Cureus
Part of SPRINGER NATURE

Open Access Case Report

Unilateral Pigmentary Retinopathy in a 43-Year-Old Woman: A Case Report

Mohammad Abbas [1], Najah Mohammad [2]

1. Department of Surgery, Ophthalmology Unit, Ghazi Al-Harriri for Surgical Specialities Hospital, Baghdad, IRQ 2. Department of Surgery, College of Medicine, University of Baghdad, Baghdad, IRQ

Corresponding author: Najah Mohammad, najah@comed.uobaghdad.edu.iq

Review began 09/12/2026
Review ended 11/01/2026
Published 11/13/2026

© **Copyright** 2026
Abbas et al. This is an open access article distributed under the terms of the Creative Commons Attribution License CC-BY 4.0., which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

DOI: 10.7759/cureus.119627

Abstract

Unilateral retinitis pigmentosa represents a rare ophthalmologic diagnosis with poorly understood pathophysiological and genetic characteristics, in contrast to the more common bilateral form of retinitis pigmentosa (RP). In this report, we present a 43-year-old woman with an unremarkable past medical history who presented with a one-year history of progressive visual loss and night blindness. Her fundus and tomography examinations showed features of RP in the right eye and a normal left eye. The diagnosis was supported by the electroretinography findings.

Categories: Ophthalmology
Keywords: electroretinography, iraq, nyctalopia, retinitis pigmentosa, unilateral

Introduction

Retinitis pigmentosa (RP) represents a genetically heterogeneous group of inherited retinal diseases related to the degeneration of rod photoreceptors, followed by the secondary loss of cone photoreceptors [1]. The disease is frequently complicated by secondary ocular complications, including posterior subcapsular cataract and cystoid macular edema [2]. Multiple inheritance patterns have been described: autosomal dominant, autosomal recessive, and X-linked. Sporadic presentations lacking a verifiable family history or identified pathogenic variant are recognized. Unilateral retinitis pigmentosa (URP) is usually described in isolated cases [3].

Clinically, RP typically manifests within the first three decades of life, with nyctalopia serving as the primary symptom. Patients subsequently experience progressive concentric constriction of the peripheral visual field, which may remain silent during the early stages due to neurosensory compensatory mechanisms, ultimately leading to disturbances in central visual acuity. Disease onset, phenotypic expressivity, and rate of progression demonstrate substantial interindividual variability. In URP, these clinical manifestations remain strictly unilateral; however, bilateral RP may begin with an asymmetrical pattern, so long-term follow-up of the unaffected eye is needed before making a diagnosis of URP [3-5].

Many diseases can mimic unilateral or bilateral RP, including infectious conditions (e.g., syphilis), drug-related and iatrogenic conditions (e.g., laser photocoagulation), metabolic and nutritional deficiencies (e.g., vitamin A and zinc deficiencies), and other retinal dystrophies [6].

Ophthalmoscopic evaluation reveals the classic diagnostic triad: waxy pallor of the optic nerve head, prominent retinal arteriolar attenuation, and intraretinal hyperpigmentation, characterized by midperipheral bone-spicule formation alongside retinal pigment epithelium mottling and granularity [7]. These findings may be mimicked by other rare pathologies, including abetalipoproteinemia and ataxia with vitamin E deficiency [3].

Electroretinography (ERG) is considered the gold standard for the diagnosis of RP, monitoring of disease progression, and differentiation from other differential diagnoses [8]. Full-field electroretinography (ffERG) assesses retinal function in response to different light stimuli, whereas dim white single-flash and flickering white-light stimuli elicit rod and cone responses, respectively. When the retina is significantly affected by RP, scotopic responses demonstrate significant reductions in the amplitudes of both a- and b-waves, which represent responses from the photoreceptors and bipolar cells, respectively [9].

URP represents a rare diagnosis that should be made after fulfilling certain criteria, including longitudinal follow-up demonstrating strictly unilateral clinical and ophthalmological features after excluding other diagnoses [10]. In this case report, we describe a young woman presenting with suspected URP.

Case Presentation

The reported case involved a 43-year-old married woman with an unremarkable past medical history. She presented with a complaint of progressive deterioration in the quality of vision in her right eye, associated

with decreased vision in dim light (nyctalopia). Her symptoms had been present for one year. There was no history of trauma, and no family members were affected by a similar condition.

