A Journey towards Improved Quality of Life of a Typist with Retinitis Pigmentosa

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Abstract

Retinitis pigmentosa (RP) is a group of inherited rod-cone degenerative pathologies that present clinically with similar signs and symptoms. Common fundus findings include bone-spicule pigment formation, attenuated blood vessels in the posterior pole, and waxy optic nerve pallor. Symptoms mostly start with progressive night blindness, mid-peripheral visual field defects, and eventual tunnel vision. A 42-year-old male patient, typist by profession presented to our clinic with complaints of decreased vision and reduced dark adaptation greatly affecting his quality of life. On examination, he was diagnosed with an advanced case of retinitis pigmentosa bilaterally and there was a bilateral posterior subcapsular cataract. His quality of life was assessed by using a simplified version of the VFQ-25 questionnaire. After successful cataract surgery and low vision rehabilitation, his quality of life was reassessed by using the same questionnaire. He had a much more positive outlook on his eyesight. Our case report reviews the life-changing personalized low vision rehabilitation of a typist with retinitis pigmentosa.

Keywords: retinitis pigmentosa, quality of life, low vision, rehabilitation
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Introduction

Retinitis Pigmentosa (RP) is a group of genetic disorders affecting the retina of the eye. It is characterized by symptoms of night blindness, decreased visual acuity, and visual fields eventually leading to legal or complete blindness. Due to the progressive nature of the condition, RP not only has physical manifestations, but it inevitably adversely affects the quality of life as well as mental and emotional well-being[1]. Various studies have demonstrated how it is strongly associated with anxiety and depression[2]. Some suggested a relationship between RP and lower education and employment rates, which consequently impact the mental health of the patients[3].Currently there is no approved therapy to halt the progression, nor to restore vision, so management revolves around slowing down degeneration (using vitamin A supplementation), managing complications, and rehabilitation to tackle its impact on daily life[4]. Various coping strategies have been investigated to help improve quality of life, including dealing with practical and emotional aspects[5]. As demonstrated by Kay et al, the use of a white cane and low vision assistive devices have a momentous impact on not only emotional welfare but also financial, social, and workplace or academic productivity[6].

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Case Report

A 42-year-old male patient was diagnosed with retinitis pigmentosa (RP) incidentally at the age of 15, when he went to visit an ophthalmologist at a tertiary care hospital for a style. He was prescribed spectacles at the time and little information was provided about the prognosis of the condition and its management. He later visited a local clinic at the end of 2020, where he was informed of the ble-

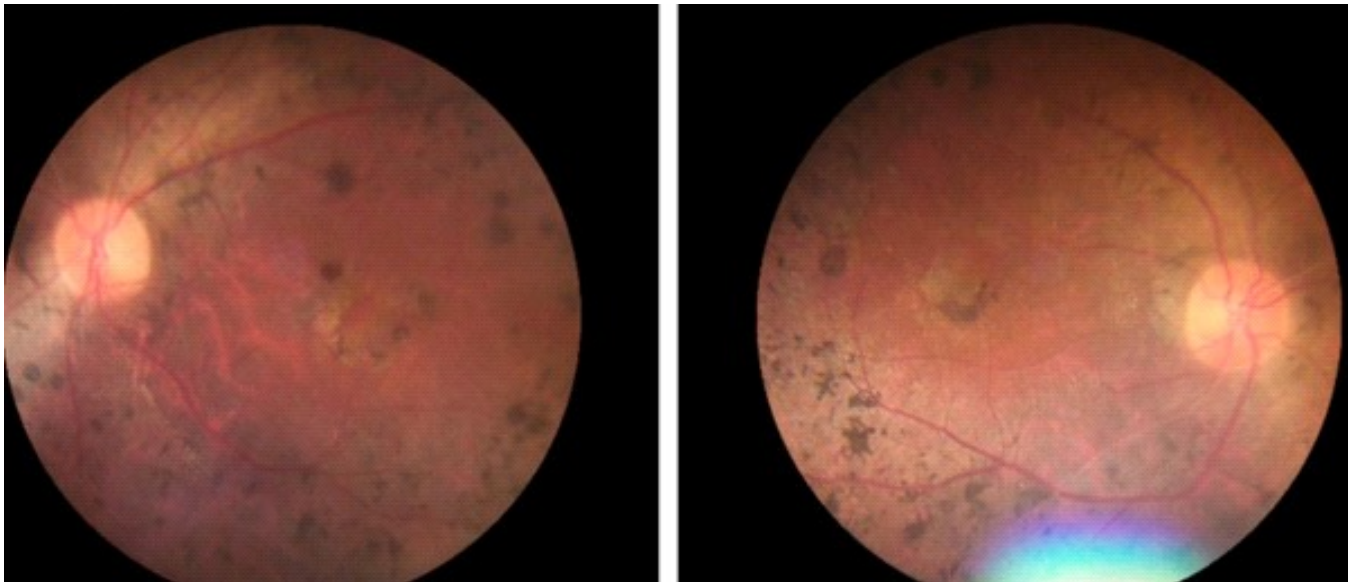

Fig 1. Fundus images of right and left eye (bony spicules, waxy yellow discs)

ak prognosis associated with RP, and he was referred to our hospital. The patient is a typist by profession.

At the time of his first visit to our hospital on 23rd January 2021, his symptoms included poor distance vision and reduced dark adaptation with no medical history. On examination, the ocular adnexa including lids, conjunctiva and sclera of both eyes were normal, with clear corneas and quiet anterior chambers of normal depth. He had bilateral cataract and ocular hypertension (intraocular pressure of 23 mmHg in both eyes). He was dilated with 1% tropicamide. Vitreous was clear in both eyes. Fundus examination revealed a waxy yellow disc cup disc ratio of 0.4 in both eyes. There were bone spicules in the periphery of both eyes (figure 1).

