CASE REPORT

WILSON'S DISEASE PRESENTING AS ISOLATED OBSESSIVE-COMPULSIVE DISORDER

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ABSTRACT

Wilson's disease (WD) is a genetic neurodegenerative disorder; it exhibits wide heterogeneity in symptoms and usually presents with liver disease and/ or neuropsychiatric manifestations. The common neurological manifestations observed are dysarthria, gait disturbance, dystonia, rigidity, tremor, dysphagia and chorea. The frequent psychiatric manifestations reported are personality and mood changes, depression, phobias, cognitive impairment, psychosis, anxiety, compulsive and impulsive behavior. Isolated obsessive-compulsive disorder (OCD) is a rare presentation of WD. Reported herein is a case of a 17-year-old boy with isolated OCD. He presented to the psychiatrist with symptoms of contamination obsessions and washing compulsions, along with compulsion of repeated feet tapping, and was treated with adequate doses of fluoxetine for 6 months but did not improve. Later on, he was diagnosed as a case of WD and showed improvement with chelating and behavior therapy. This implies the importance of the occurrence of isolated psychological symptoms in WD.

Key words: Obsessive-compulsive disorder, psychiatric manifestations, Wilson's disease

INTRODUCTION

Wilson's disease (WD) is a rare autosomal recessive genetic disorder. The genetic defect is localized to chromosome arm 13q and has been shown to affect the copper-transporting adenosine triphosphatase (ATPase) gene (*ATP7B*) in the liver.[1] The disease usually presents in the first or second decade with hepatic and/ or neuropsychiatric manifestations. Personality and mood changes, depression, altered sleep and appetite, inability to concentrate, lack of initiative, phobias, obsessive thoughts and compulsive behavior, panic attacks and lack of appropriate emotions are the common psychiatric manifestations, reported as presenting features and during treatment of WD.[2-5]

We are discussing a rare case that presented exclusively as OCD and diagnosed as WD. Probably; this presentation of WD is not published earlier in the literature.

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CASE REPORT

A 17-year-old boy presented to the psychiatrist with contamination obsessions and washing compulsions, along with repeated feet tapping, since 1 month. He started wearing only new undergarments and used to change comb and handkerchief daily. Repeated feet tapping, even during night hours, disturbed his sleep pattern also. He described these thoughts as being of his own and recognized them to be 'irrational.' Their compulsions relieved his anxiety to some extent but disturbed his studies. He did not have any history of involuntary movements, drooling of saliva, bulbar symptoms, gait difficulty, jaundice or any psychiatric illness. None of his siblings had similar illness.

He was diagnosed as a case of OCD, and the severity of symptoms and response to treatment were assessed on Yale-Brown Obsessive-Compulsive Scale (Y-BOCS) (severe, score - 30).[6] He received fluoxetine at an initial dose of 20 mg/day, which gradually increased to 60 mg/day over the next 4 weeks. After 6 months, when he did not show any improvement (Y-BOCS score - 32), CT scan of the brain was done, which revealed a hypodense lesion in right caudate; he was referred to us for ruling out the organic cause of his illness.

His general physical examination disclosed Kayser-Fleischer rings in both corneas, and per abdomen examination did not reveal organomegaly or free fluid. Mental state examination revealed an adolescent male of average built with satisfactory personal hygiene and grooming. He was cooperative, and rapport could be established. His speech was normal and affect was conveyed as euthymic. His psychomotor activity was increased, as evidenced by repetitive feet tapping and restlessness, which was explained by him as being a result of an irresistible, internal urge to perform the act. Assessment of his obsession and compulsion symptoms on Y-BOCS was severe (score - 32). His thought flow was within normal limits, and there was no evidence of either formal thought disorder or delusional beliefs. Cognitive functions were normal, and his judgment was intact and so was his insight. His verbal IQ on the Wechsler Adult Intelligence Scale-revised was 90.

On neurological examination, cranial nerves were normal. Motor system examination disclosed normal power, tone, reflexes, coordination and gait. There were no abnormal or involuntary movements. The sensations were also normal. He did not have cerebellar signs.

The biochemical investigations revealed normal hemogram, blood sugar levels, renal and thyroid profile. The liver enzyme levels – SGOT: 36 U/l (normal range=5-42 U/l); and SGPT: 33 U/l (normal range=5-45 U/l) – were normal. The serum copper level 0.66 µg/ml (normal range=0.9-11 µg/ml) and ceruloplasmin level 0.07 g/l (normal range=0.15-0.60 g/l) were low; and 24-hour urinary copper level 3143 µg/24 h (normal range=32-64 µg/24 h) was high. The electroencephalogram was normal. MRI of brain showed atrophy of right caudate nucleus, along with the altered signal intensity in right lentiform nucleus [Figure 1].

He was treated with D-Penicillamine (750 mg/

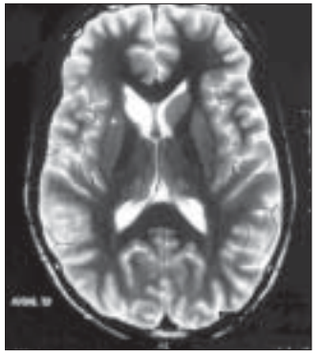

Figure 1: Brain MRI showing atrophy of right caudate nucleus, along with altered signal intensity in right lentiform nucleus

day), Zinc Acetate (150 mg/day) and cognitive behavioral therapy; and he did not experience any biochemical or clinical adverse reactions. When he showed significant improvement after 2 months of starting treatment (Y-BOCS score - 8), D-Penicillamine and fluoxetine were stopped. During 26 months of follow-up, the patient was asymptomatic on low copper diet and Zinc Acetate (150 mg/day), without any new psychiatric/ neurological symptoms.

DISCUSSION

Psychiatric symptoms are not rare in WD and have a variable incidence; about 20% of them precede hepatic and neurological dysfunction.[1-4] Personality changes such as irritability and low threshold to anger, depression and deteriorating performance were the most common. Less frequently, cognitive impairment, psychosis, anxiety, obsessive thoughts and compulsive behavior and panic attack were also seen. Rare presentations like mania,[5] schizophrenia, hypersomnolence and attention-deficit hyperactivity disorder have also been reported. Though OCD has been seen in association with many neurological illnesses,[7] isolated OCD without neurological and hepatic symptoms as presenting features of WD has never been reported.

It may be postulated that this association of WD and OCD may be either due to shared neuroanatomical or neuropsychological deficits or both. Limbic system is known to be involved in WD; and based on neuropsychological and functional imaging studies, similar areas have been shown to be involved in OCD.[8] Usually, bilateral involvement of basal ganglia occurs in WD; however, unilateral involvement of basal ganglia on imaging in our case was an uncommon finding.

Although effective treatments exist, OCD and related disorders are often underdiagnosed, undertreated and under-assessed.[6,9] Serotonin reuptake inhibitors and cognitive behavioral therapy represent the first line of treatment. Atypical neuroleptics like clozapine may be used for the psychotic and affective symptoms of WD because of their fewer extra-pyramidal side effects. In our case the initial poor response to fluoxetine may be due to the underlying disease, and the patient responded well after starting a specific chelating therapy. However, chelating agents as a treatment for OCD and related disorders have never been tried.

In conclusion, isolated OCD is a rare presentation of WD. Young patients of OCD andthose poorly responding to adequate psychiatric treatment should always be subjected to neurological examination to look for the organic cause, especially for WD. WD is a treatable disorder if diagnosed early, and the OCD symptoms in these patients can be managed with chelating and behavioral therapy.

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