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Case Report

Unilateral Retinitis Pigmentosa: A Rare Case Report

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Abstract

Retinitis Pigmentosa (RP) is a rod-cone dystrophy, the common feature being progressive deterioration in vision due to degeneration of photoreceptors and adjacent cell layers of retina. Night blindness is the initial presenting symptom but in advanced cases there could be gradual deterioration of the light-sensitive cells of the retina causing diminution of vision in daylight. It is a hereditary disorder that can be autosomal dominant, autosomal recessive or X-linked. Presentation is usually bilateral and symmetrical. Unilateral Retinitis Pigmentosa (URP) is an uncommon disease involving RP-like changes in one eye with the fellow eye being completely unaffected. Here we report a case of unilateral retinitis pigmentosa in which visual field and full field ERG was done to support the diagnosis.

Keywords: Full field ERG; Retinitis Pigmentosa (RP); Rod- cone dystrophy; Unilateral Retinitis Pigmentosa (URP)

Introduction

Unilateral Retinitis Pigmentosa (URP) is a rare manifestation involving affected eye with RP-like changes while the fellow eye being completely unaffected [1,2]. The clinical and Electroretinography (ERG) findings of RP should be well defined in affected eye to exclude from other pigmentary retinopathies for proper diagnosis. The criterion for diagnosis of unilateral retinitis pigmentosa was proposed by Francois and Verriest in 1952 which includes: [3].

- The presence in the affected eye of functional changes and an ophthalmoscopic appearance typical of primary pigmentary degeneration.
- The absence in the other eye of symptoms of tapetoretinal dystrophy with a normal Electro Retinogram (ERG).
- A suffciently long period of observation (over 5 years) to rule out delayed onset in the unaffected eye.
- Exclusion of an inflammatory cause in the affected eye.

Case Report

A 33 year old male presented with chief complain of gradual painless diminution of vision in his right eye for last 12 years. There were no any other complaints apart from vision loss. There was no history of trauma and any other systemic problems that could be suggestive of secondary pigmentary retinopathy. He was diagnosed with retinitis pigmentosa in the right eye 6 years ago in a tertiary eye hospital. There was no similar eye problem in his family members.

On examination, his presenting visual acuity was 4/60 in RE and 6/6 in left eye. Visual acuity didn't improve in RE even with pinhole or best refractive correction. On Slit lamp examination, anterior segment of the both eyes was normal. On posterior segment examination, fundus evaluation of the right eye revealed mild attenuation of blood vessels and bony spicules in peripheral and mid-peripheral area whereas fundus examination of the left eye was normal (Figure 1a,1b). Intraocular pressure of the both eyes was within reference limit.

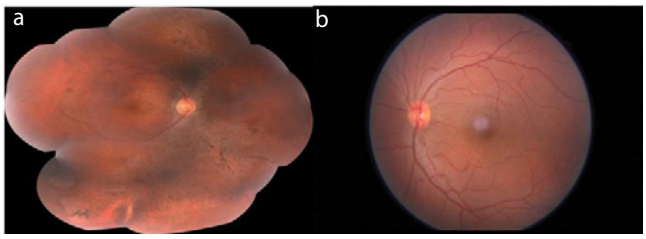

Figure 1: a. Montage view of right fundus shows mild attenuation of arterioles, bony spicules in periphery and mid periphery area. b. Normal fundus of the left eye.

His visual field investigation showed highly decreased retinal sensitivity in the right eye whereas it was within normal limit in the left eye (Figure 2a, 2b).

Full field ERG of the right eye showed decreased Rod and cone response while it was normal in the left eye. There is a reduced amplitude for RE in all test parameters with longer latency time, suggesting functional defects in rod-cone response (Figure 3a,3b). With above findings we made a diagnosis of unilateral retinitis pigmentosa.

Discussion

Retinitis pigmentosa is a hereditary disorder that can be autosomal dominant, autosomal recessive or X-linked. Because of their genetic background retinitis pigmentosa are usually bilateral and symetrical. Unilateral RP is a rare degeneration of photoreceptors which was first described in 1948 [4]. The frequency of URP is reported to be around 5% [5,6]. URP appears more frequently in adult patients. The etiology of unilateral retinitis pigmentosa is unclear and unlike RP there is no evidence to support that unilateral retinitis pigmentosa is an inherited condition [7]. In some studies, the genetic inheritance behind the unilaterality of the disease may be due to different unidentified mutation at the single loci or non-linked mutations in multiple loci [5,8]. Fundus examination, visual field testing, and ERG are necessary to confirm the diagnosis. The diagnosis of unilateral

