

Fundus Imaging Findings and Management of Idiopathic Juxtafoveal Telangiectasia

İdiopatik Jukstafoveal Telenjektazide Fundus Görüntülemesi Bulguları ve Tedavisi

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Cose Report

Olgu Sunumu

ABSTRACT

The fundus imaging findings and the management of a case with type 2 idiopathic juxtafoveal telangiectasia (IJT) is described. A 49-year-old woman referred to our clinic with progressive vision loss and best corrected visual acuity (BCVA) was found to be 20/80 OD and 20/25 OS. Corresponding to the area of macular retinal greying in fundus examination, there was a ring-shaped increased reflectance in confocal red free fundus imaging and a ring-shaped increased autofluorescence in fundus autofluorescence (FAF) imaging in both eyes. In addition, FAF imaging revealed a hyperautofluorescent area in the foveola in both eyes, possibly related to the increased retinal transparency due to photoreceptor cell layer loss in the right eye and cystic cavities in the left eye depicted by optical coherence tomography. Two intravitreal injection of bevacizumab were applied to the right eye and BCVA maintained at the level of 20/32 OD together with an overall decrease of 31 μ m in central foveal thickness after 5 months of the second injection.

Key Words: Idiopathic juxtafoveal telangiectasia, fundus autofluorescence, intravitreal bevacizumab.

ÖZ

Tip 2 idiopatik jukstafoveal telenjektazi saptanan bir olgunun fundus görüntüleme bulguları ve tedavisi anlatılmaktadır. İlerleyici görme azalması şikayeti ile kliniğimize başvuran 49 yaşındaki kadın hastada en iyi düzeltilmiş görme keskinliği (EİDGK) sağ gözde 20/80 ve sol gözde 20/25 idi. Her iki gözde fundus muayenesinde makuladaki gri alanlara uyan bölgede konfokal kırmızıdan yoksun fundus görüntülemesinde halka şeklinde artmış reflektans ve fundus otofloresans (FOF) incelemesinde halka şeklinde artmış otofloresans tespit edildi. İlaveten, FOF görüntülemesinde her iki gözde izlenen foveolar hiperotofloresansın sağ gözde optik koherens tomografi (OKT) ile saptanan fotoreseptör tabakasındaki kayıp sonucunda gelişen artmış retina saydamlığına ve sol gözde OKT'de izlenen kistik kavitelere bağlı olduğu düşünüldü. Sağ göze 2 kez intravitreal bevacizumab enjeksiyonu uygulandı ve ikinci enjeksiyondan sonraki 5. ayda santral fovea kalınlığında 31 μ m azalma ile birlikte EİDGK'nin 20/32'ye yükseldiği görüldü.

Anahtar Kelimeler: İdiopatik jukstafoveal telenjektazi, fundus otofloresansı, intravitreal bevacizumab.

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INTRODUCTION

Type 2 idiopathic juxtafoveal telangiectasia (IJT) becomes manifest with visual acuity and central retinal function loss usually beginning in the fifth to seventh decade, and diagnosed by fundus fluorescein angiography (FFA) revealing parafoveal leakage in the late phase from telangiectatic capillaries detected in the early phase.^{1,2} Herein, we report the fundus imaging findings and the management of a case with type 2 IJT.

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Figure 1: (A) Right and (B) Left fundus photograph demonstrating parafoveal retinal greying and right-angle venules.

CASE REPORT

A 49-year-old woman referred to our clinic with progressive vision loss lasting for three years in the right eye. At the initial visit, best corrected visual acuity (BCVA) was found to be 20/80 OD and 20/25 OS. Anterior segment evaluation and intraocular pressure values were within normal ranges in both eyes. Fundus examination revealed an area of macular retinal greying in both eyes (Figure 1A, B). There was a relative foveolar hyperautofluorescence in fundus autofluorescence imaging (FAF) (Figure 2C, D). Fundus fluorescein angiography (FFA) revealed parafoveal telangiectatic capillaries in the early phase (Figure 2E, F) and diffuse leakage in the late phase in both eyes (Figure 2G, H).

Central macular thickness was found to be 165 μm OD and 134 μm OS by optical coherence tomography (OCT, Stratus-OCT, Zeiss) (Figure 3A, B). In the right eye, there was a loss and disruption of the subfoveolar photoreceptor layer and in the left eye there were cyst like structures within internal retinal subfoveolar layers.

