

**RETINITIS PIGMENTOSA: A Case Report****Delfi¹, Vanda Virgayanti¹ dan Rika Ramadhani Sinambela¹**¹Department of Ophthalmology, Faculty of Medicine, University of Sumatera Utara, IndonesiaCorresponding Author : Delfi¹Email : ¹andreassusanto0805@gmail.com, ²ernani_hadiyati@unigamalang.ac.id
dan ³koharadi2017@gmail.com**Article Info :**Received : February 3rd 2022Approved : February 9th 2022Published : February 15th 2022**ABSTRACT****Keywords:**Retinitis
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Background : Retinitis pigmentosa is an inherited heterogeneous group of retinal disorders represented by rod photoreceptors progressive dysfunction with subsequent cone photoreceptors degeneration and the retinal pigment epithelium. The typical manifestations are progressive loss of visual field and night blindness. **Purpose :** The purpose of this study was to examine with the Amsler Grid on the right and left eyes, no scotomas and metamorphopsia were found. **Method :** The type of research is quantitative with an explanatory research approach, the sampling technique is saturated sampling with the number of subjects 76 employees and the data analysis technique uses PLS. **Results :** Best corrected visual acuity was 6/9 in the right eye and 6/18 in the left eye. There was arteriolar attenuation, waxy disc pallor, bone-spicule pigmentation and pigment deposits on both eyes in funduscopy examination. The patient was diagnosed with retinitis pigmentosa. The management given was vitamin A 15.000 IU/day, DHA 1.200mg/day, and counselling about retinitis pigmentosa, progressivity, and prognosis. **Conclusion :** Retinitis pigmentosa caused irreversible visual and visual field impairment. Low vision management for retinitis pigmentosa was to optimize the visual function and optimized quality of life. Genetic counselling might provide the patient and her family information of the inheritance and genetic disorders implications that could help the patient to obtain medical information regarding the disease.

INTRODUCTION

Retinitis Pigmentosa is a group of photoreceptors and Retinal Pigment Epithelium (RPE) abnormalities, which has genetic, progressive dysfunction, loss of cells, and can occur atrophy of retinal tissue (Lewandowski et al., 2021). The prevalence of this disease ranges from 1:3000 to 1:500000. This disorder is more common in males than females. Based on genetic disorders are divided into x-linked autosomal dominant and autosomal recessive (Nozu et al., 2019). Retinitis pigmentosa is a genetic disease, but in 15-63% of cases, there was no history of retinitis pigmentosa in other family members (Cideciyan et al., 2020).

Typical clinical signs are visual field loss, nyctalopia, and photopsia (Fekri, Soheilian, & Rahimi-Ardabili, 2020). Typical features found on funduscopy are bone-spicule pigmentation on the retina, arteriolar attenuation, and waxy disc pallor. Early in

the disease, this feature is absent in some cases of retinitis pigmentosa (Ilhan & Citirik, 2019). Retinitis pigmentosa without a characteristic fundal abnormality is frequently diagnosed by retinitis pigmentosa sine pigmento or paucipigmentary, which is characterized by lack of pigment accumulation or hypopigmentation on funduscopy. The purpose of this study was to examine with the Amsler Grid on the right and left eyes, no scotomas and metamorphopsia were found.

RESEARCH METHODS

The type of research is quantitative with an explanatory research approach, the sampling technique is saturated sampling with the number of subjects 76 employees and the data analysis technique uses PLS.

RESULTS AND DISCUSSION

A woman (45 years old) came to Universitas Sumatera Utara General Hospital on April 7 2021 to the vitreoretinal division of the ophthalmology clinic. She complained about the progressive blurring of vision in both eyes which started five years before (Fernández-Domínguez, Ameijide-Sanluis, García-Cabo, García-Rodríguez, & Mateos, 2020). It was also accompanied by a narrowing field of vision which was described as looking through binoculars (Deemer et al., 2018). Vision at night or dark is supposed to be getting worse. The patient admitted that she had never experienced visual disturbances and had never worn glasses. She denied any eye pain, hearing loss, and color vision impairment. She denied a history of using psychotic drugs and chloroquine. She had no underlying disease, such as diabetes mellitus and hypertension (Kazi, Akhter, Periasamy, Faruki, & Tahir, 2021).

The patient is a housewife who can still perform daily activities independently, and she is the only child who has three sons aged 10 years, 16 years, and 20 years. According to the patient's confession, no one in the family has the same disorder (ABONG'O, 2021). The patient's parents never had their eyes examined. From the results of the examination carried out on the three patient's children, it was found that the results of the examination were within normal limits, and no one had similar complaints.

