Psychological Disorders in Patients with Retinitis Pigmentosa in Iran

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Abstract
Background: To identify mental disorders and their prevalence in patients with retinitis pigmentosa (RP).
Methods: This descriptive study was carried out between January 2009 and January 2010 on 417 patients with RP, who were members of Iran RP Center. The necessary data were collected using questionnaires consisting two parts: The background characteristics and questions assessing the mental health and screening personality and psychosocial disorders, which were designed based on the Minnesota Multiphasic Personality Inventory. Data were analyzed using SPSS software with Chi-square test to assess the relationship between background characteristics and each mental disorder. Scores in the range of 30 and 70 were considered normal.
Results: Patients with RP suffered from eight mental disorders with the following prevalence: Obsessive compulsive disorder (39.3%), schizophrenia (38.1%), antisocial personality (37.6%), paranoia (36.7%), hypochondrias (35.3%), depression (31.2%), hysteria (26.9%), and hypomania (23.7%). No one had all the eight mental disorders simultaneously. Statistical analysis showed no significant relationship between obsessive compulsive disorder, paranoia, depression, and hysteria and background characteristics. A significant association was found between schizophrenia and onset of RP (P = 0.047). Furthermore, a significant association was seen between hypochondriasis and educational level (P = 0.026) as well as income (P = 0.037), and smoking (P = 0.009). There was also a significant association between hypomania and marital status (P = 0.027).
Conclusion: The findings showed that RP might lead to various mental disorders, especially obsessive compulsive disorder.

Keywords: Retinitis pigmentosa, Mental disorder, Prevalence, Iran

Introduction

Retinitis pigmentosa (RP) is a hereditary disorder, in which the progressive loss of rod and cone photoreceptor cells occurs. Patients with RP generally experience loss of night vision (nyctalopia) in adolescence, side vision in young adulthood, and central vision (tunnel vision) in later life (1-4). About 14% of RP may accompany Usher syndrome, which consists of hearing loss as well (1, 5). Retinitis pigmentosa is one of the most common causes of visual impairment in all the age groups with the prevalence of between 1/3000 and 1/5000 worldwide (3, 6-10). Retinitis pigmentosa is a common and significant finding in Iranian population as well. In studies done in cities of Shiraz and Shahroud, RP was found to be the 1st and the 3rd most common cause of visual impairment, respectively (11, 12).

Retinitis pigmentosa may be inherited as dominant autosomal, recessive autosomal, or X-related traits. However, it may sometimes be seen sporad-
Psychological disorders in patients with RP may develop a kind of a psychological disorder in a period of their life. Ignoring their mental disorder will deteriorate the RP course and finally result in their rejection by the society. Therefore, attempt to prevent, diagnose, and treat the psychological problems are necessary to improve the overall quality of life in patients with RP. To the best of our knowledge, this is the first study on assessing the common psychological disorders in patients with RP in Iran.

Materials and Methods

This descriptive, cross-sectional study was carried out between January 2009 and January 2010 on patients with RP. All the 939 patients who were members of Iran RP Center in Tehran were recruited to the study through census sampling method. This study was approved by the Ethics Committee of Nursing and Midwifery School at Islamic Azad University, Tehran Medical Branch as well as Iran RP Center. Then, the study goals were explained to the patients and a written informed consent was taken from each participant. Being older than 18 years, residence of Tehran, member of Iran RP Center, and suffering from RP for at least one year were considered as inclusion criteria. Those with mental retardness or with a history of a diagnosed mental disorder prior to RP were excluded from the study. The necessary data were collected using questionnaires composed of two parts: The first part was about socio-demographic characteristics, including age, gender, marital status, education, occupation, income, type and location of accommodation, the family relationship between parents, the onset of RP, the history of RP amongst first-degree relatives, and cigarette, drugs, or alcohol consumption (14 questions). The second part composed of questions assessing the mental health and screening personality and psychosocial disorders, which were designed based on the modified Minnesota Multiphasic Personality Inventory-2 (MMPI-2) (71 questions). The MMPI-2 is an inventory assessing mental health in psychiatric and medical settings in adults above the age 18, which has been standardized for Iranian population by Moootabi and Shahrami, and the validity and reliability of the questionnaire were confirmed (15).