A comprehensive evaluation was performed to exclude acquired causes of unilateral pigmentary retinopathy, including post-traumatic, post-inflammatory, infectious, vascular, toxic, autoimmune, and paraneoplastic retinopathies. This assessment included a detailed ocular and systemic history, medication and trauma history, comprehensive dilated fundus examination, multimodal retinal imaging, and electrophysiological assessment. Additional laboratory or systemic investigations were performed when clinically indicated. On ophthalmological examination, the anterior segment examination revealed clear corneas bilaterally, normal anterior chambers without cells or flare, and phakic lenses in both eyes. Intraocular pressures were within normal limits. There was no anterior chamber reaction or vitreous cellular activity.

As shown in Figures *1*, *2*, *3*, fundus examination of the right eye revealed attenuated retinal vessels and bone-spicule pigmentation, whereas the left fundus was normal. Optical coherence tomography (OCT) demonstrated loss of the ellipsoid zone (EZ) outside the fovea in the right eye, while the left eye showed normal retinal architecture (Figure *1*).

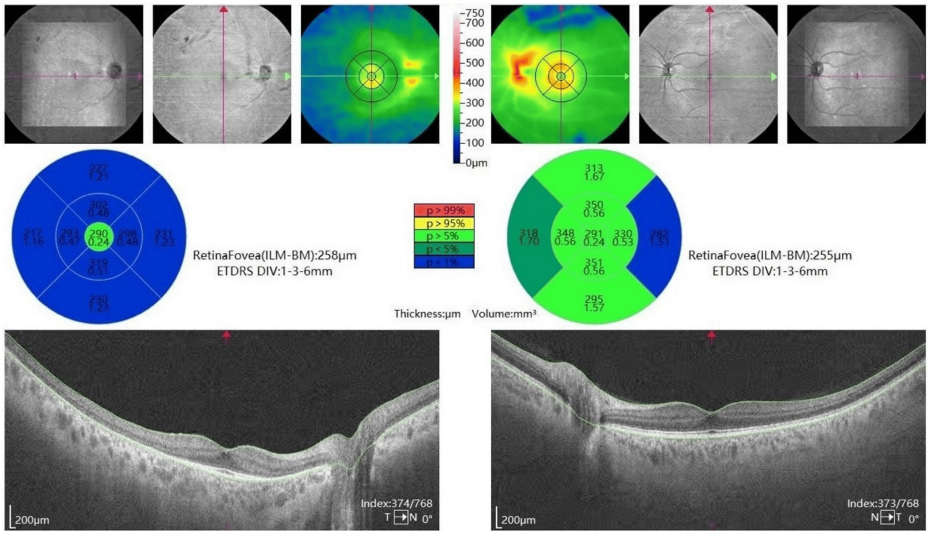


FIGURE 1: OCT of the reported case, showing loss of the EZ outside the fovea in the right eye

OCT: optical coherence tomography; ILM, internal limiting membrane; BM, Bruch's membrane; ETDRS, Early Treatment Diabetic Retinopathy Study; DIV, diameters/divisions; EZ: ellipsoid zone

