HUNTINGTON’S DISEASE PRESENTING WITH SCHIZOPHRENIC SYMPTOMS: A CASE REPORT

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A case which initially presented with schizophrenic symptoms and was being managed as a patient of schizophrenic illness with tardive dyskinesia for 12 years, was on careful review of history and examination diagnosed as having Huntington's Disease. Clinical and theoretical aspects of Huntington's disease presenting with schizophrenic symptoms, and neurological deficits which are seen in schizophrenia are discussed.

Huntington’s Disease (H D) is a neuro-degenerative disorder with a prevalence rate of 5 to 10 per 100,000 worldwide. Inherited as an autosomal dominant illness it has a fully penetrant gene. In 1983, the first genetic marker for HD was found at G8 linkage, mapped to chromosome 4 which contains the gene for HD (Gusella et al., 1983).

Clinical expression of HD is usually delayed until mid-adulthood. Disordered movements, intellectual decline, dementia and emotional disturbances are the cardinal features of HD. Intellectual decline or dementia is invariably seen, the pattern is usually of a sub-cortical dementia (Fisher, 1983). Memory impairment is seen both in recent and remote memory, whereas immediate recall and attention are relatively spared (Boll et al., 1974). Cerebellar dysfunction, seizures and rapidly progressive dementia are common with the juvenile onset type than with the adult onset HD.

Insidious change in personality is a common prodrome of HD, with antisocial behaviour, somatic pre-occupation, conduct disorder and alcoholism (Caine and Shoulson, 1983) heralding the onset in about one-third of patients. Well defined affective and psychotic disturbances are found in about half of patients of HD (Folstein et al., 1983). Affective disorder appears in 30 - 40% of patients and often attains the severity of major depressive illness. Bipolar affective disorders have also been described by Folstein et al. (1983). Schizophrenia and atypical psychotic disorders are seen in nearly 10% of patients. Symptoms range from bizarre somatic to persecutory delusions (Mayeux, 1984). None appear to be drug related and most respond to appropriate psychotropic medication. Patients with HD are at high risk for suicide, partly reflecting progressive disability and adequate motor capacity (Mayeux, 1984). Death usually results in 10 - 25 years after the onset of illness.

We report here a case of HD presenting with schizophrenic symptoms.

CASE HISTORY

A 34 year old, married, sikh male presented in the psychiatry out-patient clinic with the presenting complaints of involuntary abnormal movements of all four limbs and trunk, muttering to himself, lack of interest in his immediate environment, unexplained aggressive outbursts, suspicious of his wife, and lack of sleep. The total duration of his illness by now was 12 years, with three previous hospitalisations and partial remissions in symptoms with treatment. He had been diagnosed a case of Chronic Schizophrenia (Paranoid type) with truncal tardive dyskinesia. He had been treated with various neuroleptics mainly with injectible Flusprilene 10 mg weekly dose. In the recent past he was prescribed chlorpromazine, trifluoperazine along with trihexyphenidyl and had also received ECT'S (12 in about 3 months period) as part of his treatment from a psychiatrist in private practice. At the time of present visit, he was off all medicines for about 5 days. There was a reported deterioration in his condition.

A work-up on the patient suggested the possibility of a neurological disorder along with prominent psychiatric disturbance. Review of his illness history of 12 years, starting when he was 22 years old, while in Canada revealed that the early features of his illness were a change in behaviour, excessive smoking of cigarettes along with excessive consumption of alcohol (he started consuming alcohol at 15 years of age), inappropriate aggressiveness with impulsive violent behaviour. Within a year of this he had first hospitalisation, following a period of continuous heavy alcohol consumption. He was treated for 2 months in a psychiatric clinic, and continued to be under psychiatric care even after his discharge. One year later, again, after continuous and excessive alcohol consumption, he was hospitalized for a month. On both occasions he was treated as a case of schizophrenic illness (paranoid type) and was also recorded to have developed truncal tardive dyskinesia during the second hospitalisation. Thereafter he was on continuous treatment with neuroleptics, and had one more hospitalisation...

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On examination, his general appearance showed neglect of personal hygiene with a poor nutritional status. The mental status examination in this withdrawn person was made difficult due to the inability to communicate normally with him. He showed inappropriate affect and muttering, experienced auditory hallucinations (accusatory voice of his wife and his in-laws), had experiences of being under the control of his wife, and delusional belief that his son was not his own child. He also showed clinically elicitable disorientation to time, impaired concentration and retention, and impaired recent memory. In addition, he harboured hostility towards his father, with inappropriate self-directed aggressive acts such as banging his head against the wall unmindful of the hurt it caused. He lacked initiative and interest in any thing. There was a total lack of insight into his illness. He had dyscalculia and inability to learn new information. He was interviewed by the clinical psychologist for detailed testing for organicity, which was not possible due to lack of co-operation from the patient. However he did show impairment in time-perception, the time having slowed down for him.

