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CASE REPORT

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MRI findings in two presumed cases of central pontine myelinolysis associated with Wilson disease

Obraz MR dwóch przypadków centralnej mielinolizy mostu przypuszczalnie współistniejących z chorobą Wilsona

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Summary

MRI was carried out in two patients with Wilson disease after deterioration of their neurological status. In addition to changes in the cerebellum and lentiform nuclei, lesions were found in the central part of the pons in both of them. Pontine lesions were considered as central pontine myelinolysis due to the history of correction of hyponatremia in patient no. 1 and to the typical appearance in case 2.

key words: **Wilson disease • central pontine myelinolysis • magnetic resonance imaging**

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Background

Central pontine myelinolysis (CPM) is an acquired demyelinating disease considered as resulting from rapid correction of hyponatremia [1,2] in patients with other diseases. Although hyponatremia is thought to be the main reason of demyelination in CPM, it is also described in patients without hyponatremia [3,4].

Wilson disease (WD; hepatolenticular degeneration) is a disorder of copper metabolism, inherited as an autosomal recessive trait. The brain lesions are the most often present in the corpus striatum and putamen. Other gray and white matter structures, including brain stem, are less frequently involved [1]. Neurological and/or psychiatric symptoms are accompanied by acute or chronic liver disease.

We present two presumed cases of CPM accompanying Wilson disease. In both patients WD was diagnosed on the basis of clinical and laboratory findings.

Case reports

Case 1

A 38-year-old man presented with tremor, scanned speech and bilateral cerebellar syndrome. His neurological condition deteriorated during hospitalisation, 6 days after he had received medication due to hyponatremia.

The patient underwent MR examination of the brain at a 1.5 T unit. SE/T1-weighted, TSE/T2-weighted and FLAIR images were obtained in axial and sagittal planes. MRI revealed symmetric lesions, hypointense on T1-weighted images, hyperintense on T2- and FLAIR, located in the cerebellar hemispheres (Figure 1a), middle cerebellar peduncles and in the pons (Figure 1b). Pontine lesions presented as two hyperintense foci surrounded by the unchanged rim. Very subtle hyperintensities were also found in the thalami.

Case 2

A 36-year-old woman presented with spasticity, dysarthria, dysphagia and depression. She had chronic liver disease but

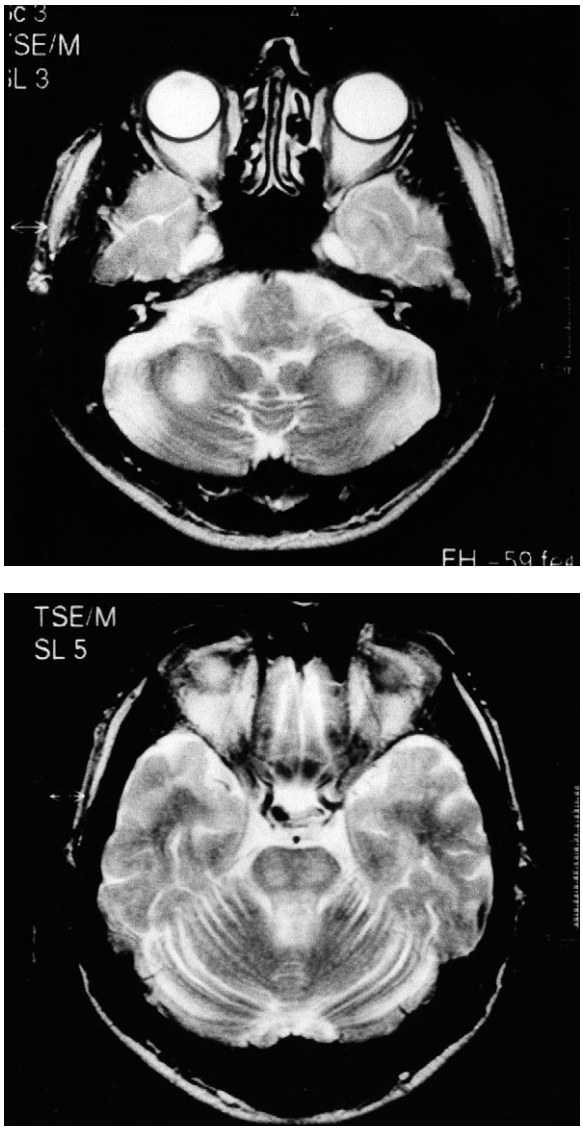

Figure 1. Case 1 – TSE/T2/ax: a) symmetrical lesions in the cerebellar hemispheres considered as resulting from Wilson disease; b) two lesions in the base of the pons, leaving the outer rim unaffected – CPM.

Rycina 1. Przypadek 1 – TSE/T2/ax: a) symetryczne ogniska w półkulach móżdżku, oceniane jako zmiany w przebiegu choroby Wilsona; b) dwa ogniska w moście, z pozostawieniem prawidłowego sygnału z jego obrzeża – centralna mielinoliza mostu

her serum natrium level was normal. She underwent cranial MRI after aggravation of dysarthria and dysphagia.

