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Neuroimage: "Hot Cross Bun" Sign (Reverse) in Case of Wilson Diseases

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Introduction

26 years old male presented with 10 months h/o tremulousness in all four limb initially, it was started with both upper limbs in form tremulousness during drinking water or eating food. Five months of an initial event he also developed tremulousness in both lower limbs more on walking than standing. He had altered speech in the form of mild slurring present for last two months. Neurological examination revealed mild dysarthria and presence of Kayser-Fleischer (K-F) ring in both eyes on the torch light and further confirmed by slit lamp examination (Figure 1).

Motor system exam revealed cogwheel rigidity with brisk reflexes with bilateral plantar mute. He had wing beating tremors with mild asymmetry (Right>left). MRI of the brain revealed, "Bisected pons" – changes like central pontine myelinolysis (CPM) (Figure2a), symmetrical hyper intensities in thalamus and asymmetrical subcortical white matter hyper intensities on FLAIR sequence (Figure 2b) and "Face of giant panda" in the midbrain (Figures 2c and 2d). MRI Brain also showed T2 hyper intensities in thalamus, midbrain involving the tegmentum pons (Figure 1e) along with a "Hot cross bun" sign (reverse) on DWI sequence (Figure 1f). In view of MRI features and K-F ring, patient was further investigated for Wilson's disease. Serum ceruloplasmin level was low .08 g/L (0.20-0.60 g/L). 24 h urinary copper excretion was elevated 414.58 mcg/day (<60 mcg/day). Thus he was diagnosed as a case of Wilson's disease, and treatment was started with Penicillamine therapy.

Neurological manifestations of Wilson's disease are related to abnormal deposition of copper in different part of brain tissues, especially in basal ganglia, thalamus, brainstem and subcortical white matter. Midbrain involvement occurs in 40% cases of brainstem with radiological signs such as "Face of giant panda sign" which is characterised by hyper intensities involving tegmentum with intact red nucleus and lateral part of substantial nigra and "CPM-like changes" with three distinct patterns for Wilson's disease: (1) the most common pattern of round shape, (2) bisected, and (3) trisected or "trident" sign occasionally reported in pons. However, all MRI features have rarely been reported in single patient but "Hot cross bun" sign (reverse) has never been reported in earlier literature of Wilson disease [1]. It has been described multisystem atrophy, spinocerebellar ataxia; cerebrotendinous xanthomatosis and pontine infarct (reverse) [2]. There was cruciform hyperintensity in the pons resulting from the degeneration of pontine neurons and loss of myelinated transverse fibres with preserved tegmentum and corticospinal tracts in "hot cross bun" sign. Here we are reporting a cruciform hypointensity in pons resulting in a reverse HCB sign in a case of Wilson's disease (Figure 2e).

Our case was interesting in displaying almost all the MRI characteristics with extended spectrum in form of reverse HCB sign in a case of Wilson disease.

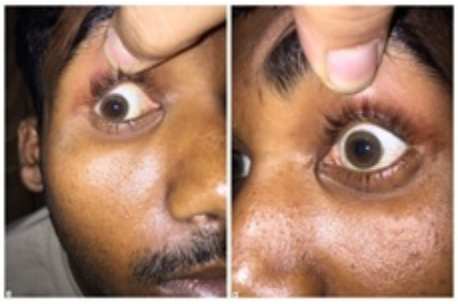

Figure 1: Revealed golden brown Kayser-Fleischer (K-F) ring in both eyes.

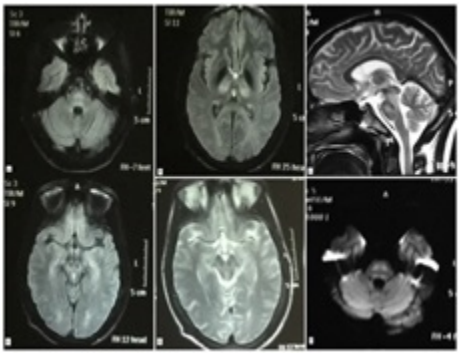

Figure 2: T2-weighted magnetic resonance imaging brain showed (a) a "Bisected Pons" appearance with relatively sparedpontine tissueas a dark horizontal line(b) bilateral symmetrical thalamic hyperintensities along with asymmetrical subcortical white matter hyperintensities. (c&d)"Face of giant panda" sign with tegmentum involvement appearing hyperintense with intact red nucleus and lateral portion of substantia nigra appearing hypointense (e) MRI Brain also showed T2 hyper intensities in thalamus, midbrain involving the tegmentum pons (f) along with a cruciform hypo intensity in pons resulting reverse "Hot cross bun" sign on DWI sequence.

References

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