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Unilateral Retinitis Pigmentosa

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Abstract: To report a case of unilateral retinitis pigmentosa. A 61 years old woman was initially seen at age 51 with a complaint of decreased vision in the right eye. All systemic studies including serologic tests were normal. Best-corrected visual acuity of the patient was 20/200 in her right eye and 20/25 in her left eye. The fundus showed typical pigment deposits of bone corpuscle shapes scattered throughout the fundus in the right eye, the left eye remained completely normal. Fluorescence fundus angiography didn't reveal any abnormalities in the left eye while FFA in the right eye showed vascular filling delayed, diffuse atrophic and bone corpuscle shapes pigment blocking fluorescence of RPE in the posterior pole and peripheral area except macular region. The visual field of the left eye was normal. Examination of color vision showed achromate in the right eye but it was normal in the left eye. Electroretinography showed rod response, cone response and combined response were no recordable in here right eye while ERG was normal in her left eye. The patient was evaluated by ophthalmoscope examination including fundus photography, fluorescence fundus angiography, electroretinography, colour vision examination and visual field examination.

Key words: Unilateral, retinitis pigmentosa, electroretinography, visual field, eye

INTRODUCTION

Unilateral Retinitis Pigmentosa (URP) is a rare degeneration of the photoreceptors. Pedraglia (1865) reported the first case. Francois and Verriest (1952) then proposed the criteria for diagnosis of URP. These are:

- The presence in the affected eye of functional changes and an ophthalmoscope appearance typical of retinitis pigmentosa
- The absence in the other eye of symptoms of an RP with the presence of a normal Electroretinogram (ERG)
- A sufficiently long period of observation (over 5 years) to rule out a delayed onset in the unaffected eye
- Exclusion of an inflammatory cause in the affected eye

The problem of diagnosing URP is not merely academic. Retinitis pigmentosa is an untreatable hereditary condition with poor prognosis (Carr and Siegel, 1973). The future health of the unaffected eye and the genetic implications for the patient's offspring must be emphasized (Kolb and Galloway, 1964).

CASE REPORT

A 61 years old woman complained about loss of visual acuity of her right eye over 20 years, especially in the dark environment accompanying with peripheral vision loss. She was diagnosed as having primary retinitis pigmentosa in her right eye in 2000 at another hospital. The patient didn't have family history of ocular disease. There was no any other history of ocular or general health problems such as ocular traumatization, uveitis, hypertension.

On ophthalmatic examination, her visual acuity was 20/200 OD and 20/25 OS. Anterior segment examination showed posterior pole caract in her right eye while the lens of the fellow eye was clear. The fundus showed typical pigment deposits of bone corpuscle shapes scattered throughout the fundus in the right eye, the left eye remained completely normal. Fluorescence fundus angiography didn't reveal any abnormalities in her left eye (Fig. 1 and 2) while FFA in her right eye showed vascular filling delayed, diffuse atrophic and bone corpuscle shapes pigment blocking fluorescence of RPE in the posterior pole and peripheral area except macular region (Fig. 3). The visual field of the right eye showed only a six degree central island, the visual field of the left eye was normal (Fig. 4).

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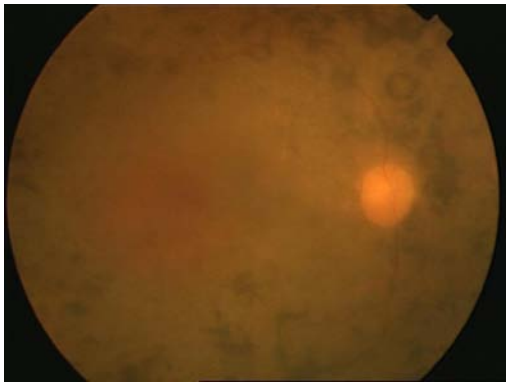

Fig. 1: The fundus in the right eye: typical pigment deposits of bone corpuscle shapes scattered throughout the fundus

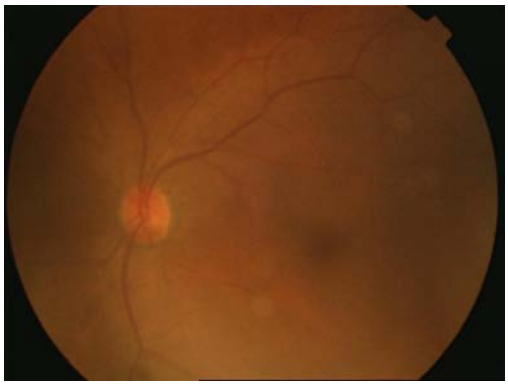

Fig. 2: The fundus in the left eye: completely normal

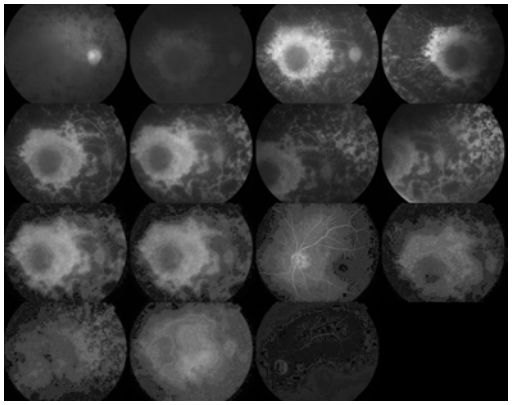

Fig. 3: FFA in both eyes: FFA in her right eye showed vascular filling delayed, diffuse atrophic and bone corpuscle shapes pigment blocking fluorescence of RPE in the posterior pole and peripheral area except macular region. FFA didn't reveal any abnormalities in her left eye

