

A Case of Coats'-Like Retinitis Pigmentosa

Meri Watanabe, Shinji Makino*, Hironobu Tampo

Department of Ophthalmology, Jichi Medical University, Shimotsuke, Tochigi, Japan

*Corresponding Author:

Name: Shinji Makino

Email: makichan@jichi.ac.jp

Abstract: We present a case of Coats'-like retinitis pigmentosa in a 22-year-old female patient. Both eyes showed the typical funduscopic features of retinitis pigmentosa, and the right eye showed scar formation due to exudative retinal detachment in the temporal peripheral retina with dilatation of the retinal vessels. Fluorescein angiography did not show dye leakage at the scar lesion or the dilated retinal vessels. The patient was generally unaware of the visual deterioration and night blindness. On the basis of this case report, we emphasize the importance of considering the development of Coats'-like exudative change in patients with retinitis pigmentosa.

Keywords: Retinitis pigmentosa, Coats'-like retinitis pigmentosa, Exudative retinal detachment

INTRODUCTION

Retinitis pigmentosa is a retinal degenerative disease characterized by the loss of photoreceptors and formation of retinal pigment deposits [1–3]. Coats' disease is a sporadic form of exudative retinal telangiectasia, and typically presents in one eye. It is common among young male patients in the first decade of life and manifests with intra- and subretinal exudates and dilated tortuous retinal vessels [1–3]. The association between retinitis pigmentosa and exudative retinopathy has been termed a “Coats'-like retinitis pigmentosa” [2] and affects approximately 1–4% of all individuals diagnosed with retinitis pigmentosa [1, 2]. We present a case of Coats'-like retinitis pigmentosa in a 22-year-old female patient.

CASE REPORT

A 22-year-old Japanese female patient was referred to our clinic for blurred vision in right eye. She noticed the decrease in visual acuity at the time of her driver's license renewal. The patients' medical history was unremarkable and there was no family history of ocular disease. Best corrected visual acuities were 0.1 for the right eye and 1.2 for the left eye, and intraocular pressure was 16 mmHg in both eyes. No inflammatory cells were observed in the anterior segment and vitreous in both eyes. Examination of the posterior segment showed greater arteriolar attenuation than expected for

the patient's age and mid-peripheral bone-spicule pigmentary changes on both sides (Fig. 1). The right eye showed scar formation due to exudative retinal detachment in the temporal peripheral retina with dilatation of the retinal vessels (Fig. 1A).

Fluorescein angiography revealed granular hyperfluorescence during the early and late phase corresponding to retinitis pigmentosa in both eyes (Fig. 2). However, there was no dye leakage at the scar lesion or the dilated retinal vessels in the right eye (Fig. 2A).

Visual field testing using Goldmann perimetry showed marked constriction in the right eye (Fig. 3A), while the visual field of the left eye showed mild constriction and absolute and relative scotomata (Fig. 3B).

Optical coherence tomography revealed macular atrophy in the right eye (Fig. 4A) and cystoid macular edema in the left eye (Fig. 4B), and the central foveal thickness was 237 μ m and 360 μ m, respectively.

The standard electroretinogram could not be recorded in both eyes. On the basis of the above findings, we diagnosed Coats'-like retinitis pigmentosa in this patient. The visual findings did not change during the 6-month follow-up period.

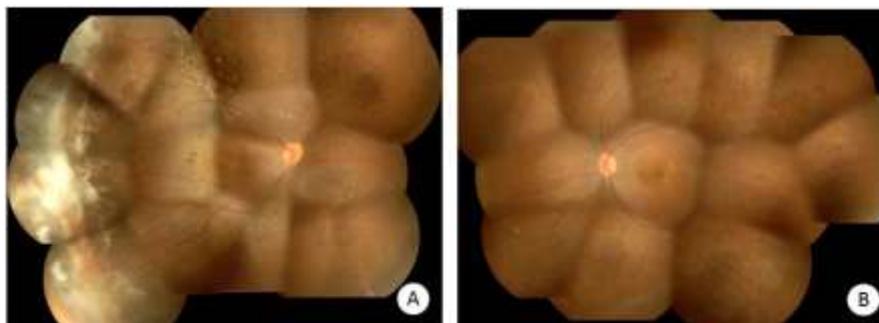


Fig. 1: Photographs of right (A) and left (B) eyes showing typical features of retinitis pigmentosa in both eyes, and scar formation due to exudative retinal detachment in the temporal peripheral retina of the right eye

