CASE REPORT

Wilson’s Disease : A Case Report

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ABSTRACT

Wilson’s disease is a rare metabolic disorder involving copper metabolism may present with hepatic, neurological and psychiatric manifestations. We present a case of Wilson’s disease with behavioral symptoms, which responded to risperidone.

Key words: Wilson’s disease, psychiatric symptoms, risperidone

INTRODUCTION

Wilson disease is a rare but major metabolic disorder involving copper metabolism and is associated with abnormal liver functions. Hepato-lenticular degeneration (Wilson disease) occurs more frequently in male, is usually detected during adolescence, half of the patients have onset before 16 years of age. Forty percent patients first show hepatic dysfunction, 40% neurological symptoms and 20% with psychiatric disorder or behavioral disorder. It mostly presents with sudden and changeable mood swings; anger explosiveness which leads to impaired performance by eventual brain damage with memory loss if untreated. It may be confused with adolescent crisis or schizophrenia. (Lipkins, 1999)

We present a case report of a patient who initially presented with emotional disturbances with psychotic features, later diagnosed as Wilson disease, in whom behavioral symptoms responded to Risperidone.

CASE REPORT

Mr. A, 11 years old male child from non-consanguinous marriage coming from middle class, based at air force was referred from pediatric OPD. He denied any complaints except that he was teased by classmates as "mad". According to parents, the child cried easily, was restless and talked irrelevantly. Symptoms started after history of mild fever during school hours and had been given antipyretic drug.

Parents had noticed changes in his behaviour when he came from school one day prior to psychiatric consultation. He was crying and complaining about the teasing by peers as "mad" and wanted reassurance and asked the mother if he is mad. He talked irrelevantly that father is dead and so mother should not put bindi on her forehead (a sign of being married and husband being alive). He was tense, anxious and was not able to study. Though sleep was normal he did not take food well. Parents initially thought there was teasing to harass him as he was clever and topper in the class, and so he is disturbed and he will be soon all right.

Child was well oriented to time, place and person, had delusion of reference but no high mood or grandiosity at the time of the first consultation.

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He is living in a nuclear family with parents and younger sister. There was no family history of any psychiatric illness. No prenatal, perinatal, or postnatal complications. Developmental milestones were normal. Child began attending school at 6 years of age. Currently studying in 6th standard, he was very good in studies.

On examination he was conscious, cooperative oriented to time, place and person. Psychomotor agitation was present, child was not able to sit on chair for long time and looked anxious and asked the clinician and wanted confirmation that he is not mad. He reported his mood as euthymic but affect was restricted. There were no hallucinations.

Provisional Diagnosis was made

1. Anxiety disorder
2. Brief psychotic disorder possibly due to general medical condition or drug induced.

He was given alprazolam 0.25 mg three times a day and was asked to come for follow-up. After 7 days when he came for second visit, he improved in terms of crying spells but still other features were present. Alprazolam was stopped and Risperidone 1 mg/day with Trihexiphenidyl 2 mg/day were prescribed. On second follow-up after 7 days, he was not crying, did not complain of teasing, was taling relevantly, but was not taking interest in surroundings. On that day he complained of abdominal pain for which pediatric reference was made with reduction of dose of Risperidone to 0.5 mg per day.

After reducing the dose there was worsening of symptoms, the child started muttering and the dose was raised to 3 mg per day he improved within a week, the dose was reduced.

When he came within a few days for follow-up, there was worsening, mood symptoms like unusual cheerfulness and sleep disturbances were present. Mood stabilizer sodium valproate 400 mg per day was added.

After a week, he was hospitalized as he was not improving and was investigated fully with routine blood counts, fundus examination, Ultrasonography (USG) abdomen as had complained of abdominal pain.

During indoor treatment, he improved almost fully within two days after increasing doses of risperidone to 3 mg per day. The fundus examination was normal and USG
showed mild hepatomegaly. Child was transferred to pediatric indoor. Pediatrician advised examination of Kayser-Fleischer ring (KF ring) to evaluate for Wilson disease. Slit-lamp examination showed KF ring, hence serum Caeruloplasmin level was advised. Serum Caeruloplasmin was 6.2 ug/dl (normal - 25-63ug/dl) level and thereafter copper level and total urine copper level were advised.

Child was given D Penicillamine 250 mg once a day with zinc sulphate monosulphate 137.5 mg (equivalent to 50 mg of essential zinc) and pyridoxine 10 mg per day for 15 days.

As child improve behaviourally, valporate was tapered off within 3 days and Risperidone 3 mg per day was continued with trihexyphenidyl 2 mg per day and asked for follow up after 15 days.

When he came back, 24 hours urine copper level was 6 (average 53.9 for people age more than 10 years) and s. copper level was 203 (normal 75-150 |Xg%). Pediatrician increased the doees of D. Penicillamine to 250 mg t.i.d and Pyridoxide 40 mg / day with zinc 137.5 mg/day.

Child is maintaining behavioral improvement on 3 mg per day Risperidone and 2 mg trihexyphenidyl HCl per day.

**DISCUSSION**

This is an unusual presentation of Wilson's disease. Psychiatric presentation of Wilson's Disease is only in 20% patients. This patient had psychotic manifestations, which responded to Risperidone without any treatment emergent side effects. The patient needed total 3 mgs of Risperidone per day for control of psychiatric symptoms. Some authorities believe that antipsychotic use is inadvisable (Tu, 1987) because it may further compromise liver functions as well as added neurological side effects. There are case reports of neuroleptic malignant syndrome as well as acute, progressive akinetic-rigid syndrome (Chroni et al 2001). Penicillamine reverses both neurological as well as behavioral symptoms of Wilson's disease (Modai et al, 1985). There is also report of effectiveness of atypical antipsychotic drugs for example Clozapine in the management of Wilson's disease. (Krim et al 2001).

There is need to further study if chelating agents alone are adequate for treatment of behavioral symptoms of Wilson's disease or there is need of addition of antipsychotic medication. More studies are required to determine relative useful ness and treatment emergent side effects if any and optimal dose and duration of medication.

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