

## Case report

### Unilateral retinitis pigmentosa

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#### Abstract

**Objective:** To report a rare case of unilateral retinitis pigmentosa and to present the clinical features, and findings of multifocal ERG and visual field of this case. **Case:** A 70-year-old-female diagnosed as Retinitis Pigmentosa in right eye 7 years back, presented with further gradual painless diminution of vision in the very eye and without any similar symptoms in left eye. On examination, the findings (including multifocal ERG and visual field) suggested the features of retinitis pigmentosa in her right eye, while the other eye being unaffected. **Conclusion:** In this rare case, the distinct features of retinitis pigmentosa are seen only in one eye, and this can be further confirmed from multifocal ERG and visual field.

**Keywords:** Unilateral Retinitis Pigmentosa, Multifocal ERG.

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#### Introduction

Retinitis Pigmentosa (RP) is a group of hereditary disorders, and the degeneration of retina causes progressive deterioration in vision, which is its common feature. It mainly affects rod cells where night blindness appears as the first feature, with gradual deterioration of the light-sensitive cells of the retina leading to diminution of vision in daylight in the advanced stages. Its occurrence is mostly bilateral and symmetrical (Weleber, 1989).

Unilateral Retinitis Pigmentosa (URP) is a rare condition where degeneration of the photoreceptors occurs involving retinitis pigmentosa-like changes in one eye, and the other eye is completely unaffected (Spadea et al, 1998).

In 1952, Francois and Verriest proposed four postulates or criteria that a speculative case of URP must satisfy. These are:

1. The presence in the affected eye of functional changes and an ophthalmoscopic appearance typical of primary pigmentary degeneration
2. The absence in the other eye of symptoms of tapetoretinal dystrophy with a normal Electro Retinogram (ERG).
3. A sufficiently long period of observation (over 5 years) to rule out delayed onset in the unaffected eye
4. Exclusion of an inflammatory cause in the affected eye (Francois & Verriest, 1952).

There are a few similar cases reported in the literature (Thakur & Puri, 2010; Chen et al, 2006).

#### Case report

A 70 year old female presented with complaints of gradual painless diminution of vision in

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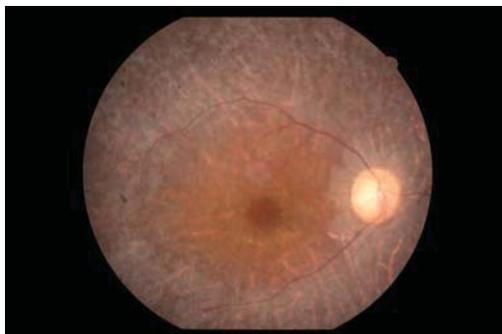
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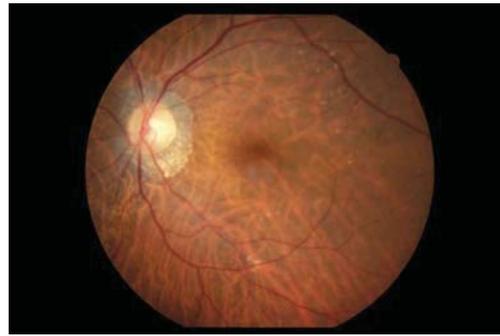
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her right eye for the last 10 years. She gave history of cataract surgery in her both eyes 7 years ago at a tertiary eye hospital of Nepal. She was diagnosed as the case of retinitis pigmentosa in her right eye (RE) 7 years ago by an ophthalmologist. There was no history of trauma to the eye or the head or any systemic illness in the past or present. Similarly, there was no history of associated inflammatory conditions that could be suggestive of secondary pigmentary retinopathy. There was no family history with similar ocular problem. Ocular examination revealed unaided visual acuity of 6/60 in right eye and 6/9 vision in left eye (LE). On slit lamp examination she was pseudophakic in her both eyes. There was no improvement of vision in RE even after refraction (retinoscopy finding -0.50 DC X 90) however vision improved to 6/6 with -0.50 Ds in LE. The intra-ocular pressures were 16 and 17 mmHg in RE and LE respectively. On posterior segment examination, there was pigmentary mottling in the peripheral and mid peripheral retina (spicules), edematous macular region, waxy pallor optic disc with attenuation of arterioles, in RE. Similarly, there was no sign of other diseases such as multifocal chorioretinitis and posterior uveitis that could be suggestive of pigmentary changes in the retina. The fundus picture of LE was within normal limit except few age related changes. These findings were suggestive of RP in RE (Fig 1 and 2.)