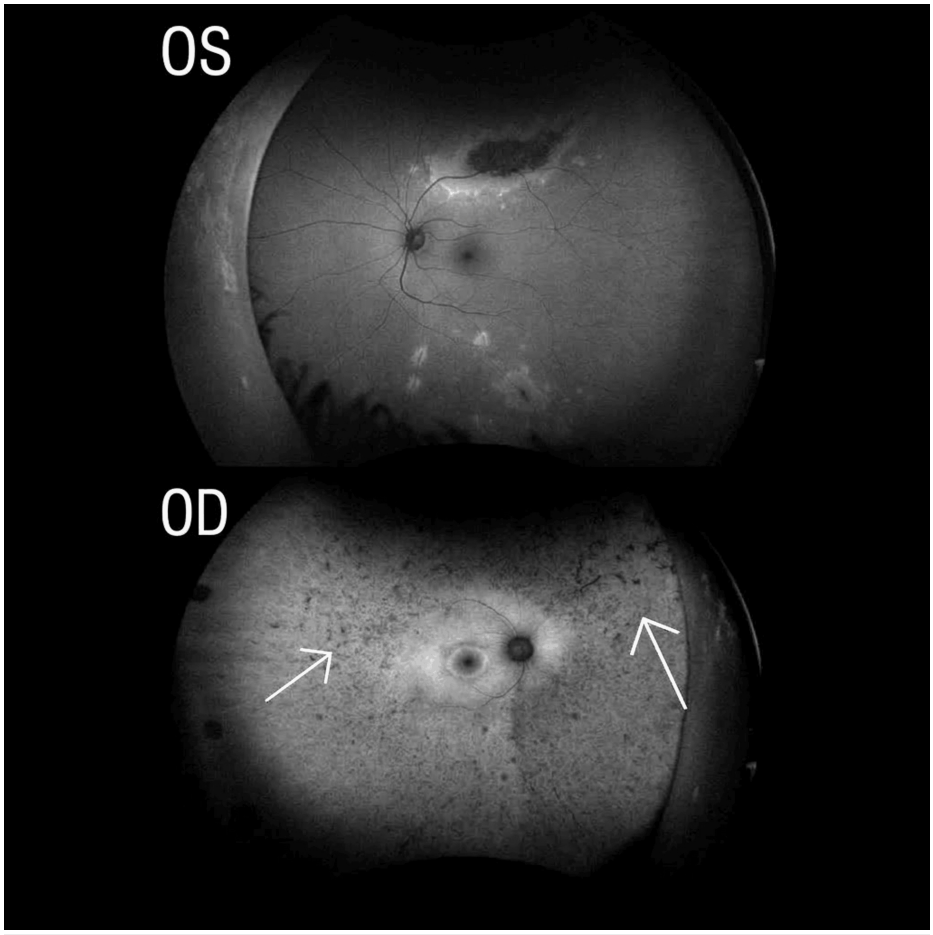


FIGURE 2: Fundus view of both eyes. The right fundus (bottom image) shows attenuated retinal vessels and bone-spicule pigmentation, as indicated by the arrows

OS, oculus sinister; OD, oculus dexter

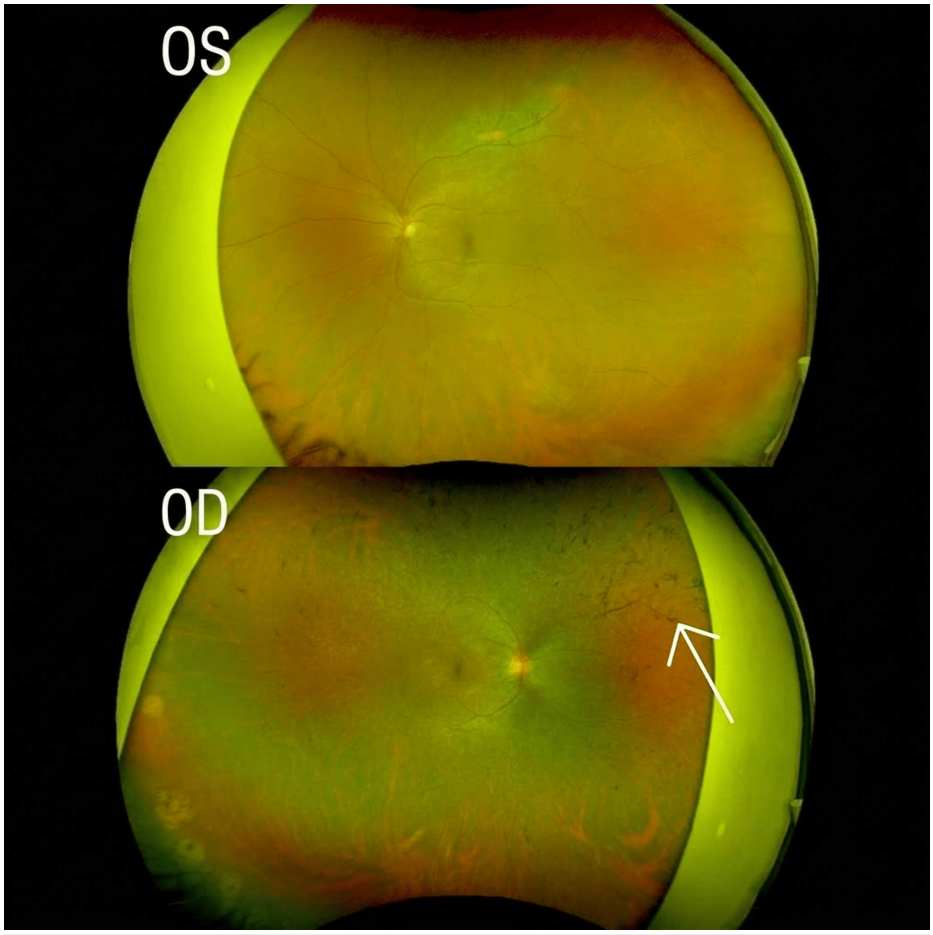


FIGURE 3: Wide-field fundus photograph of the right eye showing the aforementioned changes, as indicated by the arrows