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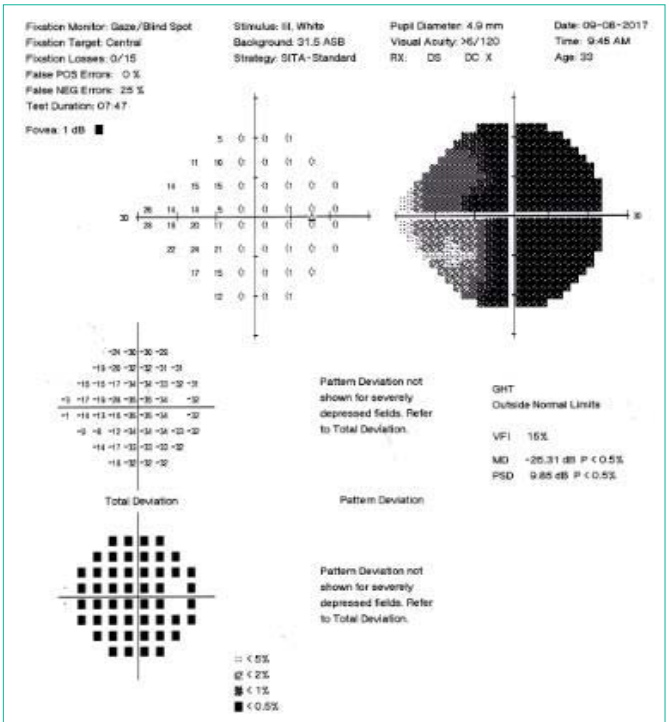

Figure 2a: Automated visual field (24-2) of the right eye shows generalized decreased retinal sensitivity.

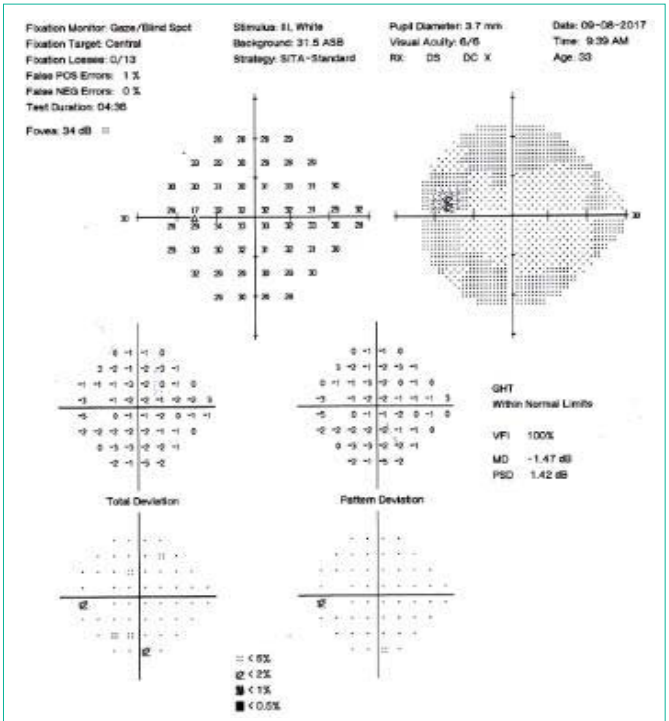

Figure 2b: Automated visual field (24-2) of the left eye was within normal limit.

retinitis pigmentosa is possible only when the François and Verriest criteria are fulfilled. Our case was compatible with Francois and Verriest unilateral retinitis pigmentosa diagnostic criteria: Long disease duration (over five years), presence of functional changes

