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Acquired Hepatocerebral Degeneration with Ataxia and Symmetric Middle Cerebellar Peduncle T2 Hyperintensities

Ataksi ile Seyreden Edinilmiş Hepatoserebral Dejenerasyon ve Orta Serebellar Pedinkülde Simetrik T2 Hiperintensiteler

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Anahtar Kelimeler: Edinilmiş hepatoserebral dejenerasyon, kazanılmış karaciğer hastalığı, toksik madde birikimi

Dear editor,

A 55-year-old male presented with progressive difficulty in walking for the last 4 years, impaired speech and droopy eyelids. He had a history of diabetes mellitus, peripheral artery disease and 20 packs/year of smoking. The patient did not report any relevant family history. On neurological examination bilateral semi-ptosis, ataxic dysarthria, bilateral dysmetria and cerebellar ataxia were found. Laboratory tests showed an elevation of direct (1.08 mg/dl, laboratory reference range: 0.0-0.2 mg/dl) and indirect (2.54 mg/dl, laboratory reference range: 0.0-0.8 mg/dl) bilirubin levels and elevated liver function (alanine aminotransferase: 136 U/l, aspartate aminotransferase: 168 U/L, laboratory reference range: 0-50 U/l) tests. Repetitive nerve stimulation was normal, and acetylcholine receptor antibody was negative. In magnetic resonance imaging (MRI), bilateral symmetric hyperintensities were detected in middle cerebellar peduncles on T2 weighted imaging (Figure 1a, b). On T1 weighted imaging, bilateral hyperintensities were observed in globus pallidus and putamen (Figure 1c). Symmetric hyperintensities were also observed in cerebral peduncles on fluid attenuated inversion recovery (FLAIR) sequence (Figure 1d). Diffusion weighted images showed more obvious signal changes in the cerebral peduncles (Figure 2). Abdominal ultrasonography revealed cirrhotic appearance in the liver, therefore abdominal computed tomography (CT) was planned. In abdominal CT, the contours of the liver were lobulated, the left and caudate lobes were hypertrophic and the right lobe was atrophic. These findings

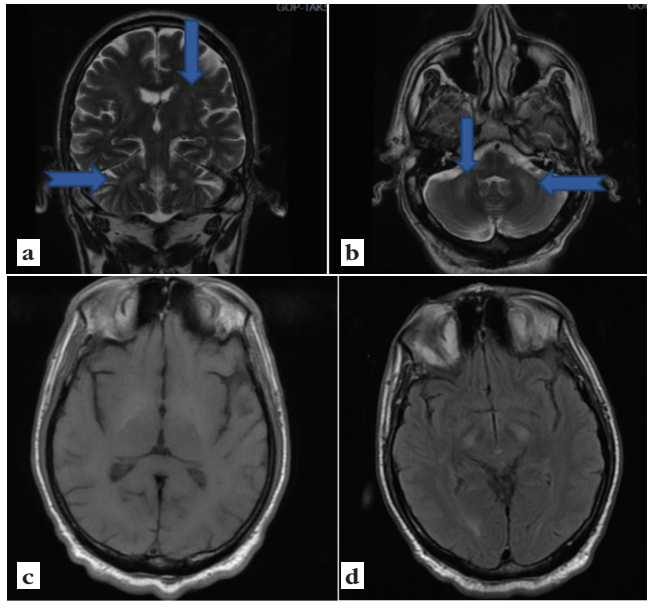

Figure 1. Coronal and axial T2 weighted images (a, b) show symmetric bilateral hyperintensities in middle cerebellar peduncles. On axial T1 weighted image (c), hyperintensities in globus pallidus and putamen. Axial FLAIR image (d), shows bilateral hyperintensities on cerebral peduncles
FLAIR: Fluid attenuated inversion recovery

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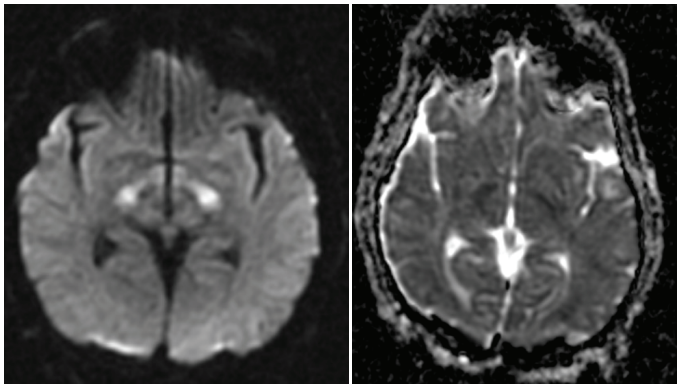

Figure 2. Hyperintensities in the cerebral peduncles appear more prominent on diffusion-weighted images

were evaluated in favor of chronic parenchymal liver disease. Serum ammonia level was high (146 µg/dl, laboratory reference range: 27-90 µg/dl). Autoimmune and infectious markers were negative. Ceruloplasmin and copper levels in 24-hour urine sample were normal. Liver damage was diagnosed as non-alcoholic fatty liver disease. With these results, the patient was diagnosed as having "ataxia-plus" subtype of acquired hepato-cerebral degeneration.

The prevalence of acquired hepato-cerebral degeneration in chronic liver disorders is estimated to be 1-2% (1). Clinical findings are heterogeneous, and movement disorders such as tremor, dysarthria, akinesia, myoclonus, hyperkinesia, gait disorders, ataxia, and parkinsonism may accompany (2). Neurological manifestations mostly tend to associate with copper of accumulation (3). Although this mechanism explains the hyperintensities on T1weighted series in the basal ganglia, it is thought that relatively less common T2 hyperintensities in middle cerebellar peduncles develop through a different mechanism. In addition to these neuroimaging findings, our patient had symmetric hyperintensities in cerebral peduncles on FLAIR and diffusion weighted images.

In conclusion, hepato-cerebral degeneration is a chronic hepatic encephalopathy characterized by parkinsonism and cerebellar findings, affecting approximately 1-2% of patients with liver cirrhosis. In our patient, hepato-cerebral degeneration was identified after cirrhosis was shown with laboratory and MRI findings.

Ethics

Informed Consent: Written consent was obtained.
Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: A.T.P., Y.Ş., Concept: A.T.P., Y.Ş., S.N.D., Design: A.T.P., Y.Ş., S.N.D., Data Collection or Processing: A.T.P., Analysis or Interpretation: A.T.P., Y.Ş., S.N.D., Literature Search: A.T.P., Y.Ş., Writing: A.T.P., Y.Ş.

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