

Catch Smells That Aren't There: A Case Report

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Citation: Apa F, Özdel İO (2018) Catch Smells That Aren't There: A Case Report. J Psychol Brain Stud. Vol.2 No.2:13

Abstract

This case report is related from a 54 years old woman who presented with a 1,5-month history of unpleasant smell senses. According to her complaints, her life has been changed due to unpleasant smells. In the examination of mental condition, her attitude has been partial harmonious, speech has been preservative, the affect has been apathetic and irritable, attention deficit, she has had hallucinations on smell milk and animal, had thoughts on self-harming due to her smell, marching ataxia and coreiform movements have been present.

The patient's family revealed that she has had a previous diagnosis of Huntington's disease; there has been also a family history. The patient has been admitted to the psychiatric inpatient unit. After 25 days of hospitalization, the amount of hallucinations has decreased considerably. Treatment has been regulated and as pimozide 2 mg, sertraline 50 mg and olanzapine 15 mg.

Keywords: Huntington's disease; Olfactory hallucination; Mild cognitive impairment; Psychotic symptom

Received: October 16, 2018; **Accepted:** October 30, 2018; **Published:** November 05, 2018

Introduction

Huntington's disease (HD) is an autosomal dominant, inherited, neuropsychiatric disorder that causes progressive motor, cognitive and behavioral symptoms [1].

HD is more common in people of European descent; the frequency in the community is around 10 to 15 in 100,000 [2]. The risk for men and women is the same. The average age of diagnosis is 30-50 years. It is rare to start before the age of 20 or after the age of 65 [3]. It is characterized by involuntary dance-like movements in the first stages chorea, akinesia and dystonia in later stages. Other manifestations include psychiatric changes and cognitive impairment [4]. Frequently reported neuropsychiatric symptoms are depressive mood, anxiety, nervousness and apathy, with a prevalence ranging from 33% to 76% [5,6]. Obsessive-compulsive symptoms and psychosis are less common, with prevalence ranging from 10% to 52% and from 3% to 11%, respectively [7]. In a study conducted, it was determined that the frequency of obsessive or compulsive symptoms was 27%, and depression, suicidal thoughts, aggression or delusions were reported more frequently in these patients [8].

Increase in the number of cytosine-adenine-guanine (CAG) trinucleotide repeats in the first exon of the Huntington's Disease gene (IT-15), which is located on the short arm of the 4th chromosome of the molecular basis of the disease [9,10].

The most striking neuropathology in HD is cerebral cortex steam laminar thinning and loss of white matter, as well as the disappearance of medium spiny neurons (MSN) in the striatum. According to a recent general opinion, because the earliest pathologic changes in Huntington's disease are located in the connective parts of the striatum, it is thought that first affected behavior is cognitive and psychiatric rather than motor signs. In fact, the observation of psychiatric symptoms in HD has shown that depression, apathy, aggression and disinhibition are common and suicide rate are more than four times the general population [11-13].

The diagnosis of Huntington's disease is made by family history of Huntington's disease, motor signs and genetic tests [7].

The pathogenetic mechanisms of Huntington's protein normal function and mutant Huntington protein are not clear today. Current drug treatment options have no effect on disease progression [1]. Training and symptomatic treatments are

effective tools for clinicians and families affected by HD, although there is no current method to change the course of this devastating disease [14].

Tetrabenazine is one of the better-worked and more effective agents to reduce chorea, despite the risk of potentially serious side effects. Newer antipsychotic agents, such as olanzapine and aripiprazole, which are more favorable for treatment of chorea and psychosis than those of old antipsychotics, may have adequate efficacy [14].

In this case, it is presented that a patient with a diagnosis of psychotic disorder due to Huntington's disease. This case report of a female patient with genetically proven HD is of particular interest.

Significance of the study is that the case which is shown that the patients could only have olfactory hallucinations as psychotic symptoms.

Case Report

N.A. 54 years old, married, primary school graduate, a woman who is not working anywhere and lives with her family. According to the story taken from her husband and children; the patient was diagnosed with HD in the other hospital's neurology clinic where they were admitted with complaints of fatigue, weight loss, gait disturbance, falls and sleep problems. In the genetic analysis, the CAG trinucleotide repeat was identified as 42 in the 1st exon of the *IT15* gene. She has been using venlafaxine 75 mg/g, olanzapine 2.5 mg/g for 6 years, and no additional disease. During the interview with the patient's relatives revealed that the last complaints of her were found on the ground 1.5 months before, that she smelled unpleasant smells, wanted to change their clothes due to smells, wanted no guest to her home because of their bad smells, wanted to throw away or burn clothes, carpets and blankets, wanted to bath 3-4 times a day and were insistent on relatives on same issue, had thoughts on self-harming due to her smell. It was expressed that she started to move away from society and did not want to coexist with the people. It was stated that there was not a period like that in her life at any time until 1.5 months ago.