The patient was examined at a 0.5 T system. MRI revealed hypointense foci on T1-weighted images, hyperintense on PD and T2-weighted images, located in the lentiform nuclei (Figure 2a) and in the pons (Figure 2b). Central part of the pons was involved and the outer rim was normal.

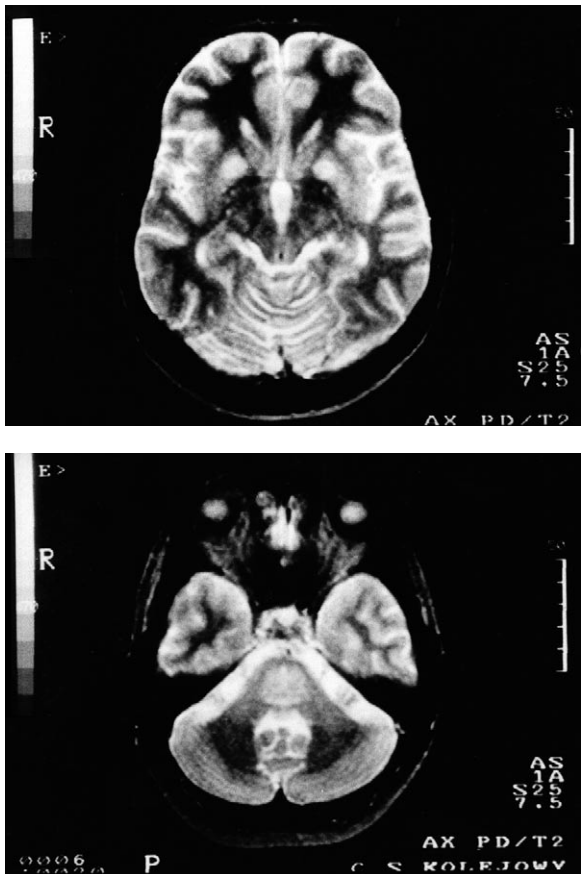

Figure 2. Case 2 – SE/T2/ax: a) symmetrical lesions in the lentiform nuclei resulting from Wilson disease; b) lesion in the central part of the pons, typical of CPM.

Rycina 2. Przypadek 2 – SE/T2/ax: a) symetryczne zmiany w jądrach soczewkowatych spowodowane przez chorobę Wilsona; b) ognisko w centralnej części mostu, typowe dla centralnej mielinolizy.

Discussion

The differentiation of neurological symptoms caused by WD and CPM is difficult or even impossible. In our patients WD has already been diagnosed; their neurological status suddenly aggravated, 7 and 4 days before MRI, respectively.

On pathological examination demyelination and loss of oligodendrocytes are found in the pontine tracts and nuclei in cases of CPM as well as astrocytosis and macrophages. As a result hyperintensity is observed on PD, T2-weighted and FLAIR images in the affected regions.

Neuronal loss and astrocytosis are also found in WD. Cavitation with lipophages and siderophages may also be present as well as depositions of copper. On MRI brain lesions usually show hypointensity on T1-weighted images and hyperintensity on PD, T2-weighted and FLAIR images although lesions that are hypointense on T2-weighted images are also described. They most likely represent iron and/or copper depositions.

The characteristic MRI pattern of CPM is a hyperintense lesion in the central part of the pons with an unaffected outer rim. Patient no. 2 showed this pattern of demyelination. In case 1 two separate foci of high signal intensity were found in the pons – this pattern has also been described [1].

MR findings of Wilson disease are reported not only in the basal ganglia but also in the brain stem. Abnormal signal intensity was found at first mainly in the pontine tegmentum [5] although on histopathologic examination degenerative changes resulting from WD have been described also in the base of the pons [6]. Further MR studies confirmed that white matter tracts are commonly abnormal in WD and lesions in the base of the pons are a part of the pontocerebellar and corticospinal tract involvement. These lesions, however, are usually small and visible all the way on the contiguous images, from the brain stem to the posterior limb of the internal capsule [7]. That was not the case in our material.

Demyelination consistent with CPM has been described in cases of WD as well [7–11]. In a report of 112 cases of CPM, WD accounted for as much as 14% of associated disorders [9]. There are very few imaging descriptions of CPM accompanying Wilson disease which makes this rare association very interesting for the investigators. The authors found only one report of 3 [4]. Imiya et al. postulate that the lesions in the base of the pons might have represented CPM in their patients although none of them had a history of rapid correction of hyponatremia whereas all of them had liver disease. It is suggested in the literature that iatrogenic correction of hyponatremia is the main reason of CPM but it is not the only one [9].

In our material patient no. 2 had liver disease resulting from WD and no history of hyponatremia and its correction. The pontine lesion was considered as CPM because of its typical MR appearance in a patient with severe liver disease and sudden deterioration of neurological condition. Patient no. 1 showed the less typical pattern of pontine lesion but his symptoms aggravated after the correction of hyponatremia.

A combination of MR findings, patients' histories and clinical data led us to the conclusion that in our two cases we deal with Wilson disease accompanied by central pontine myelinolysis. The analysis of MR images alone might be misleading since both signal characteristics and localisation of lesions in CPM/EPM and WD may be similar.

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