CASE REPORT

WILSON’S DISEASE CAN PRESENT AS PARANOID SCHIZOPHRENIA AND MANIA: TWO CASE REPORTS

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Abstract

Objective: This case report highlights Wilson's disease, a rare genetic disorder involving the liver and brain presenting clinically with psychiatric symptoms as the first manifestation. Method: We present two cases of Wilson’s disease who had the typical symptoms of Schizophrenia and Bipolar mood disorder (mania) respectively. Results: Wilson disease first presentation of psychiatric diagnosis my obscure the diagnosis who later on turned out to be suffering from Wilson’s disease. Conclusion: Although such patients are more commonly seen in neurological and hepatological settings, mental health professionals must keep in mind a high level of suspicion, once first presentations may be of psychiatric nature. ASEAN Journal of Psychiatry, Vol.12 (2): XX XX.

Key words : Wilson’s disease, Paranoid Schizophrenia, mania

Introduction

Dr. Kinnier Wilson in 1912 first described Wilson’s disease in twelve patients as hepato lenticular degeneration [1]. It is an autosomal recessive disorder caused by mutation of ATP7B gene, which is a membrane bound copper transporting ATPase [2]. The three main neurologic problems are dystonia, incoordination and tremor [2]. Others include memory loss, migraine type headache, autonomic disturbance, seizures etc. [2]. A history of behavioral disturbances with onset five years before diagnosis of Wilson’s disease is present in half of patients with neurological disease [2]. Scheinberg and Sternlieb found that 10-25% of Wilson’s disease patients initially present with psychiatric symptoms which range from depression, mania, antisocial behavior to psychosis [3].

Case One

A 20 year-old unmarried Hindu, from rural background and lower socioeconomic class, education up to 10th standard presented with sudden jerky involuntary movement of head, trunk and all four limbs for the past six months and abnormal behaviour for the past one month. He was referred to Department of Psychiatry from Department of Neurology for exclusion of any component of conversion disorder. They diagnosed the case as Progressive myoclonic epilepsy which was not responding to antiepileptics. The attacks were present during sleep with reduced frequency. He also complained of infrequent hearing voices which were commanding and persecutory in nature and used to frighten him which suggests auditory hallucination. He also described few episodes of viewing “Maa Kali” (Goddess) coming and also...
multi-coloured halo coming from Goddess and entering his body which causes a burning sensation suggestive of visual hallucination, somatic passivity and somatic hallucination respectively. The mental status examination also suggested presence of thought broad casting and feature like syndrome of Fregoli (single person is coming to him in different disguise). The patient also had delusion of control where he attributed the involuntary movements to be controlled by external forces. Throughout the mental status examination, his affect was preserved. These kinds of symptoms were atypical for his culture as reported by his father. There was no past or family history of neurological or psychiatric illness. The neurological examination was within normal limits. Investigation revealed positive KF ring in cornea under slit lamp, low serum ceruloplasmin 0.13 gram/ L (reference range 0.2-0.6 gram/L), 24 hour urinary copper was 33 µgm (reference range 32- 64 µgm in 24 hours). The results of Liver function test, Ultrasonography of whole abdomen, Hb/TLC/DLC, Fasting sugar, Urea Creatinine, Na⁺, K⁺, routine and microscopical examination of urine, EEG, ECG, CT scan of brain were all within normal limits. The liver biopsy was not performed because of the invasiveness of the procedure and the facility of estimating copper in dry liver tissue is not available at our centre. A diagnosis of Wilson’s disease was made and he was given 300 mg/ day zinc sulphate and 10 mg per day olanzapine. He is now on regular follow up and is doing well.

Case Two

A 24 year-old unmarried Hindu male, education up to Master of Science from middle socio economic class and urban background presented with repeated attacks of fall for the past three months. The fall was associated with clenching of teeth, stiffening of hands and reduced consciousness sustaining for 5-10 minutes without any history of tongue or lip bite, frothing, incontinence or injury. There was no attack during sleep and no apparent life stressor. In one single incident while he was alone he fell into a shallow well near his house and was rescued by family members. He was found to be unconscious. He was admitted to Department of Medicine, Medical College, Kolkata and was given carbamazapine 1000mg/ day in divided doses. While admitted, he developed manic symptoms for which he was transferred to Psychiatry Department as seizure was controlled. The mania was characterized by delusion of grandiosity, mood congruent delusion of persecution, decreased need for sleep, increased talkativeness, elated and irritable mood. Considering age, seizure disorder and manic symptoms, he was screened for KF ring which came out to be positive. The serum ceruloplasmin level was 0.21 (reference range 0.2-0.6 gram/L), the 24 hour urinary copper was 248 µgm (reference range 32- 64 µgm in 24 hours). The results of Liver function test, Ultrasonography of whole abdomen, Hb/TLC/DLC, Fasting sugar, Urea Creatinine, Na⁺, K⁺, routine and microscopical examination of urine, EEG, ECG, CT scan of brain were all within normal limits. There was no past or family history of neurological or psychiatric illness. The neurological examination was within normal limits. Based on presence of KF ring and high 24 hour urinary copper level, a diagnosis of Wilson’s disease was made and he was given d- penicillamine 250 mg BID along with Quetiapine 400mg/day in divided doses and carbamazapine was continued as before. The patient is now doing well and is on our continued follow up.

Discussion

Although rare, the reported incidence of Wilson’s disease is 3-4 cases per 100,000 population [4]. This disease is diagnosed by the following findings: (a) Presence of Kayser-Fleischer (KF) ring (present in 99% cases having neurological symptoms but only 30-50% cases having only hepatic involvement), (b) Urine copper level > 1.6µmol (>100µg) in 24 hours, (c) low serum ceruloplasmin level (normal range 180-350 mg/L) which is found in 80% cases. The gold standard diagnosis is by liver biopsy with quantitative copper analysis (copper values > 3.1 µmol per gram of dry liver weight) [2]. Early detection of Wilson’s disease is critical to
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prevent catastrophic outcome by introduction of chelation therapy [5]. Young patients presenting with psychiatric symptoms along with movement disorder, seizure or conversion like symptoms should be screened for Wilson’s disease even if the symptom is typically suggestive of paranoid schizophrenia or mania. For this, strong clinical suspicion and close liaison between psychiatrist and neurologist is necessary.

References


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