Consequently, the patient was offered an intravitreal bevacizumab (IVB) injection to the right eye. After a detailed informed consent was obtained, IVB injection of 1.25 mg was applied. In the 1st month follow-up visit, BCVA increased to 20/32 OD accompanied with a decrease of 30 μm in central foveal thickness. FFA revealed a significant reduction in macular leakage. Since there was a reduced but persistent leakage, a second IVB was applied two months later. Although the angiographic leakage persisted to a degree, BCVA maintained at the level of 20/32 OD together with an overall decrease of 31 μm in central foveal thickness after 5 months of the second IVB injection (Figure 4A, B).

DISCUSSION

Type 2A IJT is characterised by retinal thickening temporal to the fovea, right-angle venules, retinal pigment epithelial hyperplastic plaques, subretinal neovascularization and crystalline deposits.¹ Typical fundus appearance with parafoveal retinal greying was consistent with type 2A IJT in our patient. Intraretinal pigment epithelium clumping, parafoveal atrophy and vascular membranes that are suggested to associate with the worse visual prognosis were absent.³ Ectatic parafoveal capillaries and a parafoveal area of late-phase hyperfluorescence were found by FFA.

OCT results of our case were similar to previous findings.⁴ An apparent lack of correlation between leakage on FFA and retinal thickening on OCT existed.

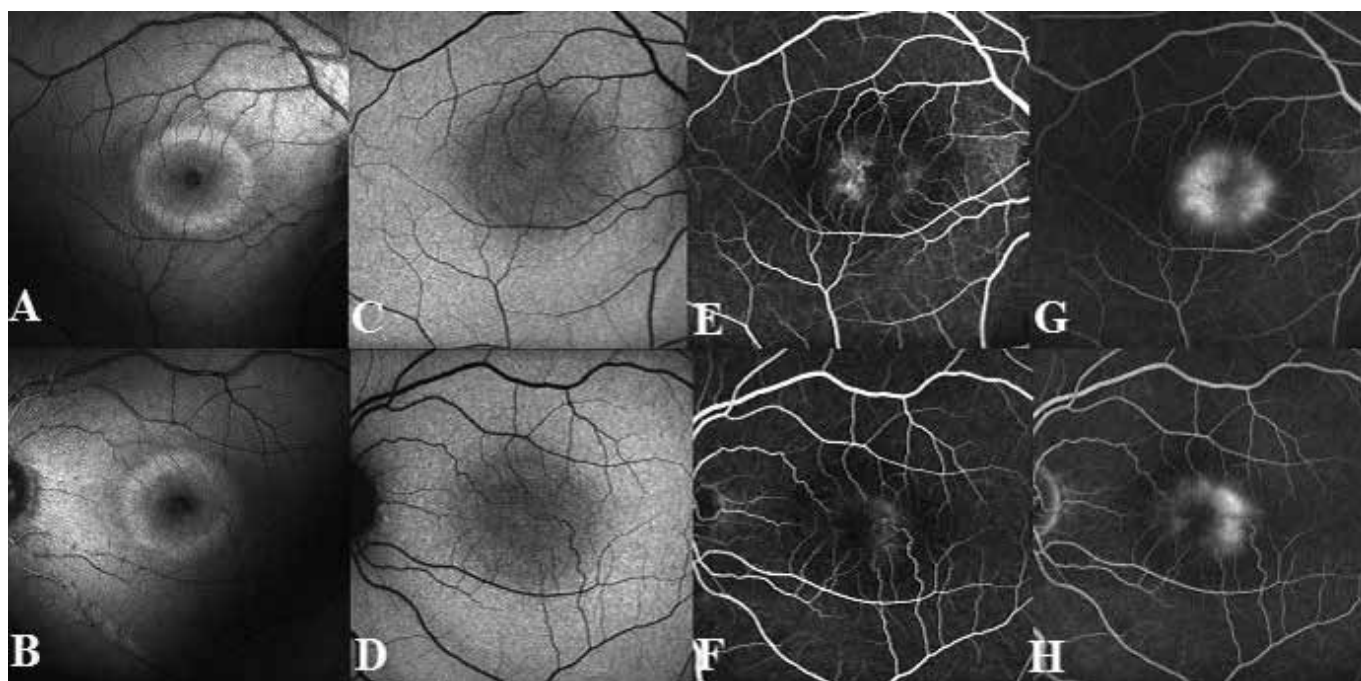


Figure 2: (A) Right and (B) Left confocal red free fundus imaging revealing a ring-shaped increased reflectance corresponding to the area of parafoveal retinal greying. (C) Right and (d) Left fundus autofluorescence imaging showing increased hyperautofluorescence corresponding to the area of parafoveal retinal greying and a hyperautofluorescent area in the foveola. Parafoveal telangiectatic capillaries were observed in the early phase of the fluorescein angiography in the right (E) and left eye (F). Diffuse leakage in the late phase of angiography was more prominent in the right eye (G) than the left eye (H).