In the examination of mental condition, self-care was adequate, attitude was partial harmonious, speech was preservative, the affect was anxious and irritable, attention deficit, the flow of thoughts were slow down, she had hallucinations on smell in the form of disturbing milk and animal, marching ataxia and coreiform movements were present. It was ascertained that in her family history, her grandfather, her grandfather's brother, her uncle (age 70 and diagnosed in 2011) and her uncle's daughter had been diagnosed HD and in addition to them, her grandmother had troubles on similar symptoms which not been diagnosed.

Due to her complaints, one month ago, after her apply to neurology clinic in İzmir, it was started pimozide 2 mg/g, it was stated that there was no benefit from medicine. After one week, because of no regression on complaints, it was ascertained that discontinue medication of pimozide, aripiprazole treatment was started and EEG and MRI imagining tests were done to investigate organic etiology after her application to psychiatry of other hospital. Due to no regression on her complaints, after the application of her to our hospital, the patient was hospitalized.

After her hospitalization to Pamukkale University Psychiatry Clinic, routine biochemistry, vitamin B 12, folate, thyroid function test (TFT), hemogram examinations were asked and ECG was taken. No pathological findings were found. The inpatient treatment was aripiprazole 10 mg/g, venlafaxine 75 mg/g, then it was gradually discontinued and sertraline 50 mg/g and olanzapine 5 mg/g were started. Patient Thematic Perception test (TPT) and Neuropsychological Test Battery (NTB) were performed.

It was observed that in her neuropsychological tests, in the sub-test of the numerical sequence related with the momentary attention, moderate deterioration on attention, moderate deterioration in recording in The Rey Auditory Verbal Learning Test (AVLT) and further deterioration on recovery (bring it back), medium loss in storage, the test performance indicating organization on Bender Visual-Motor Gestalt Coordination Test (BGT). The drawings made in this test suggest the existence of a significant deterioration in the areas of attention and memory, problems in judgment, impairment in planning, difficulty in making changes in attitude, tendency of concrete thinking, and interesting tendency to interrupt work and tasks. It was thought that the degradation in these tests would be related to demential process. Test performance in the presence of moderate corruption in the abstraction area was achieved. She had a slight difficulty in the judicial field.

The content at TPT suggested the presence of perceptual disorder, depressive traits, low coping skills, regressive attitude and attenuation in reality testing. The individual, in connection with possibly psychotic content, has included narratives that are incompatible with the content on some cards.

The patient was consulted on neurology. lectroencephalography (EEG) and contrast enhanced cranial Magnetic Resonance Imaging (MRI), MR angiography, perfusion MR for temporal lobe epilepsy was asked. MRI was usual and EEG was reported on borderline, after 10 days, the recurrent EEG was reported as normal.

It was observed that the patient, who had been intensely self-inflicted with bad smell spreading around, was trying to stay away from other people during her hospitalization and had bathing frequently, she often needed to confirm the smells she received and it was demanded that the goods in the room be changed due to bad smells by her. The dose of olanzapine was increased gradually to 20 mg, pimozide 2 mg was added to treatment.

Genetic consultation was requested for the confirmation of the Huntington diagnosis and genetic analysis repetition was requested. According to the genetic analysis on the 1st exon of the *IT15* gene it was designated that the resulting trinucleotide (CAG) repetition was 44.

After 25 days of hospitalization, the amount of hallucinations decreased considerably. Treatment at discharge was regulated as sertraline 50, pimozide 2, olanzapine 15 mg. After being discharged from the hospital, after 10 days of well-being, it was learned that when the family went to the village, she felt animal odor again, she bathed frequently and washed clothes frequently and bagged their clothes to thrown away. Treatment has been regulated and continued as pimozide 2 mg/g, sertraline 50 mg/g and olanzapine 15 mg/g.

Discussion

Huntington's disease is characterized by a triad of cognitive, motor and psychopathological disorders.

