Hypersexual features in Huntington's disease

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ABSTRACT

We report the case of a 30-year-old woman with a rare presentation of early adulthood Huntington's disease (HD) with hypersexuality. It is not known if sexual dysfunction in HD patients is due to a specific brain lesion or adverse psychosocial factors associated with HD. Although there are no evidence-based treatment guidelines for hypersexuality in HD, our patient exhibited significant improvement with olanzapine and haloperidol.

Keywords: Huntington's disease, sexual dysfunction

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INTRODUCTION

Huntington's disease (HD) is an autosomal dominant, inherited, neuropsychiatric disease that manifests with progressive motor, cognitive and behavioural symptoms.[1] Irritability and sexual dysfunction are observed to be frequent behavioural symptoms in HD, which warrant specific therapeutic approach in addition to the general treatment for HD. Published literature regarding sexuality in HD indicates that up to 85% of men and up to 75% of women experience sexual problems. Most of these problems are predominantly hypoactive sexual disorders, but increased sexual interest and paraphilia have also been reported.

CASE REPORT

A 30-year-old Indian woman presented with a history of involuntary movement of all four limbs and face for one-and-a-half years. These movements were of gradual onset with a progressive course. She also had behavioural changes in the form of frequent outbursts of anger, loss of temper and insomnia. The abnormal movements were aggravated during outbursts of anger and disturbances in mood, but were absent during sleep. She reported no weakness in any of her limbs, but was unable to perform regular household activities properly. In addition to the above complaints, she also reported increased libido exhibited by an unusual increased interest in talks on sexual matters and excessive demand for sexual intercourse for the past four months. There was no history of persistent elevation of mood; hence, a diagnosis of mania could not be established in the patient. There was also no history of intake of drugs such as phenytoin, oral contraceptives, phenothiazines, haloperidol, L-dopa, lithium, isoniazide, amphetamines and tricyclic antidepressants. Chest or joint pain, breathlessness and all symptoms that may suggest Sydenham's chorea were absent.

The patient's family history revealed that her father had similar abnormal movements with the onset at the age of 40 years. He died at the age of 55 years. Her two brothers and three children were symptom-free. Her general physical examination was non-contributory, except for mild pallor. She was normotensive and her cardiovascular system, abdomen and respiratory system were also normal. Examination of the central nervous system revealed an intact memory and normal orientation in time, space and person. Her score in the Mini-Mental State Examination was 28/30. Neurological examination showed generalised choreiform movements involving the face and limbs, that were more apparent when walking, motor impersistence (the patient could not fix her gaze at one point for more than 30 seconds or protrude her tongue for more than 30 seconds), slow and dysmetric saccadic eye movements and choreiform gait. However, deep tendon reflexes in all four limbs were normal, and the plantars were bilaterally flexor. The sensory and cerebellar systems were normal, and her bladder and bowel were intact.

The Mental State Examination revealed an adult woman of average built, with satisfactory personal hygiene and grooming. She appeared disinhibited in behaviour, as exhibited by her frequent attempts to hold the examiner's hand and to position herself as close to the examiner's seat as possible. This occurred even though she was reprimanded for such behaviour on several occasions. The examiner was able to establish rapport with the patient. Her speech was normal and her affect was irritable. Her psychomotor activity was increased, as evidenced by abnormal involuntary choreiform movements of all limbs. Her flow of thoughts was within normal limits; there was no evidence of either formal thought disorder or delusional beliefs and no discernable

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perceptual abnormality. Her cognitive functions were normal, but her judgement and insight were impaired.

Investigations revealed a haemoglobin level of 8.9 gm%. Peripheral blood smear revealed normocytic, hypochromic cells without acanthocytes, suggesting anaemia of chronic disease (HD). Erythrocyte sedimentation rate was 15 mm in the first hour; rheumatoid factor and lupus erythematosus cells were negative, and all biochemical parameters were within normal range. Chest radiograph, echocardiography and ultrasonography of the abdomen were also normal. Slit lamp examination of the cornea for Kayser Fleischer's ring was negative. Computed tomography of the head showed prominence of the lateral ventricles and flattening of the wall of the frontal horn, indicating atrophy of the caudate nucleus. Bicaudate distance between ventricles was also increased. Genetic analysis revealed 47 CAG (cytosine, adenine, guanine) repeat sequences. The patient was treated with haloperidol and had been on regular follow-up for one-and-a-half years before this study was written. There was partial response initially for 5–6 months, after which she maintained status quo. For the management of hypersexuality, olanzapine was added to the existing treatment regimen of haloperidol and was gradually titrated up to 20 mg/day, with dramatic improvement.