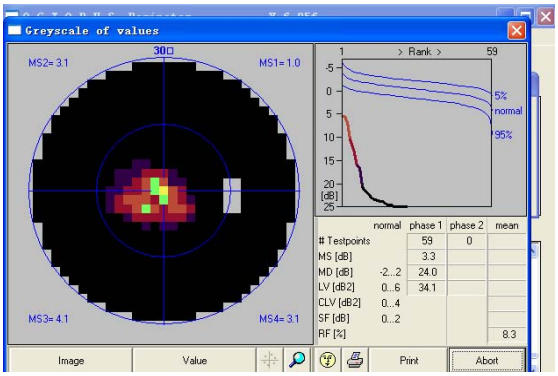

Fig. 4: The visual field of the right eye: only a six degree central island

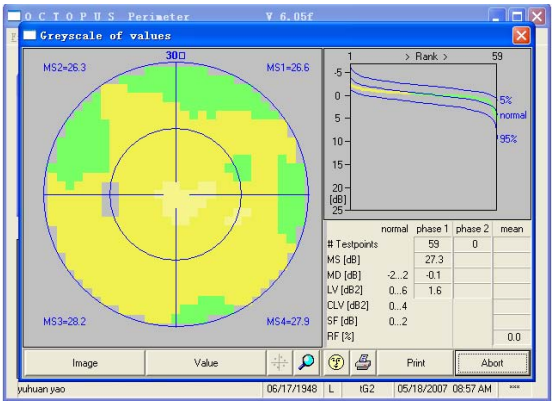

Fig. 5: The visual field of the left eye: normal appearance

Examination of colour vision showed achromat in the right eye but if was normal in the left eye (Fig. 5). Electroretinography showed rod response, cone response and combined responses were no recordable in her right eye while ERG was normal in her left eye (Fig. 6). According to her examination of both eyes, she was diagnosed unilateral retinitis pigmentosa.

As a special type of primary retinitis pigmentosa, URP is a extremely rare disease in clinic. Few reports is available in the whole world (Auerbach and Rowe, 1968; Sverak *et al*., 1968). The first case of URP was reported by Pedraglia (1865). Then, Francois and Verriest proposed the criteria for diagnosis of URP. The patient resented here confirms all the conditions. The functional examinations (ERG, colour vision and microperimetry) and morphological examinations (fundus examination and FFA) revealed obviously abnormal in the right eye while they are normal in the left eye.

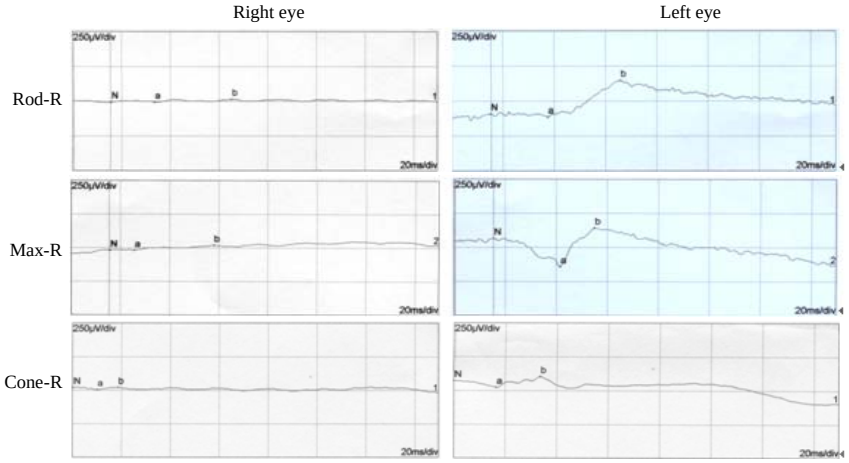

Fig. 6: Electroretinography of both eyes: rod response, cone response and combined response were no recordable in her right eye while ERG was normal in her left eye

Aparting from these features, the patient has other features. Firstly, the affected eye was a typical achromat but the other eye was normal. Secondly, the affected eye accompanied posterior pole cataract while the lens of the fellow eye was clear. Thirdly, no other members in her family involved.

The etiology of URP is unknown. There has been no proof that it is inherited (Chen *et al*., 2006). Francois and Verriest list 14 possible exogenous agents, mostly infectious. Maeder think the most likely causal factor would be some compromise of the vascular supply to the retina (Maeder and Muller, 1950). There was no satisfactory therapy for this disease, gene treatment, artificial retina and transplanting retina are investigating.

CONCLUSION

Unilateral retinitis pigmentosa is a rare condition. Ophthalmological functional and morphological examinations are necessary to confirm the diagnosis.

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