On physical examination, vital signs were within normal limits. Refractive examination for far vision correction showed that the visual acuity of the right eye was 6/9, and the visual acuity of the left eye was 6/18 (Fazeenah, 2021). Pinholes in both eyes did not progress. Colour examination with Ishihara on the right and left eyes obtained 14/14 results. Examination with Amsler Grid on the right and left eyes did not reveal any scotoma and metamorphopsia.

Eye movement examination of the patient is full orthotropia, with good eye movement in all directions (Miao, Jeon, Park, Park, & Heo, 2020). On examination of the anterior segment using a slit lamp, the results were within normal limits. Fundus examination using indirect funduscopy showed similar impressions on the right and left eyes with clear media images, well-defined round papillae, physiological a/v ratio, physiological c/d ratio, flat retina, fundus reflex (+), and found hyperpigmented spots in the form of bone spicules with the impression of retinitis pigmentosa. The patient had undergone Humphrey 24-2 examination, and the results were decreased visual field. Patients are advised to do an Electroretinogram (ERG) (Hu, Ma, & Peng, 2021).

The patient was diagnosed with retinitis pigmentosa. The management of this patient is the administration of vitamin A 15.000IU/day, DHA 1.200mgU/day, education about the disease and its prognosis, and adjustment of activity.



Figure 1. Fundus examination of the right eye



Figure 2. Fundus examination of the left eye

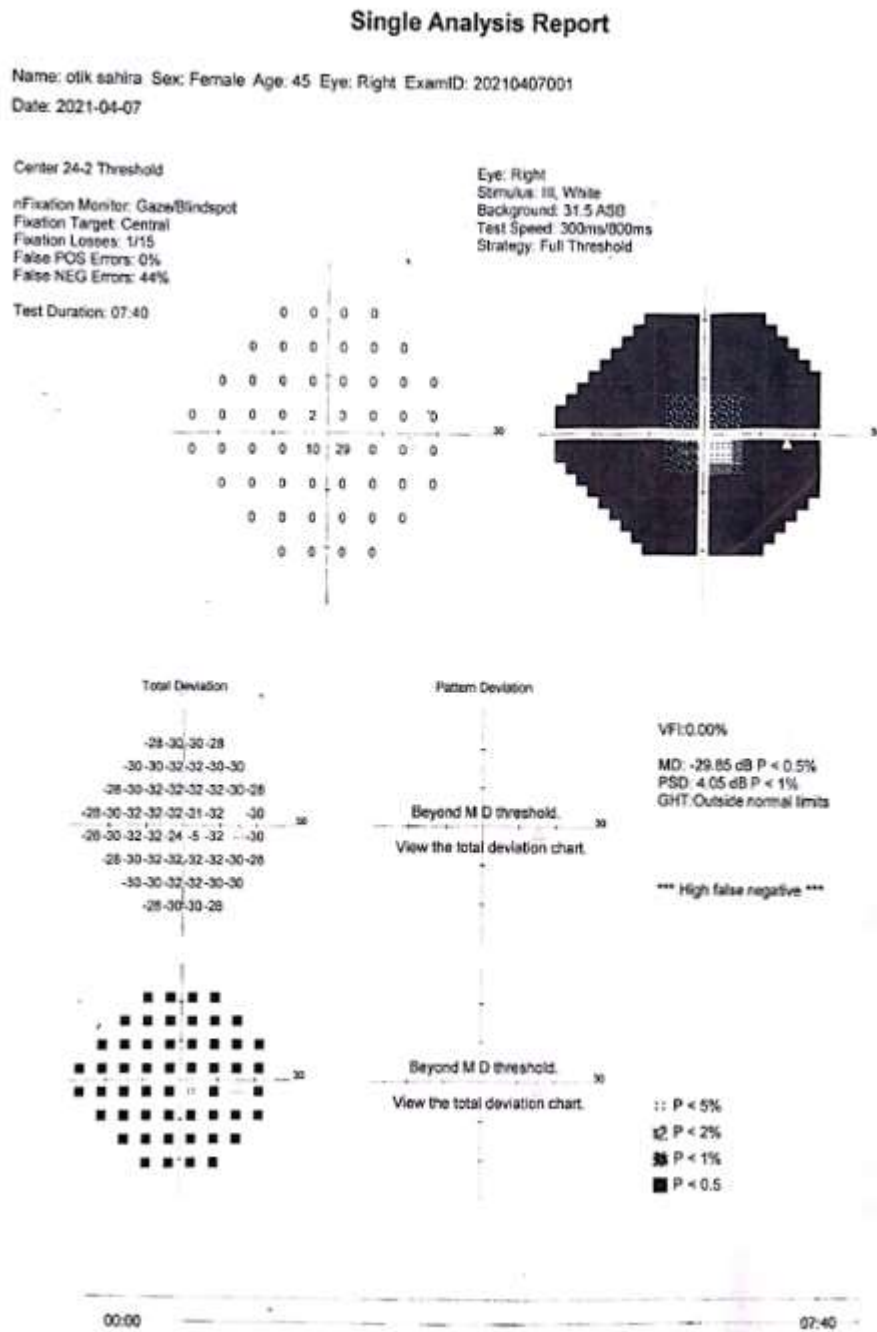


Figure 3. Humphrey Field Test Results OD

Single Analysis Report

Name: otk sahira Sex: Female Age: 45 Eye: Left ExamID: 20210407001
 Date: 2021-04-07