OS, oculus sinister; OD, oculus dexter

ffERG was performed using the Costruzione Strumenti Oftalmici (CSO) RETImax electrophysiology system (CSO, Italy) with an H-K loop corneal electrode. ERG recordings were obtained using a standardized ffERG protocol based on the recommendations of the International Society for Clinical Electrophysiology of Vision (ISCEV). The manufacturer's system provides standardized acquisition and analysis settings for ISCEV ERG protocols.

For the dark-adapted (scotopic) recordings, the patient underwent approximately 20 minutes of dark adaptation. A dark-adapted 0.01 ERG (DA 0.01) was recorded using a dim flash of 0.01 $cd{\cdot}s{\cdot}m^{-2}$ to assess rod system function (normal b-wave amplitude ⩾72 μV), followed by a dark-adapted 3.0 ERG (DA 3.0) using a standard flash of 3.0 $cd{\cdot}s{\cdot}m^{-2}$ to assess the combined rod-cone response (normal b-wave amplitude ⩾127 μV). Dark-adapted 3.0 oscillatory potentials (DA 3.0 OPs) were also recorded to assess activity arising predominantly from the inner and middle retinal layers.

For the light-adapted (photopic) recordings, the eyes were adapted to a background luminance of 30 $cd{\cdot}m^{-2}$ for at least 10 minutes. The light-adapted 3.0 ERG (LA 3.0) was then recorded using a 3.0 $cd{\cdot}s{\cdot}m^{-2}$ flash to assess cone system function (normal b-wave amplitude ⩾69 μV). The light-adapted 30 Hz flicker ERG was subsequently recorded using a 3.0 $cd{\cdot}s{\cdot}m^{-2}$ stimulus at 30 Hz under the same background illumination to assess predominantly cone-mediated post-receptoral function (normal b-wave amplitude ⩾30 μV).

ffERG demonstrated marked interocular asymmetry, with substantially attenuated responses in the right eye under both dark-adapted (scotopic) and light-adapted (photopic) conditions. The dark-adapted b-wave peak-to-trough amplitude was 18.44 μV in the right eye compared with 161.3 μV in the left eye, indicating markedly reduced rod-mediated retinal activity. Similarly, the light-adapted b-wave amplitude was 13.70 μV in the right eye compared with 86.33 μV in the left eye, demonstrating substantial attenuation of cone-mediated retinal responses (Figures *4*, *5* and Table *1*).

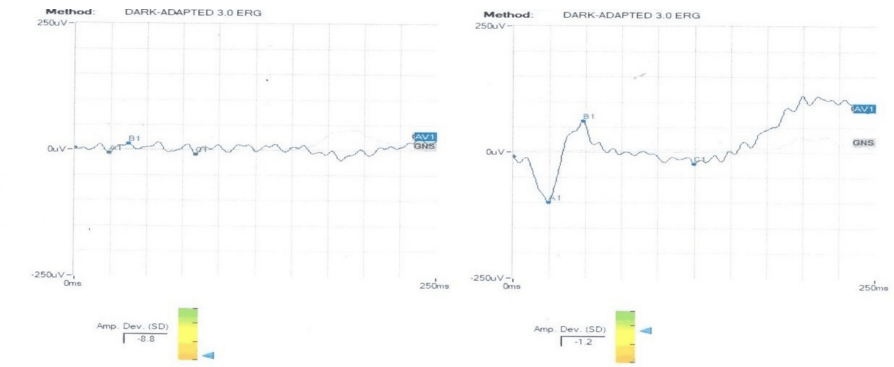


FIGURE 4: ffERG waveforms demonstrating marked attenuation of dark-adapted (scotopic, predominantly rod-mediated) responses in the right eye (right panel) compared with the left eye (left panel)

ffERG, full-field electroretinography; GNS, gain signal; AV1, averaging 1/averaged waveform 1; EGR: electroretinography

