Wilson's Disease Presenting as Parkinsonism

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ABSTRACT
Wilson's disease (WD) is autosomal recessive disease. It has varied presentation so that WD need to consider in any patient with movement disorder under the age of 50 years. The three movement disorders associated with WD are dystonia, in coordination and tremors. We report a case of a young male who presented with pin rolling tremors of right hand since 3 months. After clinical examination and laboratory investigation, diagnosis of WD was confirmed. Patient was treated with zinc acetate and diet modification. Tremors responded to treatment. This case emphasises that tremors can be the main and presenting feature of WD.

Keywords: Wilson's Disease, movement disorder, tremors, pin rolling tremors, zinc acetate.

INTRODUCTION
WD is disorder of hepatocyte copper trafficking caused by impaired function of P-type adenosine triphosphatase (ATPase) encoded by ATP-7B gene located on chromosome 13q14[1]. This ATP-7B mutation manifesting with liver symptoms, neurological symptoms, psychiatric symptoms or combination of these. Neurological presentation include dystonia, incoordination and tremor. Dysarthria and dysphagia are also common[2]. Some patient can present with parkinsonian features.

CASE REPORT
A 26 years old right handed male with no significant family history presented with complaint of involuntary movement (tremors) of right hand with slowness of movements since 3 months. Tremors were present mainly at rest and suppressed on holding some object in right hand. Tremors had progressed during 3 months duration. Patient complained of difficulty in routine activities such as dressing and feeding. There was no involvement of any other limb. There was no history of convulsion, headache, fever, vomiting, visual complaints, motor weakness, sensory impairment or bowel and bladder complaint. There was also no history of jaundice, gastric complaint or bleeding tendencies. He didn’t give history of any addiction.

On examination, patient was conscious, cooperative and oriented. Cognitive functions were normal. There was no auditory or visual abnormality with normal fundus examination. The
cranial nerves examination was within normal. Kayser- Fleischer (KF) ring was evident in both eyes. There was pin rolling movement of right wrist and fingers. Tremors decreased on holding some object. Motor system examination revealed rigidity and bradykinesia in right upper limb with normal power and normal deep tendon reflexes. Motor examination of other limb was normal. Both plantars were flexors. Patient had normal gait. Other system examination showed no significant abnormality.

Routine investigation comprising of complete blood count and renal function test was within normal limits. Liver function test showed mild indirect bilirubinemia (direct billirubin 0.2 & indirect billirubin 1.2 mg/dl). Serum ceruloplasmin was reduced (6.44mg/dl). 24 hours urinary copper excretion was significantly raised (574.6 mg/dl). Slit lamp examination confirmed KF ring in both eyes. MRI brain showed bilaterally symmetrical hyperintensities in putamen, globus pallidi and caudate nucleus. Midbrain appears hyperintense on T2WI and FLAIR sparing the bilateral red nuclei giving the appearance of “Face of Giant Panda” sign.

Figure: MRI Brain of patient of Wilson Disease Showing “Face of Giant Panda Sign”

Electroencephalogram (EEG) showed normal study. Ultrasonography of abdomen did not revealed any liver abnormality. Patient was diagnosed as a case of Wilson disease with early parkinsonism and started on tablet zinc acetate 50 mg TDS and diet modification. The patient was discharged after initial observation. At 3 month follow up, patient tremors were reduced significantly.

**DISCUSSION**

WD is autosomal recessive disease which occurs as a result of disorder of copper metabolism. The incidence of WD is about 1 in 3,00,000[3]. WD commonly presents as hepatic symptoms (40%), neurologic symptoms (40%) or psychiatric symptoms (15%)[4]. Neurological presentation includes movement disorders (tremors, poor coordination, loss of fine motor control, chorea, choreoathetosis) or rigid dystonia (mask like facies, rigidity, gait disturbances, pseudobulbar involvement)[5].

Although the diagnosis of WD is made in the first decade of life in children, the neurological symptoms of WD are mostly observed in second decade[6,7]. Parkinsonism presenting typically with bradykinesia, cog wheel rigidity or imbalance has been reported in 19 to 62% of those with neurological WD[8]. Unilateral rest tremors and parkinsonism are rarely isolated clinical features of WD but like idiopathic Parkinson's disease, rigidity and tremors are typically asymmetrical[9].

The presence of extrapyramidal features with KF ring on routine examination help us suspecting WD in this case. WD should be kept in differential diagnosis of parkinsonism and slit lamp examination should be done. Copper deposits in limbic region of the cornea known as Kayser-Fleischer's ring are seen in nearly 100% of those with neurological WD and nearly 50% in hepatic and presymptomatic WD[10,11]. Increased 24 hour urinary copper with low ceruloplasmin in our patient confirmed the diagnosis further. The well recognised sign in MRI is the midbrain “Face of Giant Panda” sign, is due to high signal in tegmentum, normal signal in red nuclei and lateral portion of pars reticulata of substantia nigra and hypointensity of superior colliculus[12]. After establishment of diagnosis of WD, patient was treated with tablet zinc acetate. Zinc acetate
therapy has been used successfully as initial treatment for neurological WD and as maintenance therapy following course of decoppering\[13\]. Because of concern of neurological worsening in those treated with penicillamine, use of other agents have been recommended for initial treatment of neurological WD. Trientine is a chelator that like penicillamine promotes urinary copper excretion and causes neurological worsening in nearly 25%\[14\].

CONCLUSION
Parkinsonian features can be presenting feature of WD and is one of the treatable cause of movement disorder. Clinician should have high suspicion about this rare cause of movement disorder to diagnose and treat early to prevent further complications.

REFERENCES