

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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Öz

Usher sendromu işitme ve görme kaybına yol açan klinik ve genetik olarak heterojen bir sendromdur. Üç tip Ushers sendromu, işitme kaybının şiddetine ve vestibüler semptomların varlığına veya yokluğuna bağlı olarak tanımlanmıştır. Ushers sendromu ile ilişkili birkaç zihinsel bozukluk görülse de, tıp camiasında aynı farkındalığın eksikliği görülmektedir. Ushers sendromu ve psikozu ile ilgili sadece birkaç olgu sunumu olmuştur ve bu olgularda görsel ve işitsel halüsinasyonlar baskın özelliştir. Bu nedenle yazar, herhangi bir halüsinasyon olmaması durumunda benzersiz bir ushers sendromu ve psikoz vakasını tartışır.

Anahtar Kelimeler: Usher sendromu, psikotik bozukluklar, olgu sunumları, nadir hastalıklar

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 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. It was found that patient gradually stopped eating food at home thinking it was poisoned and his interaction with family members was also markedly reduced. He stopped going to the work and stayed aloof. His self care also deteriorated. 1 month following the onset of symptoms family members took him to a psychiatrist, where he was prescribed medications. According to the family he took the medications for a period of 2 months and stopped it on his own as he attained pre morbid levels of functioning.

Mr. S was brought to our OPD with 12 days history of similar symptoms in the past along with complaints that his actions and feelings are being controlled by someone else. On mental status examination patient was unkempt, perplexed, restless, decreased volume and tone of speech, delusions of persecution, made act and made affect with anxious mood. Judgment was impaired with absent insight. A psychiatric diagnosis of acute schizophrenia – like psychotic disorder was made according to International Classification of Disorder – 10 (ICD-10). A differential diagnosis of

paranoid schizophrenia was considered because of the past history.

Family history revealed that his maternal cousin brother suffered from Deafness and Hearing loss and expired at a young age. Moderate to severe sensori-neural deafness was found in both ears on Pure-Tone Audiometry. Ophthalmological check-up revealed bilateral retinitis pigmentosa with tunnel vision. Other blood investigations were within normal limits. MRI of brain was essentially normal. He had no other medical/ Psychiatric co-morbidities. A diagnosis of Ushers Syndrome Type 2 was made based on Davenport and Omenn criteria (5). Mr. S was started on olanzepine 5 mg, gradually increased to 10 mg in 3 weeks. He started showing improvement within 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 concrete evidence for the causation of psychosis in Ushers syndrome. However various theories have been put forward. A common gene hypothesis for the causation of both ushers and psychosis has been put forward. Schizophrenia is a polygenic disorder. Genetic loci 11q in type I and 5q in type II associated with Usher's syndrome are also implicated in schizophrenia. In addition DISC1 on chromosome 1q42.2 which is a possible causative gene in schizophrenia is on the same chromosome and close to the USH2A gene (8-11). Neuro-anatomical similarities like smaller brain volume and focal and diffuse atrophic changes in the supratentorial brain has been noted in both the conditions (9,12). Studies have shown that there is deficient development of the theory of mind abilities in prelingually 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 throws light on the importance of educating the ophthalmologists and oto-laryngo-rhinologists 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.

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