**Figure 1:** Fundus Photograph of right eye.



**Figure 2:** Fundus photograph of left eye

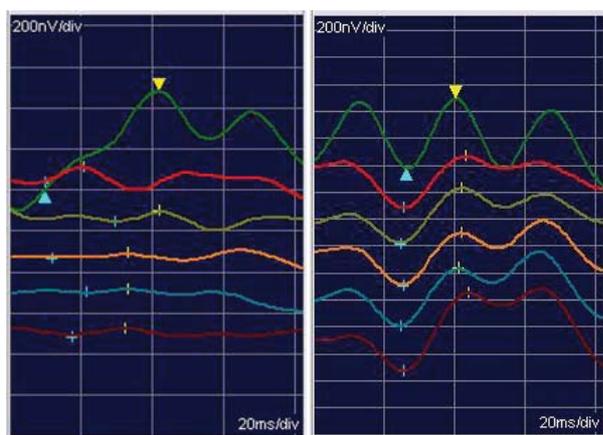
Multifocal ERG showed significant reduction of amplitude of b waves in the peripheral and mid peripheral retina of RE as compared to that of LE. Similarly, the amplitude of b waves in the macular region of RE (60.7nV/deg<sup>2</sup>) was also found to be slightly reduced than that of LE (65.9nV/deg<sup>2</sup>) (Table 1 and 2).

**Table 1:** Multifocal ERG showing reduction of amplitude on the peripheral retina of right eye.

Ring	Amp.P1 [nV/deg <sup>2</sup> ]	Amp.P1 [μV]	Amp.N1 [μV]	Pet.N1 [ms]	Pet.P1 [ms]	Area [deg <sup>2</sup> ]
1	60.7	0.49	0.071	9.8	42.2	8
2	6.64	0.072	0.009	9.8	20.6	10.8
3	3.55	0.053	0.028	29.4	42.2	15
4	1.12	0.023	0.001	11.8	33.3	20.1
5	0.65	0.017	0.006	21.6	33.3	26
6	1.14	0.039	0.034	17.6	32.4	34.3

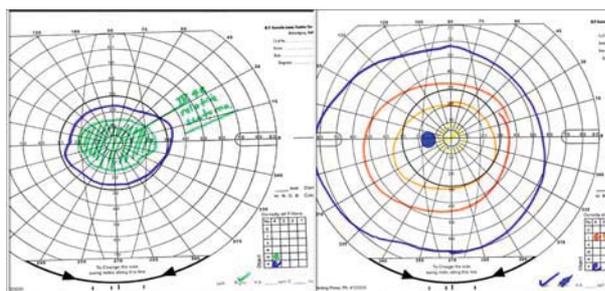
**Table 2:** Multifocal ERG showing normal amplitude on the peripheral retina of Left eye.

Ring	Amp.P1 [nV/deg <sup>2</sup> ]	Amp.P1 [μV]	Amp.N1 [μV]	Pet.N1 [ms]	Pet.P1 [ms]	Area [deg <sup>2</sup> ]
1	65.9	0.53	0.23	26.5	40.2	8
2	35.9	0.39	0.33	25.5	43.1	10.8
3	27.6	0.41	0.18	24.5	42.2	15
4	19.6	0.39	0.27	25.5	42.2	20.1
5	16.6	0.43	0.23	24.5	41.2	26
6	17.2	0.59	0.24	25.5	44.1	34.3



**Figure 3:** Diagrammatic representation of ERG waves of RE and LE respectively

Figure 3 shows combined response of rods and cones according to the retinal area divided in ring from centre to periphery. It is clear to see that there is very minimal response in the peripheral and mid peripheral areas of RE as compared to that of LE.



**Figure 4:** Visual field of RE with peripheral constriction and central relative scotoma of IV4e and that of LE which is normal, respectively.

Goldman perimetry showed constricted visual field with central relative scotoma of IV 4e in RE whereas normal visual field findings in her LE.

### Discussion

Unilateral Retinitis Pigmentosa is a rare degeneration of the photoreceptors which involves RP-like changes in one eye while the other eye is completely unaffected. The frequency of URP is reported to be around 5% of bilateral retinitis pigmentosa (Farell, 2009;

Kolg & Galloway, 1964). The condition is diagnosed only when the François and Verriest criteria are fulfilled, namely, that all infective etiologies should be excluded, the clinical signs of retinitis should be present in the affected eye and the total absence of any signs or symptoms of RP in the fellow eye should be ensured (Spadea et al, 1998; Francois & Verriest, 1952).

Till now the etiology of URP is unknown and its inheritance is unclear. However, as shown by 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 (Farell, 2009; Kajiwara et al, 1994). In URP the functional (ERG, dark adaptometry) and morphological (fundus photography/Ophthalmoscopy and FFA) findings should be normal in unaffected eye whereas in affected eye there is pigmentary mottling in the periphery, attenuated arterioles and presence of cystoid macular edema with decreased or absence of photoreceptor response in ERG. Our case meets all the criteria proposed by Francois and Verriest. Though similar cases have been reported from Nepal, our case is the first one to report ERG verified diagnosis of URP (Thakur, 2010). In this case, if dark adaptometry was performed to assess unilateral night blindness, it would have helped in further confirmation of diagnosis; however, due to unavailability of the instrument the test could not be performed.

### Conclusion

We present a case of unilateral RP without any complaints of night blindness. The condition was detected during full ophthalmological examination when the patient presented with diminution of vision of affected eye. Regular 6 months follow ups would be recommended in such cases to assess the progression of the condition and to see any changes in the fellow eye.

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