Center 24-2 Threshold

Fixation Monitor: Gaze/Blindspot
 Fixation Target: Central
 Fixation Losses: 1/111
 False POS Errors: 0%
 False NEG Errors: Not Applicable

Test Duration: 09:11

Eye: Left
 Stimulus: III, White
 Background: 31.5 ASB
 Test Speed: 300ms/900ms
 Strategy: Full Threshold

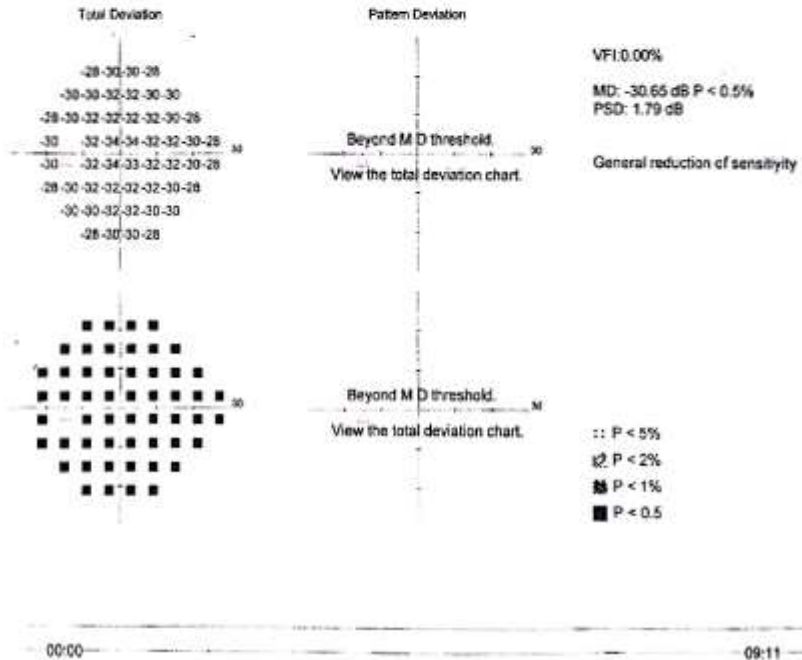
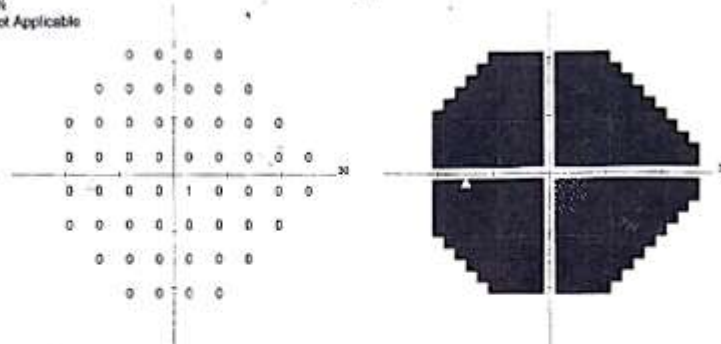