A detailed neurological examination was also carried out, which revealed generalised clumsiness, choreiform movements of all four limbs, abnormal swinging of his trunk (back & forth), and abnormal nodding of his head, finger and facial movements giving bizarre appearances. He had a dysarthria, a supranuclear gaze palsy, with marked slowing of saccadic eye movements and brisk oculocephalic response along with bilateral pyramidal and cerebellar signs and ataxic gait.

His mother had died in this hospital 10 years ago as a diagnosed case of Huntington's disorder of 2 years standing and his only sister aged 18 years underwent treatment for generalized seizures of unknown etiology for 14 years, and she died, during a status epilepticus 4 years earlier. The clinical picture of chorea, dementia, gross neurological deficits, schizophrenic symptoms, aberrant personality pattern, with a confirmed diagnosis of H D in mother and generalised seizures in sister, strongly supported the diagnosis of H D in this patient. The additional diagnosis of truncal tardive - dyskinesia was dropped as the involuntary choreiform movements were a part of H D.

He was put on neuroleptics as before, on Haloperidol increased to 30 mg/day. Trihexyphenidyl 4 mg/day was added after about 14 days when he developed tremors of hands. He responded to treatment with a marked decrease in the choreic movements and schizophrenic symptoms. He continued to maintain improvement without a complete recovery till a year and a half after his discharge, i.e. his last recorded visit to this hospital.

**DISCUSSION**

This case presenting with clinical pictures of H D. and also of schizophrenic illness is being reported here because of the diagnostic difficulties arising out of such presentations. Surveys have shown that between a third and two-thirds of cases of H D. are wrongly diagnosed initially (Bolt, 1970; Dewhurst et al., 1970). Psychiatric misdiagnoses are the most common, especially a label of schizophrenia or paranoid psychoses. With a diagnosis of schizophrenia, the choreiform involuntary movements are ascribed to 'Schizophrenic mannerisms', or as 'dyskinesia' induced by neuroleptic medication. An initial diagnosis of neurosis and affective psychosis are almost invariably corrected, whereas those of schizophrenia, neurological disorder, or dementia of other cause is less likely to be revised before the patient's death (Bolt, 1970). However, a correct diagnosis of H D. which is not a rare disease, is of great help for genetic counselling to the patient's family. As in this case, the diagnosis is always based on the clinical picture, and a positive history of H D. in the family (Braun, 1977).

The second question concerns the occurrence of schizophrenic symptoms in H D. The cause of personality changes and psychotic symptoms in H D. is still speculative. Dopamine metabolism is implicated in the pathogenesis of schizophrenia, but this issue has not been raised in H D. although post-mortem studies have revealed increased dopamine content (Spokes, 1981) and reduced levels of gamma-aminobutyric acid (GABA) in the basal ganglia and substantia nigra of brains from patients with H D. when compared to brains from neurologically normal persons. An imbalance between GABA and dopamine in such location could thus prove to be a key neuropharmacological
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Feature in H.D. at least as far as the movement disorder is concerned (Perry et al., 1973). It is also experienced that patients of H.D. have the most consistent antichoreic effects with dopamine-receptor blocking drugs or drugs that deplete pre-synaptic dopamine (Shoulson, 1979). While this explains the therapeutic response, it still does not clarify anymore the etiological heterogeneity of schizophrenic illness.

Finally, we need to know whether schizophrenia has any neurological abnormalities associated with it to enable us to differentiate between H.D. and schizophrenia in a better way. H.D. is almost always associated with dementia and varied neurological abnormalities, but this is still a point of much debate in schizophrenic illness. Patients diagnosed as having schizophrenia have been found to have increased prevalence of neurological abnormalities, indicating localised dysfunction of corticospinal tracts, basal ganglia and cerebellum, when compared with hospitalized normal volunteers (Woods et al., 1986). Cognitive impairment in schizophrenic patients such as under-estimate of their own ages of the present year and the duration of their hospital stay, i.e. temporal disorientation are associated with a significant increase in lateral ventricular size as seen on CT Scan. This is unrelated to past physical treatment, but is related to the presence of behavioural deterioration and abnormal involuntary movements often described as "tardive dyskinesia" and is not always attributed to neuroleptic drug medication (Crow et al., 1986). Goldberg et al. (1987) have speculated on the idea of presence of "dementia of the prefrontal type in schizophrenia", whereas changes located in the temporal lobe having a degree of selectivity for the left hemisphere have been reported by others (Crow et al., 1986).

Thus H.D. mis-diagnosed as schizophrenic illness is understandable, and more research into the biochemical and pathogenetic association between these two is needed not only to remove the diagnostic errors, but also to help identify specific diseases at an early stage.

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