Schizophrenia and Retinitis Pigmentosa – A case report

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Abstract

It is common for a psychiatrist to treat patients with co-morbid medical conditions. If there are co-existing sensory impairments like deafness or blindness, in the patients with the underlying psychiatric symptoms, then diagnosis and treatment becomes a challenging task. Retinitis pigmentosa (RP) is a progressive degeneration of retinal neuroepithelium which leads to impairment of visual acuity and visual fields ultimately ending in blindness. It is often found to involve other structures, particularly the nervous and endocrine systems. We are presenting a similar case that had RP and later started having symptoms of schizophrenia which was treatment resistant.

Keywords: Night blindness, Schizophrenia, Retinitis Pigmentosa.

Introduction

Retinitis pigmentosa (RP) refers to a group of hereditary retinal disorders.[1,2] It is characterized by the progressive loss of photoreceptor cells namely rods and cones. Patients with RP generally experience night blindness (nyctalopia) during adolescent period, impairment in peripheral field vision in young adulthood, and disturbance in the central vision in later parts of life.[3-5] RP is one of the commonest causes of visual impairment in all the age groups with the prevalence of 0.02% to 0.03% worldwide.[6,7] RP may be inherited as autosomal dominant, autosomal recessive or x-linked. However, it is sometimes seen sporadically and without the family history.[8,9] As per the research, nearly 14% of RP may accompany Usher syndrome, which consists of hearing loss also.[10,11] Patients with RP may develop various psychological disorders during their lifetime. If we ignore their psychiatric illnesses, it will deteriorate the course of RP. There are limited reports about the difficulties in the evaluation, management and treatment of psychiatric illnesses in a deaf-blind population.[12] Therefore, it is essential to diagnose, and treat the psychological problems to improve the overall quality of life in these patients.

Case Presentation

Mr. A, 59 yrs old, is a married male, educated till tenth standard, from a middle socio-economic status family, from urban background. He was working earlier as line man in Karnataka Electricity Board, currently not working. He was accompanied by his wife and son as informants. He presented with complaints of suspiciousness on wife for the past 27 years, complete loss of vision since 25 yrs and visual hallucinations since 19 years. Initially he had difficulty in seeing things at night and diminution of vision. Subject was taken to a faith healer and was told that someone had done black-magic on him. He started suspecting the wife that she had done black-magic on him and used to act out and beat her multiple times and was not convinced even if someone told him that it was false. Conviction became stronger after 2 years when he became completely blind and was shown to multiple ophthalmologists but was told he would not be cured. He did not have any complaints from the work place. He was taken to faith healer again in different place and was given a coconut for worshipping so that his problems would come down. Since then he started suspecting that two of his close relatives were against him and trying to harm him and was also suspecting the wife that she was also involved in it. He did not believe if his son or brother told it was false. He started saying that he could see a lady coming out of the coconut while he used to worship God and he used to see her initially two to three times a day and subsequently he started seeing the same lady multiple times around 10-15times a day. It was as clear as he was seeing a lady when he was not blind at a distance of around one meter. He used to tell it to family members but they were not seeing anyone. He started telling that he used to hear voices of a male person talking outside the house and was asking the family members to check outside. At times he used to scold and beat wife as he used to suspect her of having an affair because he used to hear sound of somebody opening the zip of trouser. His sleep was very much disturbed and used to get up multiple times saying some snake had come into house. Appetite was normal. Self care was adequate. Patient was brought for treatment mainly because of persistence of symptoms like suspiciousness on wife, visual hallucinations and sleep disturbances even on medications. He was taken to a private hospital 11 years ago for evaluation of blindness and psychiatric problems and was admitted and diagnosed to have complicated cataract possibly RP. He underwent

bilateral cataract surgery with intra-ocular lens implantation. Based on the treatment history, in spite of being compliant to all the medications in the past, he had failed monotherapy trials with risperidone, aripiprazole and olanzapine. He was tried with risperidone upto 8mg per day for three years, sleep was improved but suspiciousness and visual hallucinations persisted. Later when he was prescribed olanzapine 15mg per day for 3 years, sleep was better, visual hallucinations reduced in frequency but the persecutory delusions persisted. There was no change with aripiprazole 20mg per day even after trying for eight weeks. There was no past history of mental illness. As per the family history, he was born of third degree consanguineous marriage with birth order as three and there was history of visual impairment possibly retinitis pigmentosa in multiple first degree relatives which includes subject's elder sister, elder brother and younger brother. There was no history of substance abuse like alcohol, cocaine, LSD, cannabis, opioids. There was no history suggestive of any high risk sexual behavior. He had been well behaved, social, energetic and average in studies and was premorbidly well-adjusted A full work-up including general physical examination revealed no abnormality. Ophthalmological examination revealed perception of light in both eyes and RP. Mental status examination revealed persecutory delusions, visual hallucinations, olfactory hallucinations & somatic passivity, impaired judgment and insight was absent. Routine investigations revealed no abnormality as fasting blood sugar was 101mg/dl, hemoglobin was 11.2g/dl, thyroid profile and levels of vit-B12 in serum were within normal limits. MRI brain showed generalized cerebral and cerebellar atrophy. Electroencephalography was normal. When the subject presented to our hospital, he was treated with a combination of aripiprazole 20mg/day and olanzapine 5mg/day for 3 weeks without much improvement in his visual hallucinations and persecutory delusions. Clozapine was started considering this as treatment resistant case, and maintained up to 200 mg/day and after 4 weeks of the treatment patients psychopathology got improved but persisted to have visual hallucinations.

Discussion

Mental subnormality is a prominent part of most of the retinitis pigmentosa syndromes, the important ones being the Laurence-Moon- Biedl Syndrome, Refsum's syndrome, Usher's Syndrome, Cockayne's Syndrome and Hallgren's Syndrome.[13-16] However, there are very few case reports of documented case of schizophrenia in association with retinitis pigmentosa without any other abnormality. A study from Iran evaluated 417 patients with retinitis pigmentosa which reported that schizophrenia was the co-morbidity in 38.1% of patients.[17] As per the literature review, there are very few articles about RP and psychiatric disorder with the majority of them focusing on pathophysiology and etiology of the psychiatric disorders in Ushers Syndrome.[18] Research suggest that there is a link between schizophrenia and retinitis pigmentosa associated syndromes. Even in the absence of any consistently replicated linkage findings in schizophrenia, there is some evidence for genes at certain chromosomal regions, especially at 6p24-p22 and 8p22-2, to be associated with the condition.[19] Schizophrenia along with many other metabolic disturbances and RP have been found to have coding regions in chromosome 6 encoding the major histocompatibility complex involved in immune regulation.[20] It is possible to hypothesize that RP associated syndromes, retinoid cascade and schizophrenia have similar gene loci and may have common pathogenic mechanisms. We want to conclude by saying that we are yet to find the reason what might have contributed for the treatment resistant condition in our patient and posed a challenge in treating this case. As a consequence, it would be difficult for the mental health professionals and nursing staff to handle and treat these kinds of complicated patients. As suggested by some authors it is diagnostically important to consider the etiological, developmental, adaptive and social factors in interpreting patient condition. Our diagnostics and pharmacological treatment did not differ much from other published cases; however the best results were achieved with a start of clozapine. Still, there is a need to unravel the management of schizophrenia in the cases like above.

Limitations

We did not use any scales for assessing the psychopathology or for comparing the response with medications.

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