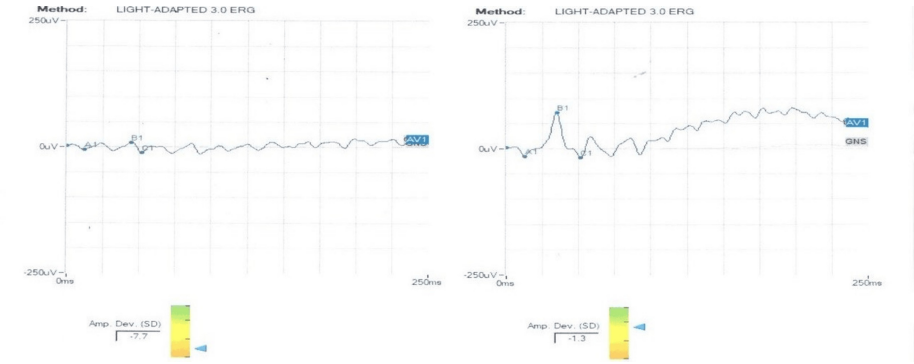


FIGURE 5: ffERG waveforms demonstrating marked attenuation of light-adapted (photopic, cone-mediated) responses in the right eye (right panel) compared with the left eye (left panel)

ffERG, full-field electroretinography; GNS, gain signal; AV1, averaging 1/averaged waveform 1; ERG: electroretinography

Condition/eye	Parameter	a-wave (ISO/A1)	b-wave (B1)	Late component (C1)	a-to-b peak-to-trough (ISO-B1/A1-B1)
Dark-adapted (right eye)	Amplitude (μV)	-6.41	12.02	-9.16	18.44
	Implicit time (ms)	23.44	37.11	83.50	13.67
Dark-adapted (left eye)	Amplitude (μV)	-99.43	61.88	-22.92	161.3
	Implicit time (ms)	23.93	47.85	124.51	23.91
Light-adapted (right eye)	Amplitude (μV)	-5.17	8.53	-11.74	13.70
	Implicit time (ms)	12.21	44.43	51.76	32.23
Light-adapted (left eye)	Amplitude (μV)	-14.49	71.84	-17.26	86.33
	Implicit time (ms)	12.70	34.67	51.27	21.97

TABLE 1: Electroretinographic parameters under dark-adapted (scotopic) and light-adapted (photopic) conditions demonstrated marked reductions in response amplitudes in the right eye

The a-wave represents the hyperpolarization of photoreceptors (rods and cones) in response to light. The b-wave reflects the activity of depolarizing bipolar cells and Müller glial cells in the inner retina. The c-wave is a slow potential generated largely by RPE responses to sustained illumination. Implicit time (peak time) is measured in milliseconds (ms) from the onset of the stimulus to the peak or trough of the waveform. The a-wave to b-wave peak-to-trough amplitude is the composite amplitude measured from the trough of the a-wave to the peak of the b-wave, representing combined outer and inner retinal function.

RPE, retinal pigment epithelium; ISO, isoelectric line

The ERG values corresponding to Figures *4*, *5* are presented in Table *1*. The patient had no history of ocular trauma, infection, or symptoms suggestive of inflammation. Additionally, there was no history suggestive of systemic disease (e.g., syphilis or systemic lupus erythematosus), and the patient was not taking any medications. Table *2* summarizes the basic hematological/biochemical, and toxicological investigations performed. Biochemical screening for syphilis was not performed because of limited resources.

Parameter	Result	Range	Units
Hb	13	Female: 12.1-15.1	g/dL
MCV	88	80-100	fL
WBC count	6.1×10³	(4.5-11.0)×10³	/µL
Platelets	150×10³	(150-450)×10³	/µL
ESR	15	Female: 0-20	mm/hr
Creatinine	0.9	Female: 0.59-1.04	mg/dL
Urea	33	13-43	mg/dL
AST	20	10-40	U/L
ALT	30	7-56	U/L
ALP	64	44-147	U/L
CRP	<0.5	<0.5	mg/dL
Virology screen	HBV (HBsAg), HCV (anti-HCV), HIV-1/2: negative		
Toxicology screen	Standard panel (amphetamines, opiates, cocaine, cannabis, benzodiazepines): negative		

TABLE 2: Basic biochemical investigations of the patient

All values are within the normal reference ranges. Reference ranges are based on institutional laboratory reference for adults.