To examine the scientific reliability of the instrument, the Cronbach’s alpha index was applied. Eventually, 417 subjects remained for final analysis. Data were analyzed using SPSS software (the Statistical Package for the Social Sciences, Version 16.0, SPSS Inc., Chicago, Illinois, USA) through Chi-square test to assess the relationship between socio-demographic characteristics and each mental disorder. Scores in the range of 30 and 70 were considered normal.

Results

The socio-demographic characteristics of the patients with RP are demonstrated in Table 1. As it shows, the majority of the participants were men (55.4%) in the age range of 26 to 35 years, and single (48.7%). Furthermore, most of the patients experience RP from their childhood (40.9%) and had the parental family relationship (60.7%). Based on MMPI questionnaire, patients with RP suffered from eight mental disorders with the following prevalence: Obsessive compulsive disorder (39.3%), schizophrenia (38.1%), antisocial personality (37.6%), paranoia (36.7%), hypochondrias (35.3%), depression (31.2%), hysteria (26.9%), and hypomania (23.7%). No one had all the eight mental disorders simultaneously. Table 2 depicts the frequency of coexisting mental disorders.
Table 1: Socio-demographic characteristics of patients with retinitis pigmentosa

| Variables                          | Number, n | Percent |
|-----------------------------------|-----------|---------|
| Total                             | 417       | 100     |
| Gender                            |           |         |
| Male                              | 231       | 55.4    |
| Female                            | 186       | 44.6    |
| Age, years                        |           |         |
| 18-25                             | 64        | 15.5    |
| 26-35                             | 138       | 33.6    |
| 36-45                             | 90        | 21.8    |
| 46-55                             | 71        | 16.5    |
| >55                               | 54        | 12.5    |
| Marital Status                    |           |         |
| Single                            | 206       | 48.7    |
| Married                           | 188       | 45.7    |
| Divorced                          | 6         | 1.5     |
| Separated                         | 2         | 0.5     |
| Widowed                           | 15        | 3.6     |
| Educational Status                |           |         |
| Primary School                    | 59        | 14.2    |
| Secondary School                  | 121       | 29.2    |
| Diploma                           | 126       | 30.4    |
| Post-Diploma                      | 30        | 7.2     |
| Bachelor                          | 56        | 13.5    |
| Master                            | 16        | 3.9     |
| PhD/Doctorate                     | 9         | 1.7     |
| Occupation                        |           |         |
| Unemployed                        | 130       | 31.2    |
| Student                           | 55        | 13.2    |
| Clerk                             | 71        | 17.0    |
| Laborer                           | 21        | 5.0     |
| Self-Business                     | 47        | 11.3    |
| House-wife                        | 68        | 16.3    |
| Retired                           | 25        | 6.0     |
| Type of accommodation             |           |         |
| Being Owner                       | 258       | 62.2    |
| Being Tenant                      | 154       | 36.6    |
| Living in Home organization       | 5         | 1.2     |
| Age of retinitis pigmentosa onset |           |         |
| Infancy                           |           |         |
| Childhood                         | 20        | 4.8     |
| Adolescence                       | 171       | 40.9    |
| Adulthood                         | 122       | 29.3    |
| 104                               | 25.0      |
| Location of residence/ North      |           |         |
| South                             | 70        | 16.8    |
| East                              | 111       | 26.6    |
| West                              | 71        | 17.0    |
| Center                            | 111       | 26.6    |
| Parental family relationship      |           |         |
| Yes                               | 253       | 60.7    |
| 164                               | 39.3      |
| Family history of RP amongst 1st-degree relatives | | |
| Yes                               | 101       | 24.2    |
| 316                               | 75.8      |
| Income, Toman/ < 350 000           |           |         |
| 20                                | 4.8       |
| 350 000-500 000                    | 124       | 29.1    |
| 500 000-700 000                    | 224       | 54.2    |
| > 700 000                         | 49        | 11.9    |
| Smoking cigarette or other tobaccos |           |         |
| Yes                               | 34        | 8.2     |
| No                                | 383       | 91.8    |
| Consuming alcohol                 |           |         |
| Yes                               | 22        | 5.3     |
| No                                | 395       | 94.7    |
Table 2: The frequency of mental disorders in patients with retinitis pigmentosa

| Variables       | Positive | Negative |
|-----------------|----------|----------|
|                 | Number   | Percent  | Number   | Percent  |
| Hypochondrias   | 147      | 35.3     | 270      | 64.7     |
| Hysteria        | 112      | 26.9     | 305      | 73.1     |
| Paranoia        | 153      | 36.7     | 264      | 63.3     |
| Antisocial      | 157      | 37.6     | 260      | 62.4     |
| Obsessive compulsive | 164 | 39.3 | 253 | 60.7 |
| Depression      | 130      | 31.2     | 287      | 68.8     |
| Schizophrenia   | 159      | 38.1     | 258      | 61.9     |
| Hypomania       | 99       | 23.7     | 318      | 76.3     |

