под термином „ампигиозни“. Описан је први пут од Џона и осталих. Циљ приказа случаја је веома специфичан случај јер је клинички ток био прогресиван са губитком видне функције.

Приказ болесника 31-годишњи мушкарац, без ранијих офталмолошких обољења, хоспитализован је на Клинику за офталмологију Клиничког центра Крагујевац због пада вида на десном оку, и са скотомима и метаморфопсијом на левом оку. Клинички ток ретиналних лезија на левом оку личио је на промене описане код акутне постериорне мултифокал-

не пигментне епителопатије, а на десном оку промене су биле између акутне постериорне мултифокалне пигментне плакоидне епителопатије и серпигиозног хороидитиса. Дијагноза рефракторног плакоидног хориоретинитиса је потврђена након клиничких, лабораторијских, вирусолошких и ангиографских испитивања.

Закључак Прогресиван клинички ток болести, уопуњен мултимодијалним имицингом и опсежном лабораторијском дијагностиком, довели су нас до дијагнозе рефракторног плакоидног хориоретинитиса. Комбинована антиинфламаторна и имуномодулаторна терапија су довеле до стабилизације видне функције левог ока за разлику од десног, где није дошло до опоравка.

Кључне речи: антиинфламаторна терапија; имуномодулаторна терапија; мултимодијални имицинг; примарне инфламаторне хориокапиларопатије

Примљен • Received: 22/12/2015

Ревизија • Revision: 27/04/2016

Прихваћен • Accepted: 15/06/2016