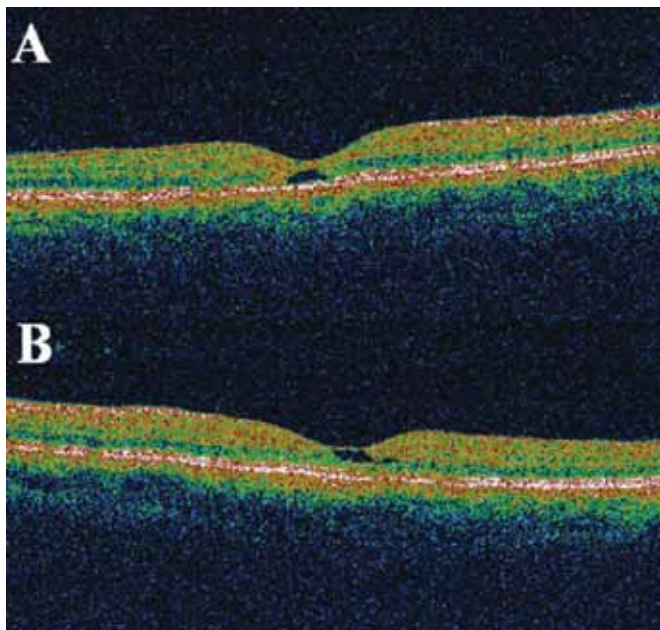


Figure 3: Optical coherence tomography revealed (A) a loss and disruption of the subfoveal photoreceptor layer in the right eye and (B) cyst like structures within internal retinal subfoveal layers in the left eye.

In the right eye, there was a loss and disruption of the subfoveal photoreceptor layer and in the left eye there were cyst like structures within internal retinal subfoveal layers. Inner and outer retinal cystoid spaces were reported to be mainly found in IJT.⁵ Barthelmes and colleagues reported that inner and outer retinal cystic cavities did not result from exudation since there was no accompanying macular thickening and these spaces appeared to have lower optical reflectivity than did cavities occurring secondary to exudation.⁵

Corresponding to the area of retinal greying, there was a ring-shaped increased reflectance in confocal red free fundus imaging (Figure 2A, B) and a ring-shaped increased autofluorescence in FAF imaging (Figure 2C, D). In addition, FAF imaging revealed a hyperautofluorescent area in the foveola in both eyes, possibly related to the increased retinal transparency due to photoreceptor cell layer loss in the right eye and cystic cavities in the left eye depicted by OCT.

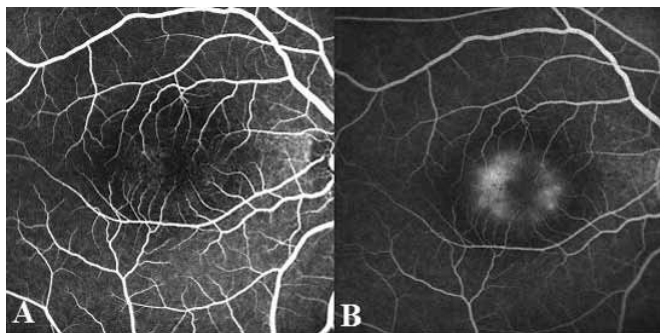


Figure 4: (A) Early and (B) Late phase of fluorescein angiography in the right eye after three months of the second intravitreal bevacizumab injection.

Laser photocoagulation, intravitreal triamcinolone acetonide and anecortave acetate injection treatments were all used in the treatment of IJT. It has been stated that VEGF may participate in the pathogenesis of macular telangiectasia.^{3,6} Although there is no proven effective treatment, recently IVB injection is suggested to decrease angiographic leakage in several preliminary studies.^{2,7,8} Bevacizumab is a recombinant humanized monoclonal antibody that inhibits VEGF and shown to be safe and effective in various ocular diseases with abnormal growth of new vessels or destabilized blood-retinal barrier like neovascular age-related macular degeneration, proliferative diabetic retinopathy, macular edema related to central retinal vein occlusion and diabetic maculopathy. Besides, bevacizumab treatment eliminates ocular side effects like cataract progression and glaucoma seen after intravitreal triamcinolone acetonide injection. Despite the increase in visual acuity, decrease in leakage in FFA and central macular thickness, FAF findings remained unchanged in the present case. Although repeated injections may be required, intravitreal bevacizumab seems to be a promising treatment in type 2A IJT especially in the short term follow-up.

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