

## Is central pontine myelinolysis a sign of pre-symptomatic neurologic form of Wilson disease?

*Santral pontin myelinosis pre-semptomatik nörolojik tutulumlu Wilson hastalığının bir işareti olabilir mi?*

To the Editor,

The diagnosis of Wilson disease (WD), a rare metabolic disorder, is based on clinical presentation of hepatic and/or neurologic involvement or routine screening due to the index case. Magnetic resonance imaging (MRI) was frequently normal in patients with hepatic form of WD (2). Central pontine myelinolysis (CPM) is usually an acquired disorder associated with rapid correction of hyponatremia, high amount of and chronic alcohol intake, and rarely, a neurologic form of WD (3). We describe a patient who had an image of CPM noted via MRI in spite of the absence of neuropsychiatric symptoms or findings of a neurologic form of WD.

A 26-year-old male was admitted to our hospital with upper gastrointestinal system bleeding and massive ascites. Physical examination revealed massive ascites, cachexia and palmar erythema. Upper gastrointestinal endoscopy showed lower esophageal varices and portal hypertensive gastropathy. Laboratory parameters were as follows: alanine aminotransferase (ALT) 30 IU/L, aspartate aminotransferase (AST) 34 IU/L, gamma glutamyl transpeptidase (GGT) 80 IU/L, total bilirubin 0.8 mg/dl, albumin 3.0 g/dl, and prothrombin time (PT) 12 sec. Viral markers for A-E were negative. The patient denied drinking alcohol. Family history for liver diseases or any other diseases leading to sudden death or neurological deterioration were all negative. His general status was good, and normal range of body stature was also noted. Serum ceruloplasmin level was 13.4 mg/dl, excretion of copper in urine was in normal range, Kayser-Fleischer ring examination by slit lamp was positive, and finally fine needle biopsy revealed excess copper accumulation in the liver. Cranial

MRI revealed CPM, atrophy in the cerebellum and cerebrum, hyperintensity on T2 images in the region of cerebral white matter, parietal and periventricular regions, optic radiation, and corticospinal tracts. However, there was no abnormality on neurological examination, with normal range of serum electrolytes.

Wilson disease (WD) is an autosomal recessive disease in which a defect in the copper excretion system leads to an accumulation in some tissues, especially in the liver and brain (4). Cranial MRI usually reveals some pathologic changes in patients with the neurologic form of WD, and the lesions in MRI mostly represent bilateral basal ganglionic lesions or diffuse atrophy of the brain. In contrast, MRI is frequently normal in patients with the pre-symptomatic neurologic form of WD, and moreover, CPM has not been determined in patients with the hepatic form of WD (2,3). The association between CPM and neurological form of WD was determined previously via histological and MRI examination, and the studies revealed a high rate of CPM in the neurologic form of WD (3,5-7). Sinha et al. (6) reported that all of the WD patients with CPM-like lesions had progressive neuropsychiatric symptoms. Prashant et al. (7) investigated the presence of some MRI findings among early-onset extrapyramidal disorders, and found a high percent (62.5%) of CPM-like abnormalities in WD patients.

In conclusion, CPM is not a rare finding in the neurologic form of WD, whereas it has not been reported before as a manifestation of the hepatic form of WD. Furthermore, it is not known whether or not the presence of CPM in the hepatic form of WD is a predictor of neurological WD.

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## Metastatic liver malignant melanoma of unknown origin

*Primeri bilinmeyen metastatik karaciger malign melanomu*

*To the Editor,*

Malignant melanomas usually originate from the skin (90%) (1). Cutaneous or subcutaneous nodules and lymph nodes are the most and abdominal viscera the least common sites of metastases of malignant melanomas. We discuss herein a patient with malignant melanoma in the liver of unknown origin.

A 60-year-old female admitted to the hospital with upper abdominal discomfort and a weight loss of 10 kg in the last two months. Abdominal ultrasound (USG) revealed liver enlargement (177 mm in diameter) with a mass lesion covering the right lobe and infiltrating the hilus of the left lobe. Laboratory analyses were in normal range. Serum alpha protein level was in normal range, and viral hepatitis serology was all negative except for antibody against hepatitis B antigen. Abdominal computed tomography (CT) showed multiple mass le-

sions in the liver, the largest in segment 5 with a diameter of 2 cm; some were observed to have central necrosis and enhanced with contrast agent in the arterial phase. The fine needle biopsy from the mass lesion in the liver revealed the diagnosis of malignant melanoma with EMA focally weak-positive, HMB45 focally strong-positive, and S100 diffuse-positive. Chromogranin was negative and mucin was also negative in neoplastic cells. This result was consistent with metastasis of malignant melanoma to the liver. There was no melanocytic origin or history of an excised melanocytic or pigmented lesion. Chest X-ray, ophthalmoscopy, examination of anogenital region, and upper and lower gastrointestinal endoscopy were all normal. Thereafter, the patient was diagnosed as stage 4 metastatic malignant melanoma of the liver of unknown origin according to the criteria of M.D. Anderson and not to be treated.

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