In recent years, in the knowledge of many different types of dementia have been increase and correspondingly, the possibility of detecting neurodegenerative diseases far before the time of clinical presentation has arisen. On the Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5), mild cognitive impairment has been identified as a pioneer or prodromal stage in progressive brain disease. Mild cognitive impairment in Huntington's disease has been documented in at least 40% of people of carrying gene mutations and numerous studies have shown that cognitive retardation can be detected ten years before motor manifestations occur. There are many publications showing early cognitive decline in Huntington's disease and there are also signs of psychiatric symptom progression before the onset of motor symptoms. Earlier diagnosis may be possible for many people due to the earlier clinical practices of prodromal Huntington's disease by mental health professionals ten years ago differently from neurology professionals [15].

Our patient has also had a tendency to deterioration in memory, attention and planning and concrete thinking. It has been thought that she has been at the level of mild cognitive impairment.

The prevalence of depressive mood in Huntington's disease ranges from 33% to 69% [16]. The development of depressive symptoms in Huntington's disease may be a direct consequence of cerebral degeneration. In addition to this, depression can be a psychological response to being at risk for developing Huntington's disease, growth in an insecure and harmful environment and/ or a psychological reaction to being aware his/her illness [17].

In our case, our patient's complaints were in the way of leaving society, not wanting to be together with people, low-spiritedness, not enjoying life. She said that her thoughts about harming herself often come to mind because her complaints did not pass.

The relationship between impulse and aggression of irritability arising in HD has been shown. When it occurs, amygdala and medial orbitofrontal cortex dysfunction play a key role [18].

In a study conducted in 2015 with a cohort of 90 mutation carriers followed for 2 years, it was found that while the incidence of irritability was 23%, the initial irritability was maintained at 70% of the mutation carriers, the increase in irritability was strongly associated with the increase in apathy [19].

Apathy and irritability in Huntington's patients are seen at similar rates, about half of the patients [20]. Anterior cingulate cortex plays a role in motivation and social behavior in humans. In Huntington's disease, dysfunction in the subcortical components of the anterior cingulate gyrus often manifests itself as apathy [21,22]. Our patient was apathetic and irritable at first sight. According to her relatives, it was stated that this was the case for the last 1, 5 months, beginning with psychotic and depressive complaints.

The occurrence of obsessive and compulsive symptoms in Huntington's disease is of particular interest. Both HD and obsessive-compulsive disorder (OCD) share a similar neuropathology of the basal ganglia and orbito-frontostriatal circuits [7]. Obsessive-compulsive symptoms are 10-52% (22). In a study of 1642 HD cases reported in 2010, approximately four quarters of those with obsessive-compulsive symptoms (27.2%) have been notified receiving OCD therapy. In this study, it has been determined that individuals with comorbid HD and OCD have been economically worse, been elderly patients and had longer disease duration than those without obsessive-compulsive symptoms. It has been notified that in individuals with HD and OCD, psychiatric comorbidities such as depression, suicidal thoughts, aggression, delusions and hallucinations are dramatically higher [8].

Psychotic symptoms usually occur when the action statement is already clear. Previously, psychosis was often described as the main psychiatric feature of Huntington's disease and could be diagnosed later in the disease [23]. Huntington's disease patients were often misdiagnosed as dementia praecox or schizophrenia until the first half of the 20th century. At the present time, the prevalence of psychotic symptoms has been reported at a lower rate, ranging from 3% to 11%, probably due to the early and better recognition of the disease [24]. The underlying mechanisms of psychotic symptoms are unknown, but the same symptoms appear in schizophrenia and therefore genetic risk factors for schizophrenia may be associated with the development of psychosis in HD [25]. Psychotic symptoms are associated with HD, a consequence of a relative hyperdopaminergic condition [24].

In our case, our patient also had the delusion of thinking that the bad smell was spreading around and the odor hallucination. It is mentioned here, a rare case of Huntington's disease that has been diagnosed 5 years ago by genetic analysis and with added symptoms cognitive and neurological findings in her process and generated psychosis clinic, treatment process and HD in the literature and accompanying psychiatric cases. With the case reports in the process and the work to be done for treatment, it is thought that psychiatric symptoms in HD will provide more information about the clinic and its treatment. Our case is remarkable in terms of she has only had a psychotic symptom such as olfactory hallucination and suicidal ideation, accompanied by dysfunction and cognitive decline with movement disorder. It should not be forgotten that HD cases may rarely come with psychotic symptoms.

This case suggests that the patient rarely encountered in this way should be considered as an opportunity for education on the other hand, psychiatric assessment of this type of case should not be ignored as a component of the inspection of the Huntington's disease.

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