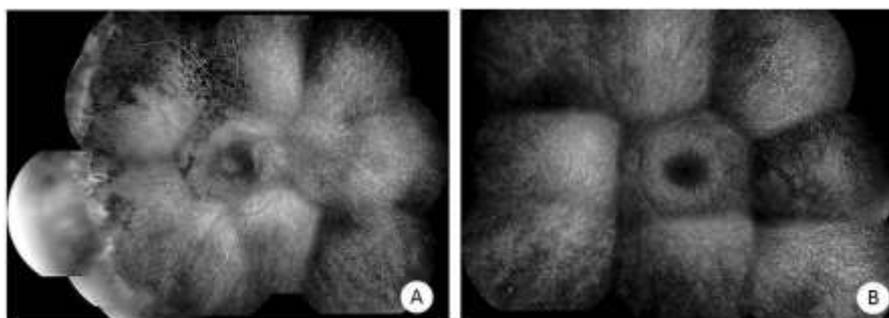


Fig. 2 Fluorescein angiography of the (A) right and (B) left eyes showing granular hyperfluorescence in both eyes, no dye leakage at the scar lesion and the dilated retinal vessels in the right eye.

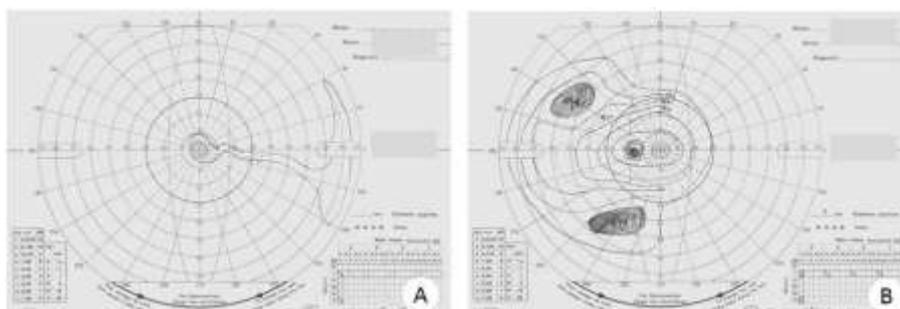


Fig. 3: Goldmann perimetry showing marked constriction in the right eye (A) and mild constriction and absolute and relative scotomata in the left eye (B)

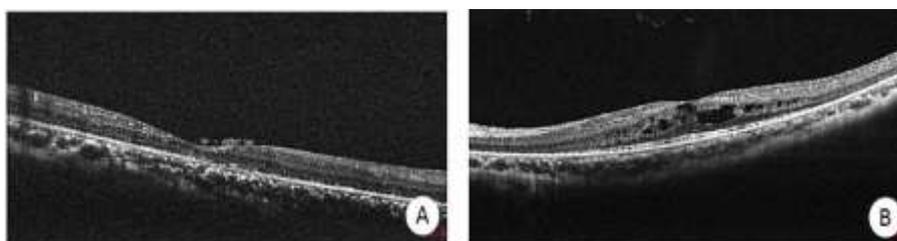


Fig. 4: Optical coherence tomography images showing macular atrophy in the right eye (A) and cystoid macular edema in the left eye (B)

DISCUSSION

Coats'-like retinitis pigmentosa is an atypical form of retinitis pigmentosa. This condition is characterized by vascular abnormalities (aneurysmal dilations and telangiectatic retinal veins), yellow extravascular lipid depositions, and retinal detachment [1, 2, 4-6]. Various studies have suggested that Coats'-

like retinitis pigmentosa differs from the typical Coats' disease with regards to age (occurs at a relatively older age), sex (no predominance), bilateral eye involvement (more often bilateral), progression (more severe prognosis), and retinal location (mainly inferior quadrants and multifocal) [1, 2, 4-6].

The underlying pathogenesis of Coats'-like retinitis pigmentosa is unknown, but several possible explanations have been proposed. One hypothesis is that the condition may occur as a vasodilatory response to the toxic products of photoreceptor/retinal pigment epithelium degeneration [2]. An alternative theory suggests Coats'-type retinopathy may result from chronic microvascular leakage followed by secondary retinal detachment. Retinal hypoxia may occur due to the separation from the choroid, and later, development of telangiectatic abnormalities [1].

Although our patient was largely unaware of the decreased visual acuity and night blindness, we believe this may be attributed to the gradual progression in retinal degeneration from childhood through to adulthood.

CONCLUSION

Finally, we emphasize that it is important to consider the possible development of Coats'-like exudative changes in all patients with retinitis pigmentosa.

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