His aided visual acuity with his own glasses was 6/30 in the right eye and 6/30-2 with difficulty in the left eye measured with Snellen visual acuity chart. Near visual acuity was measured with Keeler near visual acuity chart and it was N 24 with difficulty in both eyes at his preferred working distance with his distance vision glasses. He was not using separate near vision glasses before. A refraction trial was given but there was no improvement with refraction because of the central posterior subcapsular cataract. The patient was counselled regarding cataract surgery, in the hope that it would improve his vision and started on Travatan (travoprost) to lower the intraocular pressure.

His ultimate goal was to be able to continue his job as a typist, which required him to be able to read clearly at 40 cm and to see computer screens. He demanded to be fit with stronger glasses and hoped for visual rehabilitation. His increasing dependence on his wife had rendered him frustrated and he had to stop hobbies such as reading newspapers and writing because of his progressive loss of vision. He was slowly confining himself, especially at night, and rarely visited friends and relatives due to the difficulty he faced navigating new localities. He was struggling with basic tasks such as selecting clothes and climbing staircases. He was constantly worried about his eyesight and about embarrassing himself and his family due to his condition.

A questionnaire was filled in in order to assess the patient's general health, ability to carry out basic daily tasks like getting dressed, finding items, reading etc. and the emotional/mental

effects of the disease on the patient.

An uneventful cataract surgery was performed on the left eye on 4th April 2021. He was happy with the visual outcome a day after the surgery as his unaided visual acuity was 6/60 in the operated eye. There were no remarkable findings on examination and the intraocular lens was in position. An uncomplicated right cataract surgery was performed 3 weeks later, on 27 April 2021.

After successful phacoemulsification of both eyes, he attended on 20th May 2021 for a follow-up visit with chief complain of blurred distance and near vision. Intraocular lenses of both eyes were in position. No active remarkable findings were present. His unaided visual acuities were 6/60 in both eyes for distance and N24 at near vision, using Snellen's and Keeler Near vision charts. With refraction trial his best corrected distance visual acuity was 6/18 in both eyes measured with Snellen vision chart and the best-corrected near vision was N18 on Keeler near vision chart at his preferred working distance. He was happy with the new correction. Patient was not satisfied with hand held and stand held magnifier due to a small field of view and preferred bifocal glasses for his computer work. Distance portion of bifocal was corrected for intermediate /computer distance and the near portion was corrected for standard paper text from which he has to type. He was given separate distance vision glasses and bifocal glasses for his computer and near work. He was recommended polycarbonate lenses with antiglare and blue cut effect for clear vision.

The same questionnaire was filled again on his follow up visit on 4th December 2021. It demonstrated that now he navigates around his house with ease, is even able to roam around his locality at night, and requires very little assistance in a familiar setting. However, he still struggled with new environments. He now enjoys watching television and watching videos on his phone with no difficulty at all and is fully capable of carrying out work-related documentation and activities, including those that require him to read text and official documents.

Overall, he has a much more positive outlook towards his eye-sight. Although he still suffers from the occasional bouts of frustration or anxiety, as a whole he feels less worried about his vision. He is happy to be more independent with everyday tasks such as picking out clothes, finding objects from crowded shelves, and can easily locate stationery and paperwork at his job, making work enjoyable with support from his colleagues.

Discussion

RP is one of the commonest forms of inherited blindness, affecting more than 1 million people worldwide, hitting Pakistan particularly hard due to frequent cousin marriages[3,7]. Our case reviews the poor quality of life of a 42 years old male typist with RP. Visual impairment due to RP has a negative impact on the performance of an individual's daily activities, such as self-grooming, driving, and cooking. The ability to complete daily tasks gradually decreases as the disease progresses[1], as was evident in our patient.

RP has greater psychological and emotional effects as compared to patients with other major blinding diseases such as age-related macular degeneration and diabetic retinopathy, as these diseases are more curable and occur later in life as compared to RP that has an earlier onset and is not treatable[5]. A meta-analysis performed by G Garip and A Kamal investigated coping strategies to help improve the quality of life. These included maintaining autonomy and independence, dealing with practical and emotional issues, experiences with healthcare professionals and social support workers, coping strategies (positive and negative), and the effects on the career of the patient[5]

Rehabilitation is an effective way of improving the quality of life of patients with low vision and blindness which cannot be treated or prevented[8]. In our patient the successful cataract surgeries and low vision rehabilitation played a remarkable role in improving his personal as well as professional life, consequently reducing his stress and depression levels.

The level of anxiety in RP patients was significantly related to the limitation of the NEI-VFQ25 questionnaire[9]. Quality of life can be significantly increased by improving the reading speed of RP patients[10], as was evident in the comparison of pre-rehabilitation and post-rehabilitation questionnaires of our patient. As his reading quality improved, we also noticed a positive impact on his overall wellbeing and mental health, as this significantly impacted his career and professional life.

Conclusion

Retinitis Pigmentosa can have a detrimental effect, not only on the vision of sufferers, but also on their mental health. Our report's aim is to highlight the importance of diligent and effective low vision rehabilitation, which can in turn greatly reduce visual impairment related stress in RP patients. This is very clearly demonstrated in our patient's VFQ score, which was considerably improved after his cataract surgery and low vision rehabilitation as compared to his first visit.

Declarations

Consent regarding publishing has been acquired from the patient. In order to maintain confidentiality, data generated and analyzed during this study are not publicly available. However, they are available from the corresponding author on reasonable request, and with permission from the patient.

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