Case Report

BIOMEDICAL Journal of Scientific & Technical Research

ISSN: 2574 -1241

DOI: 10.26717/BJSTR.2022.45.007150

# Schizophrenia-like Psychosis and Other Psychological Manifestations of Huntington Disease

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**ARTICLE INFO**

**Received:** July 04, 2022

**Published:** July 13, 2022

**Citation:** Abu Zarrin Zakaraia, Hazura Hamzah, Nor Ilyani Abdullah, Chong Siew Koon. Schizophrenia-like Psychosis and Other Psychological Manifestations of Huntington Disease. Biomed J Sci & Tech Res 45(1)-2022. BJSTR. MS.ID.007150.

**ABSTRACT**

Huntington Disease (HD) is a progressive neurodegenerative disorder with hallmark features of movement disorders, impaired cognitive functions and neuropsychiatric manifestations. Management for HD is usually multidisciplinary as its physical manifestations are equally debilitating as its psychological features. There is no definitive cure for HD to date, and mortality generally ensues between 5 to 20 years from the onset of illness.

**Keywords:** Huntington; Schizophrenia; Psychosis

## Introduction

Huntington Disease (HD) is an autosomal dominant neurodegenerative disorder, characterized by movement disorders, impaired cognitive functions and neuropsychiatric manifestations. It affected men and women equally with mean age of onset at 40 years old [1]. While uncontrolled movements especially on the upper parts of the body often present as the hallmark feature of the disease, as much as 6%-25% of patients with HD also suffers from neuropsychiatric presentations [2]. We describe a case of HD with schizophrenia-like psychosis together with cognitive impairment and movement disorder, which complicates the overall prognosis of illness.

## Case Report

Mr. M, a 53 year old Malay gentleman, was a retired teacher diagnosed with Huntington Disease by Neurological department at the age of 50 years old. It started with subtle but progressive cognitive decline 7 years prior to presentation, followed by involuntary movement 2 years later. Disturbing psychotic symptoms subsequently emerged in 2019, prompting family members to seek treatment at Hospital Tanah Merah (HTM). Mr. M had strong family history of HD. His father was diagnosed with HD and passed away a few years after suffering from the illness. All of patient's four siblings experienced a similar fate, with an earlier onset compared to Mr. M. His youngest brother passed away few years after being diagnosed. The rest of his siblings are currently on treatment. Further history revealed that the deterioration of his cognitive functions was progressive, but he did not seek treatment, dismissing early symptoms as simple forgetfulness. He subsequently experienced involuntary movements (chorea), which is brief, jerky and unpredictable in nature. It would start abruptly from his left upper limb before spreading to involve all limbs, including his head. The peculiar movements were beyond his control and occurred exclusively when he was awake. Progression took an insidious course; movements which was initially painless became associated with excruciating and sustained muscle contractions.

Pain caused him great distress and adversely affected his quality of life. In 2019, psychotic symptoms started to occur, manifesting as auditory hallucinations which were mainly commanding in nature. Mr M also complained of visual hallucination saying that he saw figures of unidentified people coming to visit him at home. He began to feel insecure, which developed into a persecutory delusion towards his in-laws. As his delusion worsened, Mr M became impulsive with frequent bouts of aggression towards his family members. Ever since, Mr M required intensive psychiatric intervention and had multiple admissions due to unmanageable behavior. Psychotic symptoms improved with a combination of antipsychotics and mood stabilizer. Unfortunately, functional limitations persisted due to the worsening of involuntary movements. He admitted that his mood was low at times. Apparently, Mr M was not the only individual to be affected by the ailment. His in laws were so traumatized by his aggressive and unpredictable behavior that they refused to let him in. Instead, he was forced to stay in his mother's house, locked away in a room. He was also prohibited from seeing his wife and children. Prolonged isolation and being shunned by his own family took a heavy toll on his overall morale.

