Indocyanine Green Angiography and Scanning Laser Ophthalmoscope Microperimetry in Punctate Inner Choroidopathy: Case Report

Punktat İç Koroidopatide İndosiyanin Yeşili Anjiyografisi ve Scanning Lazer Oftalmoskopi Mikroperimetresi

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Geliş Tarihi/*Received:* 06.12.2008
Kabul Tarihi/*Accepted:* 30.03.2009

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ABSTRACT Our purpose is to describe indocyanine green angiography (ICG) and scanning laser ophthalmoscopy (SLO) findings in a patient with punctuate inner choroidopathy (PIC). The clinical record of a patient with PIC evaluated at the National Eye Institute (NEI) using fluorescein angiography (FA), ICG and SLO was reviewed. ICG showed hypofluorescent spots in early and late phases that corresponded to areas of leakage in FA. These hypofluorescent spots disappeared following immunosuppressive treatment. SLO mapped the location of scotomas that correlated to FA and ICG. ICG and SLO can be useful adjuncts to FA and clinical exam in assessing disease activity and guiding immunosuppressive therapy in patients with PIC.

Key Words: Indocyanine green; choroid; uveitis, posterior

ÖZET Bu yazıda amacımız punktat iç koroidopatili bir hastada indosiyanin yeşili anjiyografisi (İYA) ve tarayıcı lazer oftalmoskopisi (SLO) bulgularını tanımlamaktır. Punktat iç koroidopati teşhisi ile Ulusal Göz Enstitüsü (NEI)'nde takip edilen bir hastanın klinik bilgileri, floresein anjiyografi (FA), İYA ve SLO bulguları incelendi. İYA'da, FA'da görülen sızıntı bölgelerinde, hipofloresan alanlar izlendi. Bu hipofloresan noktaların, immünsüpresif tedavi sonrası kaybolduğu görüldü. SLO mikroperimetresinde tespit edilen skotomların lokalizasyonu, FA ve İYA ile uyumluluk gösterdi. İYA ve SLO, punktat iç koroidopatili hastalarda, hastalığın aktivitesinin takibinde ve buna bağlı olarak immünsüpresif tedavinin yönlendirilmesinde, FA'ya yardımcı olacak faydalı araçlardır.

Anahtar Kelimeler: İndosiyanin yeşili; koroid; posterior üveit

Turkiye Klinikleri J Med Sci 2009;29(2):536-9

The term punctate inner choroidopathy (PIC) was first described by Watzke and Packer et al in 1984 although similar cases have been described by Doran and Hamilton previously.[1,2] It is an inflammatory chorioretinal disease occurring mostly in healthy young women with myopia. Patients present with acute scotoma and photopsias and on examination small yellow spots (100-300 micron) localized in the retinal pigment epithelium (RPE) and inner choroid are observed. The lesions, that are generally limited to the posterior pole, look similar in appearance to multifocal choroiditis but there is no inflammation of the vitreous or anterior chamber. Resolution of symptoms and evolution of lesions into atrophic scars typically occur in the course of several weeks. Visual acuity is only

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moderately affected unless a subfoveal neovascularization develops, which may occur in approximately 25-40% of eyes as has been described by Watzke and Packer et al, Brown and Folk and Gerstenblith and Thorne et al.[1,3,4]

Different ICG angiographic characteristics as both-hypofluorescent and hyperfluorescent have been described in patients with PIC both. However anatomic and functional changes in PIC have never been correlated using SLO-microperimetry. Herein, we present the ICG and SLO (SLO 101, Rodenstock, Ottobrunn, Germany) characteristics of a patient with active PIC in correlation to FA and clinical findings before and after treatment.

CASE REPORT

A 26-year-old white female with myopia (-4.00 sphere) who noted paracentral relative scotoma in the right eye (OD) with sudden drop in visual acuity following a severe sinusitis 3 years prior to her presentation was given a presumptive diagnosis of acute posterior multifocal placoid pigment epitheliopathy with no treatment. Her visual acuity at that time was 20/40 in the right eye and 20/20 in

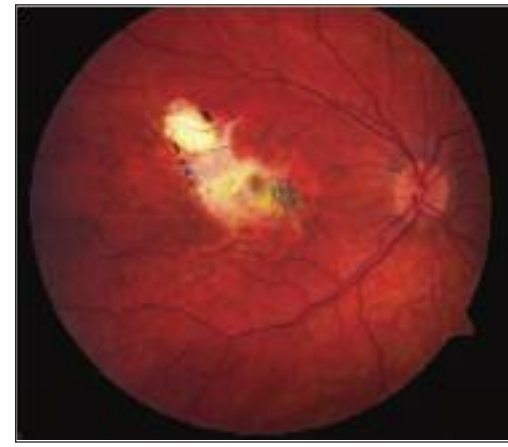

FIGURE 1: Color photo of the right eye showing a large macular scar.