The Cronbach’s alpha value was 0.8, which is satisfactory, and the power of study was 80% (if $\beta = 20\%$, $1 - \beta = 80\%$). Statistical analysis showed no significant relationship between obsessive compulsive disorder, paranoia, depression, and hysteria and background characteristics ($P > 0.05$). A significant association was found between schizophrenia and onset of RP ($P = 0.047$). Furthermore, a significant association was seen between hypochondrias and educational level ($P = 0.026$) as well as income ($P = 0.037$), and smoking ($P = 0.009$). There was also a significant association between hypomania and marital status ($P = 0.027$).

Discussion

In our study, of 417 patients with RP, 231 (55.4%) were men, which is compatible with other studies. Because RP has x-related trait as well (13, 14), it is more prevalent in men while women are mostly carriers. Since the diagnostic value of MMPI-2 test decreases in subjects less than 18 years, we recruited patients above 18 years to our study. The most and the least of our patients have experienced RP from their childhood and infancy, respectively. This may be due to disability of infants to express the visual symptoms. Besides, since the specific amblyopic examination is done in childhood, the diagnosis of RP increases in that life period. Furthermore, because the parents are aware of the genetic transmission of RP to their children, they mostly take their child to an ophthalmologist for specific examinations, such as electroretinography.

Of 417 patients in our study, 60.7% had parental family relationship. Retinitis pigmentosa is a hereditary disease; hence, consanguineous marriage increases its frequency. This is in line with other studies, including those performed in Iran (11, 12). Mehdizadeh et al. performed a study in Shourideh Educational Center, which is the only educational center for blinds in Shiraz, in the South of Iran, and has students at different levels, from preschool to high school. Of 145 students, 65 (44.8%) had RP, which was the single most common cause of visual loss. Large proportion of these blind children (47.5%) had a positive history of consanguineous marriage in their parents (first cousins marriage) (12). Consanguineous marriage seemed to be the most probable cause of increased frequency of genetic etiologies of childhood blindness. Therefore, avoidance of consanguineous marriage may reduce the number of blindness caused by RP.

In a study on 970 patients with RP, significant depression, anxiety, and phobia were reported ($P < 0.0001$) (16). Risk of depression in patients with RP was 25.1% more than that in the general population (17). Patients with RP that experienced depression in the same period had poorer visual function in comparison with those without depression ($P < 0.0005$) (17).

In 2009, Rijavec and Grubic reported a 28-year-old woman with Usher syndrome (RP with hearing impairment), who developed psychological disorders, including eating disorder, psychosis, panic, anxiety, and obsessive compulsive disorder (18). In another study in 2012 on 26 children with Usher syndrome, (23%) had a mental and behavioral disorder (19).

John Hopkins University followed up 50 patients with RP from 2007 to 2010 and reported that
depression developed in patients with RP. The more depressed they were, the less visual function they had (17). In a study carried out on 144 patients with RP, relationship between depression and the vision-related quality of life was assessed. It was concluded that RP patients with depression had poorer vision-related functions. Therefore, it was suggested to diagnose and treat depression in order to enhance the overall quality of life in patients with RP (20). Depression was more frequent in patients with RP than those with glaucoma \((P<0.0005)\) (21).

Our results showed that depression was mostly reported in women with RP, especially those who were divorced and were in the age range of 46 to 55 years. Generally, depression is more prevalent in women. Besides, getting divorced and increasing age are themselves predisposing factors for depression as well (22).

Yet no treatment modality has been introduced for RP. Therefore, patients should be addressed towards rehabilitating institutions that help them obtain new skills and return to their life. Furthermore, psychiatric counseling is required for patients with RP to accept and adjust to their condition.

Conclusion

The findings showed that RP might lead to various mental disorders, especially obsessive compulsive disorder. Therefore, psychological support, which can be provided by either professionals or supportive patients’ family, is often crucial in the course of RP. Further studies are required to evaluate the effect of orientation and training on the mental disorders in RP as well as comparing the psychological disorders in RP with other ophthalmological diseases.