Figure 4. Humphrey Field Test Results OS

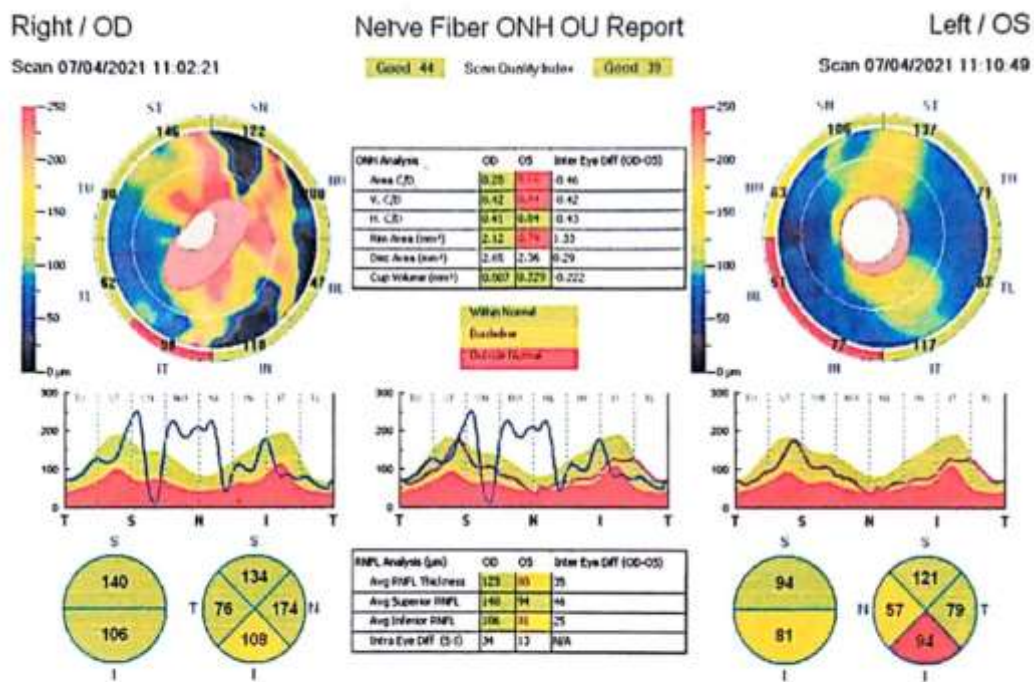


Figure 5. Spectral Domain-OCT results for the right and left eyes

Retinitis pigmentosa is a progressive and genetic retinal dystrophy. The initial symptoms of retinitis pigmentosa in the form of night vision disturbances and peripheral visual field defects occur slowly. The onset of retinitis pigmentosa ranges from infancy to age 50 and is more common in men than women (Birtel et al., 2018). In this case, the patient is a 45-year-old woman. The symptoms in this patient are the typical retinitis pigmentosa, namely bilateral, nyctalopia, and narrowing of the visual field, which have been experienced slowly since ± 5 years ago. On Humphrey 24-2 perimetry examination showed peripheral visual field disturbances and retinal thinning during SD-OCT examination to support the diagnosis of retinitis pigmentosa.

Retinitis pigmentosa usually only affects the eyes or is called primary retinitis pigmentosa, but 20-30% of cases have non-ocular manifestations called syndromic retinitis pigmentosa. The most common syndrome is Usher syndrome, which manifests as hearing loss with or without vestibular dysfunction and visual impairment (Stiff et al., 2020). This patient had primary retinitis pigmentosa because it only affected the eye without other systemic disorders. The patient has never taken psychotic drugs and chloroquine; this excludes the differential diagnosis of drug toxicity which has symptoms resembling retinitis pigmentosa in its early stages. Early poisoning symptoms with psychotic drugs such as phenothiazines are nyctalopia and blurred vision, especially in the central part. Chloroquine poisoning can cause decreased visual field symptoms.

Fundus examination in retinitis pigmentosa can reveal specific signs, including vascular attenuation, bone-spicule pigmentation on the retina, and waxy disc pallor. On fundoscopic examination of this patient, only bone spicules were found in both eyes.

The Amsler Grid examination did not reveal any scotoma or metamorphosis. The Ishihara examination in this patient was within normal limits. In retinitis pigmentosa, color vision was normal until the visual acuity worsen ($\leq 20/40$). Colour vision might worsen in early cases if the cone cells core present in the macula are abnormal.

Management of these patients aims to maximize remaining vision and optimize quality of life. The motivation was also essential in encouraging the patient so that they may adapt to their new situation. The patient received medical therapy in vitamin A 15.000 IU/day and DHA 1.200mg/day. Nutritional supplements have been advocated as therapy for retinitis pigmentosa. Another study reported that vitamin A palmitate (15.000 IU/day) high doses slowed the decline in ERG and visual field responses in eyes with retinitis pigmentosa by about 20% per year. A slight advantage of omega-3 and omega-6 fatty acid supplementation has also been reported (Djuricic & Calder, 2021).

The prognosis for this patient is because there was no systemic disease that could threaten this patient's life. To retinitis pigmentosa is an irreversible disease with a tendency to worsen and the visual field in this patient has narrowed.

CONCLUSION

Retinitis pigmentosa is a disease that causes an irreversible and progressive decrease in visual acuity and visual field. Treatment of retinitis pigmentosa aims to maximize the remaining vision and improve and optimize the patient's quality of life. Genetic counselling can provide patients and their families with information about the inheritance and implications of the genetic disorder of retinopathy pigmentosa and can help them to obtain medical information regarding this disease. The management of this patient is the administration of vitamin A 15.000IU/day, DHA 1.200mg/day, and counseling regarding adjustment of daily activities.

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