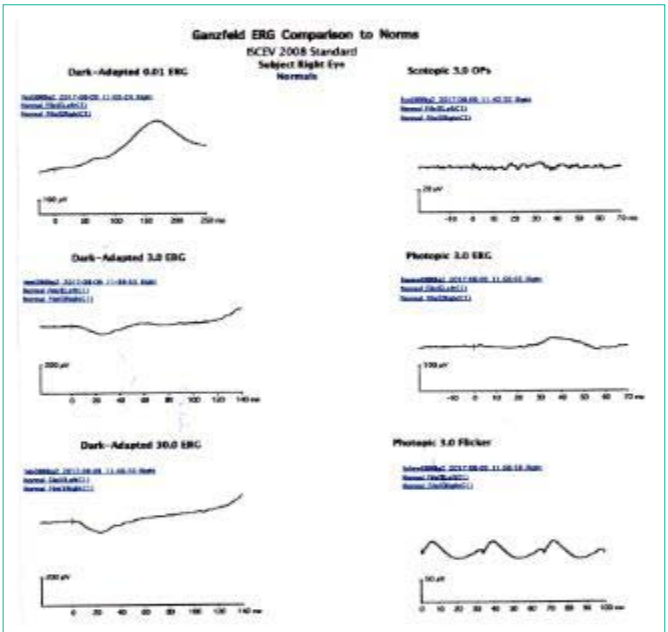

Figure 3a: Full field ERG of the right eye shows reduced amplitude in all test parameters with longer latency time, suggesting functional defects in rod-cone response.

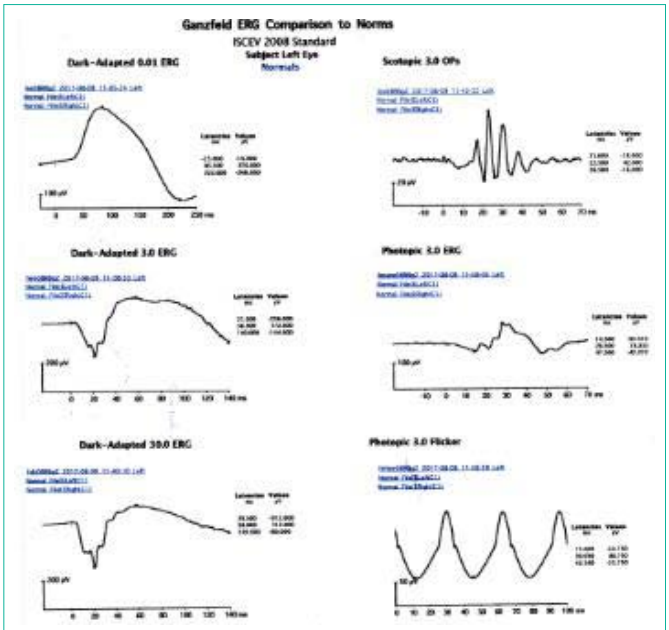

Figure 3b: Full field ERG of the left eye shows normal pattern.

and an ophthalmoscopic appearance of typical primary pigmentary degeneration in affected eye, no clinical and electrophysiological change in unaffected eye, and exclusion of an inflammatory cause [3]. In some literatures, it is shown that the ERG abnormalities are noted even without any fundus finding [9]. Our case is the first one to report unilateral retinitis pigmentosa confirmed by full field ERG and automated visual field testing from Nepal.

Conclusion

Till now there is no exact treatment protocol for retinitis

pigemntosa. Unilateral retinitis pigmentosa is rare form of rod-cone dystrophy. Clinical signs and symptoms, at least 5 year follow up period, ERG and visual field testing are required to support the diagnosis. Regular follow-ups would be recommended to evaluate the progression of the disease condition and to observe any changes in the fellow eye.

References

1. Carr RE, Siegel IM. Unilateral retinitis pigmentosa. Arch Ophthalmol. 1973; 90: 21-26.
2. Spadea L, Magni R, Rinaldi G, Dragani T, Bianco G. Unilateral Retinitis pigmentosa: Clinical and Electrophysiological Report of Four Cases. Ophthalmologica; 1998; 212: 350-354.
3. Francois J, Verriest G. Retinopathie pigmentaire unilaterale. Ophthalmologica. 1952; 124: 65-87.
4. Dreisler KK. Unilateral retinitis pigmentosa: two cases. Acta Ophthalmol. 1948; 26: 385-393.
5. Farell DF. Unilateral retinitis pimentosa and cone-rod dystrophy. Clin Ophthalmol. 2009; 3: 263-270.
6. Kolb H, Galloway NR. Three cases of unilateral pigmentary degeneration. Br J Ophthalmol. 1964; 8: 471-479.
7. Thakur A, Puri L. Unilateral retinitis pigmentosa. Clin Exp Optom. 2010; 93: 102-104.
8. Kajiwara K, Berson EL, Dryja TP. Diagenic retinitis pigmentosa due to mutations at the unlinked peripherin/RDS and ROM 1 loci. Science. 1994; 264: 1604-1608.
9. Berson E. Retinitis pigmentosa: Some observations on the course. Principles and practice of clinical electrophysiology of vision St Louis: Mosby. 1991; 501-509.

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