Ethical considerations

Ethical issues (Including plagiarism, Informed Consent, misconduct, data fabrication and/or falsification, double publication and/or submission, redundancy, etc.) have been completely observed by the authors.

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The authors declare that there is no conflict of interests.

References

1. Hamel C (2006). Retinitis pigmentosa. Orphanet Journal of Rare Diseases,1(1):40.
2. Kanski JJ (2009). Clinical ophthalmology: a synopsis. 6th ed. Butterworth-Heinemann Medical, England.
3. Sahel J, Bonnel S, Mrejen S, Paques M (2010). Retinitis pigmentosa and other dystrophies. Dev Ophtalmol, 47:160-7. 4.
4. Hood DC, Birch DG (1996). Abnormalities of the retinal cone system in retinitis pigmentosa. Vision Res, 36(11):1699-709.
5. Boughman JA, Vernon M, Shaver KA (1983). Usher syndrome: definition and estimate of prevalence from two high-risk populations. J Chronic Dis, 36(8):595-603.
6. Ammann F, Klein D, Franceschetti A (1965). Genetic and epidemiological investigations on pigmentary degeneration of the retina and allied disorders in Switzerland. J Neurol Sci, 2(2):183-96.
7. Boughman JA, Conneally PM, Nance WE (1980). Population genetic studies of retinitis pigmentosa. Am J Hum Genet, 32(2):223-35.
8. Jay M (1982). On the heredity of retinitis pigmentosa. Br J Ophthalmo, 66(7):405-16.
9. Puech B, Kostrubiec B, Hache JC, Francois P (1991). Epidemiology and prevalence of hereditary retinal dystrophies in the Northern France. J Fr Ophthalmo, 14(3):153-64.
10. Berson EL, Rosner B, Sandberg MA, Hayes KC, Nicholson BW, Weigel-DiFrancesco C, et al (1993). A randomized trial of vitamin A and vitamin E supplementation for retinitis pigmentosa. Arch Ophthalmo, 111(6):761-72.
11. Hashemi H, Khabazkhoob M, Emamian MH, Shariati M, Fotouhi A (2012). Visual impairment in the 40- to 64-year-old population of Shahroud, Iran. Eye (Lond), 26(8):1071-7.
12. Mehdizadeh M, Afarid M, Atrazadeh A (2005). Causes of Childhood Blindness among
Students of Blinds' School in Shiraz, Iran. IJM5,30(2):58.

13. Smith AJ, Bainbridge JW, Ali RR (2009). Prospects for retinal gene replacement therapy. Trends in Genetics,25(4):156-65.

14. Hassan-Karimi H, Jafarzadehpur E, Blouri B, Hashemi H, Sadeghi AZ, Mirzajani A (2012). Frequency Domain Electroretinography in Retinitis Pigmentosa versus Normal Eyes. J Ophthalmic Vis Res,7(1):34-8.

15. Mootabi F, Shahrami E (1988). Psychometric characteristics of MMPI-2 in apparently healthy population in Tehran, Iran [Master thesis for clinical psychology]. Iran University of Medical Sciences, Iran.

16. Strougo Z, Badoux A, Duchanel D (1997). Psychometric problems associated with retinitis pigmentosa. J Fr Ophtalmol,20(2):111-6.

17. Hahm BJ, Shin YW, Shim EJ, Jeon HJ, Seo JM, Chung H, et al (2008). Depression and the vision-related quality of life in patients with retinitis pigmentosa. Br J Ophthalmol,92(5):650-4.

18. Rijavec N, Grubic VN (2009). Usher syndrome and psychiatric symptoms: a challenge in psychiatric management. Psychiatria Danubina,21(1):68.

19. Dammeyer J (2012). Children with Usher syndrome: mental and behavioral disorders. Behav Brain Funct,8:16.

20. Hahm B-J, Shin Y-W, Shim E-J, Jeon HJ, Seo J-M, Chung H, et al (2008). Depression and the vision-related quality of life in patients with retinitis pigmentosa. British Journal of Ophthalmology,92(5):650-4.

21. Igarashi Y, Sato E, Ito A, Miyachi O, Ikejiri M, Hanawa T, et al (2003). Comparison of Yatabe-Guilford personality test results in retinitis pigmentosa and glaucoma patients. Jpn J Ophthalmol,47:1-5.

22. Sadock BJ, Sadock VA (2010). Kaplan and Sadock's pocket handbook of clinical psychiatry. 5th ed. Lippincott Williams & Wilkins, USA.

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