A Case of Status Epilepticus: A Giant Panda Dropped the Hint

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ABSTRACT

Wilson's disease can manifest as neurological disorder without hepatic involvement. However, first presentation as status epilepticus is extremely rare. Herein, we report a case where a 21-year-old male presented with status epilepticus. Clinical background, biochemical tests and typical magnetic resonance imaging findings in the form of "Face of giant Panda sign" confirmed the diagnosis of Wilson's disease.

Key words: Giant panda; status epilepticus; Wilson's disease

Introduction

Wilson's disease (hepatolenticular degeneration), is an autosomal recessive hereditary disease of human copper metabolism, being characterized by excessive accumulation of copper in the body particularly the brain, liver, kidney and cornea. The clinical presentation varies but neuropsychiatric manifestations are common. Hepatic derangements like acute hepatitis, chronic hepatitis, cirrhosis of liver and acute fulminant hepatic failure can occur in early childhood. Neurological manifestations appear in the second decade and early symptoms are incoordination, tremor, dysarthria, dystonia, rigidity and difficulty with fine motor tasks. Seizures can appear during disease course or during treatment, but initial presentation as status epilepticus constitutes a distinctly unique scenario.[1]

Case Report

A 21-year-old male born to non-consanguineous parents, was rushed to the emergency department of SSKM hospitals with history of generalized tonic-clonic seizures (lasting more than 2 hours without regaining of consciousness in between. Seizures were associated with frothing from mouth, up rolling of eyeballs and urinary incontinence. There was no history of headache, vomiting, visual disturbances or focal deficits. He had no history of substance abuse or prior head trauma. He was managed as a case of status epilepticus in the Intensive Care Unit (ICU) with lorazepam (0.1 mg/kg intravenously) immediately on admission and phenytoin (20 mg/kg bolus intravenously followed by 6 mg/kg intravenously as maintenance dosages). Prior history of recent onset behavioral disturbances in the form of disinterest in the surroundings and decreased interaction with friends and relatives with occasional outburst of temper for last 15 days was present. But past history was unremarkable for liver dysfunction or seizure disorder. None of his siblings had similar illness. On examination, his vitals were stable. No jaundice or asterixis was noted. Abdominal examination did not reveal organomegaly or free-fluid. No signs of meningeal irritation was noted. Kayser-Fleischer rings were observed in both corneas, which were later confirmed by slit lamp examination. Comprehensive neurological examination, done after patient regained consciousness revealed generalized cogwheel rigidity in all 4 limbs, postural tremors of both upper limbs, hyperreflexia with normal muscle strength, sensory, cerebellar and cranial nerve (including fundi) examination. Routine laboratory tests including complete hemogram, electrolytes, liver and renal function tests were normal. Cerebrospinal fluid (CSF) Study was within normal limits. Axial T2-weighted magnetic resonance imaging (MRI) of the brain showed normal red nuclei and substantia nigra surrounded by hyperintense tegmentum, which gave rise to

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the characteristic 'face of the giant panda' sign in the midbrain, classically described in patients of Wilson's disease [Figure 1]. Analyzing the history and clinical findings and MRI findings, an atypical presentation of Wilson's disease was considered and appropriate tests ordered. Wilson's disease (WD) was finally confirmed by documentation of Kayser-Fleischer rings in both eyes, a low serum ceruloplasmin level (11.5 mg/dl reference range 20–60 mg/dl) and increased 24-hour urinary copper excretion (589 μg/24 hr reference range 25–50 μg/24 hr). Subsequently he was put on zinc sulphate 150 mg per day along with D-pencillamine 250 mg per day and was discharged in stable condition. Follow-up at 3 months showed signs of recovery in the patient.