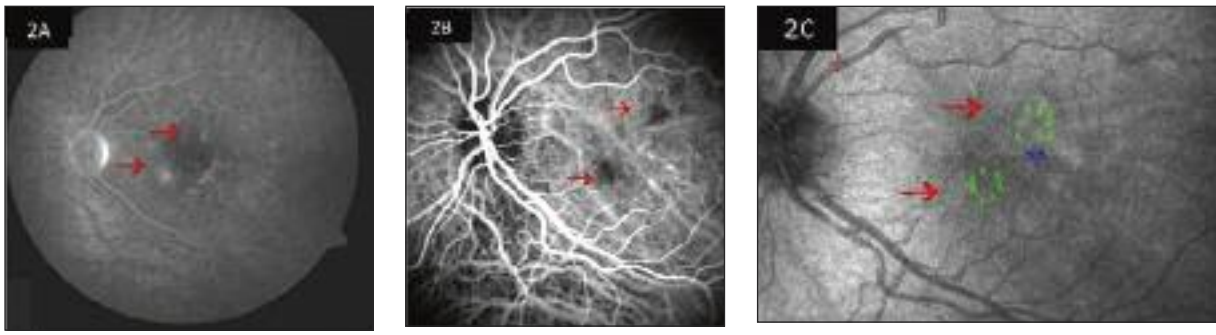

FIGURE 2: Fluorescein angiography of the patient during active disease shows 2 foci of hyperfluorescence (2A) corresponding to the 2 areas of hypofluorescence on ICG (2B) and deep scotoma seen on SLO-microperimetry (2C). The area inferonasal to fovea is a deeper scotoma than the area superior to fovea (a higher letter in alphabetical order indicates a shallower scotoma, thus, letter "A" indicates a deeper scotoma than "K" and "K" deeper than "U"). Note that a more subtle leakage on FA (superior to fovea) leads to a shallower scotoma.

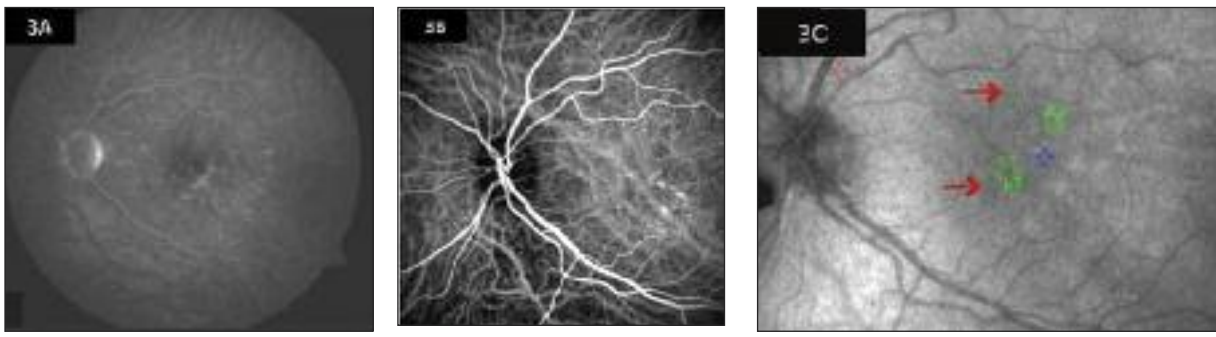

FIGURE 3: Following immunosuppressive treatment resolution of hyperfluorescence on FA (3A) and hypoflorescence on ICG (3B) was accompanied by improvement in the 2 scotomas (higher sensitivity as indicated by letter "U") on SLO-microperimetry (3C).

