A Rare Case of Psychosis in Ushers’s Syndrome in Absence of Hallucinations

Halüsinasyon Yokluğunda Ushers Sendromunda Nadir Görülen Bir Psikoz Olgusu

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Abstract

Usher syndrome is a clinically and genetically heterogeneous syndrome leading to hearing and vision loss. Three types of Ushers syndrome have been described based on severity of hearing loss and presence or absence of vestibular symptoms. Although several mental disorders have been seen to be associated with Ushers syndrome, the awareness of the same is lacking in the medical community. There have been only few case reports of Ushers syndrome and psychosis, with visual and auditory hallucinations being the predominant feature in these cases. Hereby the author discusses a unique case of ushers syndrome and psychosis in absence of any hallucinations.

Key words: Usher's syndrome, psychotic disorders, case reports, rare diseases

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INTRODUCTION

Usher’s syndrome is a heterogeneous autosomal recessive disorder, primarily associated with sensori-neural hearing loss, retinitis pigmentosa and in some variants vestibular dysfunction. It is also known as Hallgren syndrome. Usher-Hallgren syndrome, Retinitis pigmentosa-dysacusis syndrome and Dystrophia retinae dysacusis syndrome. Usher’s syndrome has a worldwide prevalence of 3.2-6.2/100,000 (1,2). Although no there is no literature on prevalence of Usher’s syndrome in India, in a longitudinal study (3) 14.28% of patients with syndromic deafness were diagnosed with Usher’s syndrome. Epidemiological studies report varying prevalence rates of mental disorders in these patients ranging from 15% to 60% (4). Herein, we report a case of a middle aged man with Usher’s syndrome with psychosis.

CASE

Mr. S, 35 year old male patient presented to the psychiatric out-patient department (OPD) in a tertiary care center with episodes of altered behaviour since past 3 years, most recent episode since 12 days. He also had a history of decreased vision since past 10 years and hearing difficulty since 8 years. He presented with an acute episode of suspiciousness 3 years back with strong beliefs of black magic being performed on him, and was extremely fearful as he felt his family members and co-workers were trying to harm him. He had been taking olanzapine for a few days of initiating treatment with minimal side effects. He reached pre-morbid levels in 2 months in is currently on regular follow-ups.

DISCUSSION

The data regarding Ushers syndrome and associated psychosis is conflicting, with some studies showing prevalence 4.5% (6) while others as high as 23% (7). Most common form psychosis diagnosed in these individuals was of schizophreniform spectrum. It was found that visual and auditory hallucinations were predominant in these individuals. The discussed above was also diagnose as acute schizophrenia-like psychosis. However, the highlight of this particular case is the absence of auditory hallucinations with presence of made phenomenon. There has been no consensus on whether there is deficient development of the theory of mind abilities in pre-lingually deaf patients. Studies have shown that there is deficient development of the theory of mind abilities in pre-lingually deaf patients contributing to the development of delusional beliefs (13).

Another theory of relevance is stress-related theory. The chronic stress due to auditory and visual impairment leads to increased vulnerability in the patient for psychiatric disorders such as depression and psychosis (9). The current study highlights the presence of psychiatric illness in individuals with Ushers syndrome. This puts light on the importance of educating the ophthalmologists and otolaryngologists regarding recognition of mental illnesses in these individuals and timely referrals, thereby providing them with holistic treatment.

Consent: Written informed consent was obtained from the patient for publication of this case report.

Conflict of interest: Authors declare that there is no conflict of interest between the authors of the article.

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