HBV: hepatitis B; HBsAg: hepatitis B surface antigen; HCV: hepatitis C virus; HIV: human immunodeficiency virus; CRP: C-reactive protein; Hb: hemoglobin; MCV, mean corpuscular volume; WBC: white blood cell; ESR: erythrocyte sedimentation rate; AST: aspartate aminotransferase; ALT: alanine aminotransferase; ALP: alkaline phosphatase

Discussion

While typical RP demonstrates bilateral and highly symmetric involvement, URP represents an exceptionally rare phenotypic variation. Based on the reported literature, URP appears to be more prevalent among adults, consistent with the patient presented in this article [10-12].

The established diagnostic criteria for URP comprise four essential requirements [5]: (1) Unilateral pathognomonic features: characteristic functional deficits and ophthalmoscopic findings typical of rod-cone dystrophy restricted to the affected eye. (2) Normal contralateral eye: absence of clinical manifestations, visual field defects, and electrophysiological abnormalities, confirmed by a normal ffERG. (3) Sufficient longitudinal follow-up: a minimum observation period of five years to exclude delayed-onset or markedly asymmetric bilateral disease. (4) Exclusion of secondary causes: ruling out acquired infectious, inflammatory, vascular, or traumatic etiologies in the affected eye.

The reported patient did not have any family history suggestive of similar symptoms. Nevertheless, scattered case reports in the literature have described sporadic associations with specific gene mutations and positive family histories [3,13,14].

A major limitation of this case was the absence of long-term follow-up, which is essential for establishing a definitive diagnosis of URP. Diagnosing true URP remains challenging, as some suspected cases may represent pseudoretinitis pigmentosa secondary to acquired etiologies, whereas others may reflect markedly asymmetric manifestations of bilateral RP that become apparent only through prolonged observation [10]. Although the clinical findings, multimodal imaging, and exclusion of identifiable secondary causes supported the diagnosis of presumed URP, electrophysiological confirmation of strictly unilateral involvement remained uncertain. ffERG demonstrated marked attenuation of both scotopic and photopic responses in the right eye compared with the fellow eye, indicating pronounced interocular asymmetry. Furthermore, longitudinal assessment was not possible because the patient did not return for subsequent examinations. Therefore, the diagnosis remains presumptive, and the possibility of markedly asymmetric bilateral RP cannot be definitively excluded.

The differential diagnosis in this case included infectious, paraneoplastic, toxic, traumatic, and autoimmune causes. There was no significant medical history, relevant clinical finding, or biochemical abnormality to suggest an alternative systemic etiology, including systemic lupus erythematosus, syphilis, or malignancy.

The patient also denied any history of head or ocular trauma and had no previous ophthalmological evaluations suggestive of a preexisting retinal disorder. Given the absence of supportive clinical or laboratory findings, further extensive investigations, such as comprehensive autoimmune serological testing or systemic imaging, were not pursued. Unfortunately, genetic testing could not be performed because the required test was unavailable, although the patient reported no family history of similar complaints. Table *3* summarizes the published articles reporting unilateral RP [7,10-12,14-17].

Author	Age of the case	Gender	Eye affected	Presentation duration	Family history	Follow-up duration
Weller JM, et al. [7]	43 years	Female	Not reported	Not reported	Not mentioned	30 years
Stamate AC, et al. [10]	30 years	Male	Left eye	10 years	No	1 year
Gupta S, et al. [11]	33 years	Male	Right eye	12 years	No	6 years
Aragao REM, et al. [12]	60 years	Female	Right eye	Not reported	No	Not reported
Mukhopadhyay R, et al. [14]	63 years	Female	Right eye	Incidental	Yes	Not reported
Milibari D, et al.-patient 1 [15]	32 years	Female	Left eye	4 years	No	5 years
Milibari D, et al.-patient 2 [15]	65 years	Male	Left eye	20 years	No	5 years
Thakur A, et al. [16]	24 years	Female	Left eye	Not reported	No	Reported 6 years after diagnosis
Farrell DF [17]	31.3 (mean, nonfamilial) and 38.6 (familial) years	8 Females, 6 Males	Unilateral; individual eye not specified	Not reported	Yes in 5/14 cases	Not reported

TABLE 3: Summary and comparison of published articles reporting unilateral RP

Source: [7,10-12,14-17]

