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Case Report

A rare case of hepatolenticular degeneration with double panda sign on imaging

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ABSTRACT

Wilson's disease is an inborn error of copper metabolism that is characterized by deficiency of ceruloplasmin, the serum transport protein for copper. Copper is collected in the liver, and after hepatic binding sites are saturated, it is released. Systemic disease then develops and there is abnormal accumulation of copper in the brain, particularly in the putamen and globus pallidus. Presenting this case of a 32-year-old male patient who presented with peculiar features for Wilson's disease.

Keywords: Dermatomyositis, Classical, Wilson's disease

INTRODUCTION

Wilson disease is considered a rare type of autosomal recessive (AR) disorder in which abnormal copper accumulation in tissues, caused by mutations in ATP7B gene is seen. Investigations in patient reveals total body copper which is elevated and there is deposition of copper in Golgi complexes and mitochondria which results in oxidative damage through Fenton's process to the liver, brain, eyes, kidneys and skeletal system.[1] Imaging in patient with Wilson's disease shows a characteristic feature termed as the "face of the giant panda", which is a feature resulting from the sparing of the red nuclei within a diffuse midbrain hyperintensity. The age of presentation in Wilson's disease is from early childhood to 50 years.[2] Wilson's disease has a slight male predominance (52%).[3]

Our case shows this characteristic finding and in addition shows another rare finding on imaging.

CASE REPORT

A 32-year-old male presented to the OPD with rest tremors of right upper limb and left lower limb for a year. He had bradykinesia and slowness of activities of daily living. He was evaluated for young onset Parkinson's disease. KF rings were seen (Figure 1).

Serum ceruloplasmin was markedly decreased at 11 mg/dL (normal 18 to 55 mg/dL).

MRI revealed T2 flair hyperintensities in midbrain and pons (Figure 2) mimicking a giant panda and cub of a giant panda, collectively called as giant panda sign, suggestive of Wilson's disease. He was treated with dopaminergic medication and copper chelating agents.

Figure 1: KF rings.

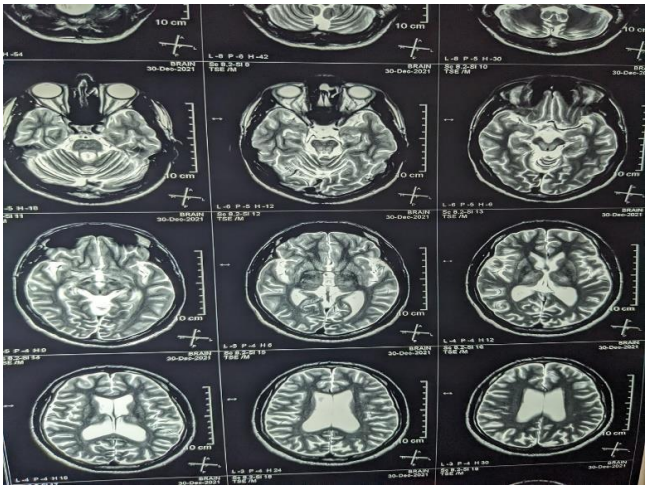

Figure 2: T2 flair hyperintensities in midbrain and pons.

DISCUSSION

An estimated worldwide prevalence of Wilson's disease is one case per 30,000 live births in most populations.[4] The midbrain "panda sign" on T2-weighted MRI has been previously described as preservation of normal signal intensity in the red nuclei and lateral portion of the pars reticulata of the substantia nigra, high signal in the tegmentum, and hypointensity of the superior colliculus.[5]

Another peculiar finding known as "face of the miniature panda" was also observed within the pontine tegmentum. It is due to relative hypointensity of the medial longitudinal fasciculi (MLF) and central tegmental tracts which will make the "eyes of the panda" combined with the hyperintensity of the aqueduct of sylvius opening into the fourth ventricle called as nose and mouth of the panda respectively which is bounded inferiorly by the superior medullary velum. The superior cerebellar peduncles form the panda's "cheeks."[6,7] The face of the giant panda sign" was first described by Hitoshi et al. On diffusion-weighted MR, recent lesions show a restricted diffusion which is likely due to cytotoxic edema.[8] On MR spectroscopy, there will be reduced N-acetyl aspartate/ creatinine ratio in the involved areas and increase in myoinositol/ creatinine ratio.[9] Clinical and MR imaging improvement occurred in 80% of patients with effective de-coppering treatment [10]

CONCLUSION

Edema, necrosis, and spongiform degeneration are the histopathological changes that are observed in Wilson's disease involving the brain. MRI not only provides biochemical information on heavy metal distribution in brain tissue but also gives an insight into the pathologic and anatomic correlates of clinical signs and symptoms in Wilson's disease. Interval changes seen on follow-up MR imaging have good correlation with clinical symptoms and can be useful in evaluating the clinical response to treatment of children with Wilson's disease.

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