the left eye (OS). She subsequently developed submacular hemorrhage with choroidal neovascular membrane (CNVM) and underwent surgical removal of the CNVM with resultant macular scar and a vision of 20/60 OD. Soon after she developed multiple paracentral scotomas and photopsias OS with visual acuity of 20/30 OS. She was diagnosed with PIC, treated intermittently with oral prednisone and had a relatively stable course for approximately 3 years. She was referred to NEI with recurrent worsening of paracentral scotomas and photopsias OS. On exam at NEI her visual acuity was 20/30 OD, and 20/20 OS. Her intraocular pressures were within normal limits bilaterally. The ophthalmic examination showed completely quiet anterior chamber (AC) and vitreous, a macular scar (1 x 2.5 DD disc diameter) OD and mild RPE changes around the fovea OS (Figure 1). FA OS showed 2 foci of leakage in mid to late phases in the macula; one inferonasal to the fovea and another more subtle leakage superior to the fovea (Fig 2A). ICG using Heidelberg retina angiography showed hypofluorescence in the corresponding areas (Figure 2B). An SLO at the same time showed decreased sensitivity (deep scotomata) corresponding to the 2 spots of leakage on FA and hypofluorescence on ICG (Figure 2C). She was treated with prednisone (60 mg/day) and a periocular steroid injection in the left eye with resolution of symptoms over 3 weeks. A repeat FA and ICG in 3 weeks showed no leakage or blockage respectively (Figures 3A and 3B) and the SLO revealed an improvement in the depth of the corresponding scotomas (Figure 3C). She was eventually started on methotrexate as steroid-sparing agent. Her total follow-up at NEI was 2 years since her first presentation at NEI.

DISCUSSION

Punctate inner choroidopathy is an inflammatory chorioretinal disease occurring mostly in healthy young women with myopia. Patients with PIC originally described by Watzke and Packer et al did not have any clinically evident intraocular inflammation.[1] Many authors including Brown and Folk and Jampol and Wiredu feel that the cases described in various series in the literature represent different presentations of a single disease process.[3,5]

Blind spot enlargement is often seen in MEWDS but can also be seen in MFC and PIC. Brown and Folk et al indicated that patients with PIC often have visual field defects and ERG abnormalities much larger than would be expected from the number of visible chorioretinal lesions.[3] De Meyer and Lafaut et al believe that focal areas of inflammatory activity as well as secondary choroidal neovascularization may cause leakage in fluorescein angiography.[6] The FA leakage in our patient was attributable to inflammatory activity, however the presence of a choroidal neovascular membrane should always be ruled out.

Serous detachment over the choroidal lesions has been observed by Brown and Folk et al in some patients with PIC and this may be one explanation for the leakage on FA.[1] In our patient thickening of the overlying retina as evidenced by diffuse retinal edema on OCT during the active disease that resolved following treatment was seen.

According to Slakter and Giovannini et al in PIC, hypofluorescence on ICG may be seen not only at the site of visible fundus lesions but also in normal-appearing areas, indicating additional areas of choroidal lesions.[7] Bouchenaki and Cimino et al showed that ICG findings in diseases with primary inflammatory choriocapillaropathy (i.e MEWDS, APMPPE, serpiginous choroidopathy) consisted of hypofluorescent areas up to the late phase of angiography characteristic for choriocapillaris non-perfusion sparing larger stromal choroidal vasculature.[8] Akman and Kadayifcilar et al and Tiffin and Maini et al also reported similar findings in ICG and indicated that larger choroidal vessels running through the hypofluorescent areas suggest that vasculitis may be confined to small choroidal vessels.[9,10] However in our patient, the hypofluorescent areas were relatively well-defined and appeared as dark "punched-out" areas. We believe that these hypofluorescent areas represent localized choroidal hypoperfusion secondary to an inflammatory process. It remains unclear whether these hypofluorescent spots on ICG angiography are specific or can occur in any inflammatory disease that affects the choro-

id. Concurrent presence of hyperfluorescence (leakage) on FA suggests that inflammatory process in choriocapillaris involves deep retinal vasculature as well. Whether this is due to an intraretinal microangiopathy secondary to inflammation or a simultaneous compromise in RPE/outer blood-retina barrier is unclear. Cimino and Auer et al reported on correlation between ICG findings and visual function in inflammatory choriocapillaropathies and their value in follow-up.[11] Rohrschneider and Bültmann et al showed that fundus perimetry with SLO allows for an accurate correlation between morphologic alteration and functional impairment.[12] In our patient, with resolution of active lesions these hypofluorescent areas on ICG disappeared. At the same time leakage on FA disappeared and the depth of scotomas on SLO decreased. Interestingly, SLO was sensitive enough to show the difference in severity in 2 foci of leakage on FA with one deeper and one shallower scotoma. The fact that there were still scotomas, albeit shallow, despite the disappearance of leakage and hypofluorescence on FA and ICG respectively may indicate that SLO may be more sensitive to the changes in RPE and photoreceptors that occur as a result of the inflammatory process in PIC.

With this patient we were able to confirm improvement in function using SLO following treatment that corresponded to improvement in fluorescein angiogram and ICG changes. Although our experience with SLO in management and follow-up of PIC is limited; we believe that it may be a useful adjunct to FA and ICG in the diagnosis and management of PIC and, possibly of other white dot syndromes.

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