RP: retinitis pigmentosa

Conclusions

Here, we describe a case of presumed URP in a young female who presented with clinical and ophthalmologic features over a one-year period. The absence of similar findings in the contralateral eye and of clinical features suggestive of alternative diagnoses supported the diagnosis of URP. This rare presentation highlights the importance of comprehensive ophthalmologic evaluation and appropriate electrophysiological testing in establishing the diagnosis.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Acquisition, analysis, or interpretation of data: Najah Mohammad, Mohammad Abbas

Drafting of the manuscript: Najah Mohammad, Mohammad Abbas

Critical review of the manuscript for important intellectual content: Najah Mohammad, Mohammad Abbas

Concept and design: Mohammad Abbas

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

References

1. Verbakel SK, van Huet RA, Boon CJ, et al.: Non-syndromic retinitis pigmentosa. Prog Retin Eye Res. 2018, 66:157-86. 10.1016/j.preteyeres.2018.03.005
2. Antonio-Aguirre B, Swenor B, Canner JK, Singh MS: Risk of cystoid macular edema following cataract surgery in retinitis pigmentosa: An analysis of United States claims from 2010 to 2018. Ophthalmol Retina. 2022, 6:906-13. 10.1016/j.oret.2022.04.018
3. Nguyen XT, Moekotte L, Plomp AS, Bergen AA, van Genderen MM, Boon CJ: Retinitis pigmentosa: current clinical management and emerging therapies. Int J Mol Sci. 2023, 24:7481. 10.3390/ijms24087481
4. Dias MF, Joo K, Kemp JA, Fialho SL, da Silva Cunha A Jr, Woo SJ, Kwon YJ: Molecular genetics and emerging therapies for retinitis pigmentosa: basic research and clinical perspectives. Prog Retin Eye Res. 2018, 63:107-31. 10.1016/j.preteyeres.2017.10.004
5. Francois J, Verriest G: Retinitis pigmentosa unilateral [French]. Ophthalmologica. 1952, 124:65-88. 10.1159/000301252
6. Hamel C: Retinitis pigmentosa. Orphanet J Rare Dis. 2006, 1:40. 10.1186/1750-1172-1-40
7. Weller JM, Michelson G, Juenemann AG: Unilateral retinitis pigmentosa: 30 years follow-up . BMJ Case Rep. 2014, 2014:10.1136/bcr-2013-202236
8. McCulloch DL, Marmor MF, Brigell MG, Hamilton R, Holder GE, Tzekov R, Bach M: ISCEV Standard for full-field clinical electroretinography (2015 update). Doc Ophthalmol. 2015, 130:1-12. 10.1007/s10633-014-9473-7
9. Marmor MF: The electroretinogram in retinitis pigmentosa . Arch Ophthalmol. 1979, 97:1300-4. 10.1001/archopht.1979.01020020042009
10. Stamate AC, Burcea M, Zemba M: Unilateral pigmentary retinopathy-a review of literature and case presentation. Rom J Ophthalmol. 2016, 60:47-52.
11. Gupta S, Kumar V: Unilateral retinitis pigmentosa case report. Int J Ophthalmol Vis Sci. 2021, 41:2.
12. Aragao REM, Barreira IMA, Bastos ASV, Carneiro GJAM, Oria TPC: Unilateral retinitis pigmentosa: case report. Rev Med UFC. 2015, 55:54-8. 10.20513/2447-6595.2015v55n2p54-58
13. Marsiglia M, Duncker T, Peiretti E, Brodie SE, Tsang SH: Unilateral retinitis pigmentosa: a proposal of genetic pathogenic mechanisms. Eur J Ophthalmol. 2012, 22:654-60. 10.5301/ejo.5000086
14. Mukhopadhyay R, Holder GE, Moore AT, Webster AR: Unilateral retinitis pigmentosa occurring in an individual with a germline mutation in the RP1 gene. Arch Ophthalmol. 2011, 129:954-6. 10.1001/archophthalmol.2011.171
15. Milibari D, Magliyah M, Semidey VA, Schatz P, ALBalawi HB: Unilateral retinitis pigmentosa associated with possible ciliopathy and a novel mutation. Clin Pract. 2022, 12:491-500. 10.3390/clinpract12040053
16. Thakur A, Puri L: Unilateral retinitis pigmentosa. Clin Exp Optom. 2010, 93:102-4. 10.1111/j.1444-0938.2009.00435.x
17. Farrell DF: Unilateral retinitis pigmentosa and cone-rod dystrophy. Clin Ophthalmol. 2009, 3:263-70. 10.2147/opth.s5130