Discussion

Epileptic seizures focal or generalized tonic clonic, are very uncommon as initial manifestations of Wilson's disease. Although, rare as first manifestations, seizure frequency is significantly increased in this condition. Dening *et al*., found a seizure prevalence of 6.2% in Wilson's disease, which is 10 times higher than in general population.[1] Seizures may be focal or generalized tonic clonic in nature. Similar to the index case, Shukla *et al*., reported a case of Wilson's disease where the patient presented with generalized tonic-clonic seizures. The underlying etiology was established by predominant extrapyramidal manifestations on clinical examination and supportive biochemistry and typical MRI findings.[2] Copper deposition in the brain may cause seizures by inhibition of membrane ATPase.[3] Neuronal loss, gliosis and cavitation may also be responsible for focal seizure activity. Again, hepatic encephalopathy can lead to seizures in patients with hepatic involvement. D-penicillamine, used as a chelator therapy in WD, can cause lowering of seizure threshold secondary to pyridoxine deficiency or by increased mobilization and subsequent cortical deposition of copper precipitating seizures.[4] Benbir *et al*., reported a case of Wilson's disease due to hypocupremia due to overzealous treatment by copper chelator D-penicillamine. The explanation offered was that copper is an integral component of key metallo-enzymes like cytochrome-c oxidase (role in electron transport and oxidative phosphorylation) that have a critical role in the structure and function of the nervous system.[5] Seizures can occur at any stage of the disease, but most commonly after beginning the treatment.

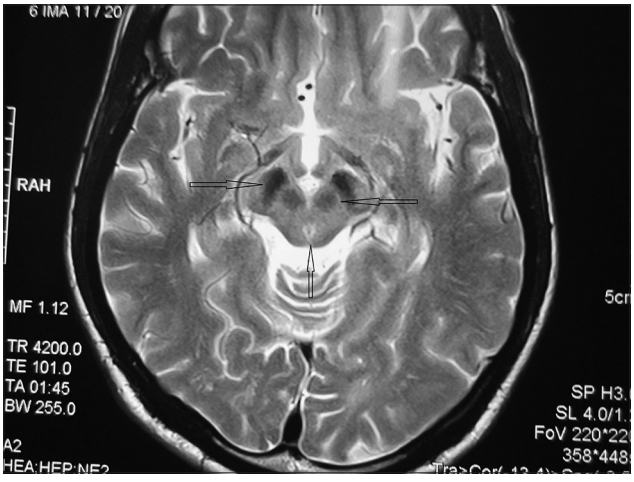

Figure 1: Showing Axial T2-weighted MRI of the brain at the level of the midbrain showing the characteristic 'face of the giant panda' sign, with normal red nuclei (eyes of giant panda) and pars reticulata of substantianigra (ears of giant panda) against a background of hyperintensity in the tegmentum, as well as hypointensity of the superior colliculi (chin of giant panda)

MRI changes are seen in virtually all neurologically symptomatic patients. MRI of the brain is not only a useful diagnostic modality in WD, but can also be used to assess disease severity and response to treatment. In patients with predominant hepatic involvement, T1 hyperintensities are noted in the globuspallidus, putamen and mesencephalon. In neurologically symptomatic patients, T2 hyperintensities are noted in the putamen, caudate nuclei, thalami, midbrain and pons.[6] Atrophy of the cerebrum, cerebellum and brainstem with ventricular dilatation may be seen in other cases. Kim *et al*. mentioned an interesting case in which a 28-year-old male of WD developed seizures due to cortical degeneration, which were initially misdiagnosed as dystonia. The patient also had a notable feature of extensive white matter lesions in MRI. The WD with extensive white matter lesions represents a rare neuropathological subgroup, the pathogenesis of which is not clearly determined. The abnormalities are mainly noted in the frontal lobe and most of these patients happen to be non-treated patients with a very severe form of the disease with particular poor prognosis.[7]

The 'face of the giant panda' sign, seen on axial T2-weighted images of the midbrain, is regarded as characteristic of WD. The sign, originally described by Hitoshi *et al*.is produced as a result of high signal intensity in the tegmentum with preserved normal signal intensity in the red nuclei (eyes of the panda) and lateral portion of the pars reticulata of the substantianigra (ears of the panda), and hypointensity of the superior colliculi (chin of the panda).[8] Exact pathogenesis of this finding is not known, but probably paramagnetic effects of the deposition of the heavy metals such as copper and iron may be responsible.[9] The 'face of the giant panda' sign is not commonly encountered; but if present, it is diagnostic, being the only MRI feature that distinguishes WD from other early onset of extrapyramidal disorders.[10]

Conclusion

Wilson disease is a very rare cause of status epilepticus. However, in a fitting clinical scenario, this uncommon presentation of Wilson disease must be borne in mind. MRI brain showing the typical "Face of giant Panda sign" clinches the diagnosis. Biochemical parameters showing copper overload lend supportive evidence.

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