DISCUSSION

In 1872, George Huntington (1850–1916), a medical practitioner, made the first complete description of a disorder among the population of Long Island in New York State.[2] The disorder was then named 'Huntington's disease' after him. This disease is distinguished by the triad of autosomal dominant inheritance, choreoathetosis and dementia. Its cause has been linked to abnormal expansion in the length of a CAG triplet repeat sequence in a gene on chromosome 4p, now called the Huntington gene. HD has a population frequency of about 7–10 per 100,000 population and usually starts in adults 40–50 years of age. HD manifesting in early adulthood is rare.[3] Neuropathologically, the brain in HD may show cerebral atrophy ranging from mild to marked, with a corresponding reduction of about 30% in the total brain weight. On cut surfaces, the main abnormality is atrophy of the caudate nucleus (57%), putamen (64%) and globus pallidus. Neurotransmitters such as gamma aminobutyric acid, substance P, acetylcholine, enkephalins and cholecystokinin are decreased while the levels of hormones such as somatostatin, neurotensin and thyrotropin-releasing hormone are increased.[4,5]

The development of abnormal movements are usually the first signs of HD, and are most evident in the hands and face. Patients face difficulty in performing a sequence of hand movements. Movement disorder is usually slowly progressive, and may eventually become a disability. Attention, judgement, awareness and executive functions may be seriously deficient at an early stage, but memory is frequently not impaired until late in the disease. These abnormal movements, accompanied by disturbance in mood, particularly depression, are common. The high suicide rate in patients suffering from HD is noteworthy.[6] HD pursues a steadily progressive course, resulting in death at an average of 15–20 years after onset.

George Huntington described "two married men with HD who are constantly making love to some ladies, not seeming to be aware that there is any impropriety in it and they never let out an opportunity to flirt with a girl".[2] Altered sexual functions consisting of changes in sexual interests and behaviour are commonly observed in HD patients, which affects not only the patients but also their family members.[7,8] Studies have shown that spouses and children of HD patients suffer from various sociopsychiatric consequences.[9] Hypersexual behaviour is more commonly observed in men, with reported prevalence rates ranging from 3.9%–30%, as compared to the prevalence rates of 2.1%–25% in women. Dewhurst et al, in their study of the sociopsychiatric consequences of HD, reported that 30 out of 102 patients displayed abnormal sexual behaviour, 19 (18.6%) of whom showed hypersexuality.[9] Bolt found that only 20 out of 334 (6%) patients experienced increased libido or sexual deviation,[10] while Oliver reported 6% of the patients displaying similar behaviours.[11] Increased sexual interest has been reported in about 30% of HD males and almost 25% of HD females.[12] Of the 134 patients who attended an HD management clinic interviewed by Craufurd et al, using the Problem Behaviours Assessment for HD, uninhibited sexual behaviour was reported by 6% of the patients, while demanding or persistent sexual behaviour was described in 5%.[13] A behavioural profile characterised by irritability, mental inflexibility and obsessive-compulsive or perseverative behaviours has been found to be commonly associated with hypersexual behaviour.[13]

Our patient presented with behavioural disturbances such as irritability and frequent outbursts of anger along with hypersexual symptoms. The presentation of HD in early adulthood (as in our case) is rare, and that with hypersexuality is even rarer. Most of the available literature on HD reports that loss of libido and hyposexuality are much more common than

hypersexuality. This case illustrates the need to be aware of this known but unusual presentation of HD in routine clinical practice. In this case, the patient was put on a combination of olanzapine and haloperidol for behavioural disturbances and hypersexuality, and dramatic improvement was observed after one month of treatment.

The behavioural aspects of HD are more variable and more difficult to classify than the motor and cognitive changes observed, and perhaps due to this reason, they have received relatively little attention from researchers till date.[14] The only available data regarding the management of the behavioural disturbances, including hypersexuality, is from case reports. These behavioural disturbances have significant impact on patients and their family, on treatment compliance, and consequently have serious implications on the overall prognosis and quality of life in HD patients. It is thus imperative to ensure timely assessment and management of behavioural complaints and hypersexuality in HD patients.

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