Mr M was put on acute service care by our community team when he started to express suicidal thoughts. The treating team doubled their efforts and he was visited on frequent and regular basis to monitor symptoms and to ensure treatment compliance. We held several family sessions involving patient's mother, wife, siblings, in laws and children in a concerted effort to psycho educate them regarding the nature of the illness, the expected outcomes, medication administration and strategies to deal with disturbing behavior. Family intervention was integral to help his family cope with the sequelae of his illness as well as showing them ways to manage his psychotic symptoms. Gradually, they were able to come in terms with his symptoms, becoming more supportive and accepting towards Mr M. We applied 'shared decision making' concept throughout the treatment process, to ensure that his family members actively participate in the management and future plans for Mr M. The outcome of this multimodal therapy was forthcoming and Mr M was able to come out of his isolation and participate in social as well as family activities.

## Discussion

HD is a rare autosomal dominant inherited neurodegenerative disorder. The prevalence of HD in Asian country is 0.40/100,000. Meanwhile, the prevalence of HD in Australia, North America and Europe are 5.70/100,000 [2]. The mean age of onset is approximately 40 years, and about 5% will develop symptoms before 20 years, giving rise to the term 'Juvenile Huntington Disease' (JHD) [3]. To date, there is limited information regarding the local prevalence of HD, with the first case of HD in Malaysia being described in 1994 [4]. The signs and symptoms that characterizes HD are movement disorders, cognitive impairment and psychiatric manifestation. Cognitive impairment is the core characteristic of HD and tend to develop several years prior to the diagnosis [5]. As with the case of Mr M, cognitive impairment had affected his work performance and caused great distress in his life. However, one interesting feature of this case is the delayed onset of his illness compared to his siblings. Harkening to his occupational background, cognitive demands as a teacher could possibly play a crucial role in slowing the progress of disease pathology. Such an association was also postulated by Garcia-Gorro, et al. [6]. Chorea is the prominent movement disorder in HD which could be very disturbing and painful, often leading to mood instability in HD patients. In the case of Mr. M emotional lability and irritability coincided with the bouts of pain from chorea and dystonic movements. This was further aggravated by the myriad functional limitations especially pertaining to independent mobility. Psychosis among HD patient is rare, with prevalence between 9%-17%. As rare as it may seem, tackling such issue is a daunting task. Neuroleptics had to be judiciously titrated to strike a fine balance between ameliorating psychosis and the aggravation of dystonic movements. According to Goh, et al. [7], psychotic symptoms tend to become more attenuated with the decline of cognitive function.

As cognitive function tends to predate the onset of psychosis, most patients would present to a psychiatric facility with an advanced stage of disease. In this case, psychotic symptoms occurred a few years from the onset of illness, in the form of auditory hallucinations together with paranoid and persecutory delusions. These are the symptoms which prompted Mr M's family, as well as the family of many other sufferers of HD to actively seek treatment despite having subtle warning signs prior to onset. Delayed treatment has multiple repercussion such as lower quality of life, psychological distress and a ruined familial relationship. Fortunately, Mr. M showed significant improvement after receiving treatment using multimodal approach by our team. He was treated with Tab Olanzapine for his psychotic symptoms, Sodium Valproate as mood stabilizer and Clonazepam as treatment for chorea, under strict supervision from his wife and his mother. Though Mr. M responded well with the medications, his was still constantly monitored and received optimum care by our community psychiatric team to ensure that he complied with the treatment plan.

## Conclusion

This report portrays the various neuropsychiatric facets of HD. Needless to say, HD presents as a challenge to every treating clinicians as the range of disorders are manifold. In this case, notable features are distressing schizophrenia-like symptoms, the challenge of titrating medications, relieving pain from unwanted

movements as well as tackling family related issues. This encounter is written succinctly with hopes that it may serve as one of the multiple references available in the treatment of neuropsychiatric manifestations of HD. As a final note, we opine that holistic multimodal management is the best approach to aid patients suffering from psychiatric symptoms as a sequela of HD. Although the final morbid outcome of HD is inevitable, the journey of a patient through the course of illness need not be traversed with such a condescending note. Prompt treatment with empowered family support may greatly improve the quality of life of our patients. And the task of us psychiatrist, is to guide them there.

## Acknowledgement

Authors would like to express their gratitude to the multidisciplinary teams involved in the care of this patient, making the treatment process a seamless endeavor.

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**ISSN: 2574-1241**

DOI: 10.26717/BJSTR.2022.45.007150

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