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CASE REPORT

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Olivopontocerebellar artrophy in MRI spectroscopy – case report

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Summary

Background: Olivopontocerebellar atrophy (OPCA) is an adult-onset disorder. It may occur in a sporadic or familial form.

Case report: We present the case of a 62-year-old man with clinical symptoms of progressive cerebellar ataxia.

Conclusions: Radiography presented atrophy of the pons and cerebellum. Clinically the syndrome results in progressive ataxia and bulbar dysfunction. Spectroscopy MR shows decrease in NAA/Cr ratio in cerebellum and pons – typical patterns of atrophy.

Key words: Olivopontocerebellar atrophy (OPCA) • H1MRS

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Background

Olivopontocerebellar atrophy (OPCA) occurs familially as a genetically conditioned Holmes form (autosomal dominant or recessive pattern of inheritance) as well as a sporadic form of Marie-Foix-Alajouanine (acquired form). According to latest views the olivopontocerebellar atrophy is classified to the group of spinocerebellar ataxias – large heterogenic group of progresive neurodegenerative diseases of the central nervous system. The sporadic forms are classified to the group of multisystem atrophies. The peak occurrence is in middle aged people.

The pathomorphologic image shows atrophies within the cortex and nuclei of the cerebellum, ganglia of the pons, olives, white matter and grey matter. Atrophies of frontal, parietal lobes, sometimes hyperintensive lesions on T2 weighted images (of demyelinational character) of the white matter of brain stem, cerebellum, less often – spinal cord, and atrophic lesions of basal nuclei are also observed. The histopathologic image of lesions within the CNS is related to the degenerating processes which lead to massive loss of neurons in the cerebellar granular layer and axons of the Purkini cells with following gliosis. Moreover, lesions are also found within the white matter – demyelination of spinocerebellar tracts. Occupancy of olivopontocerebellar system is secondary to the occupancy of the Purkini cells and ought to be associated with retrograde transsynaptic degeneration. In view of such hypothesis the 1HMRS examination, especially the evaluation of NAA and excitotoxic neurotransmitters (glutamates-Gln,Glu "hidden" in the Glx band) can provide important information.

Typical clinical symptoms of the Olivopontocerebellar syndrome include: ataxia, tremor, dysarthria, gait dysfunction, Parkinson symptoms. The CT and MR imaging techniques show mainly atrophy of the structures in posterior cranial fossa, i.e. cerebellum and brain stem (fig. 1a, 1b). Moreover, cortical atrophy of frontal and parietal lobes is also possible, as well as the less frequent demyelization in brain stem, cerebellum and spinal cord which are presented as hyperintensive lesions on T2 images and FLAIR sequence of MR examination.

Case report

A 62-year-old patient with ataxia progresing over the last 4–5 years and gait dysfunctions was referred to the Department of Imaging Diagnostics for definite diagnosis. The MR of cerebellum and previous tomographic examinations revealed

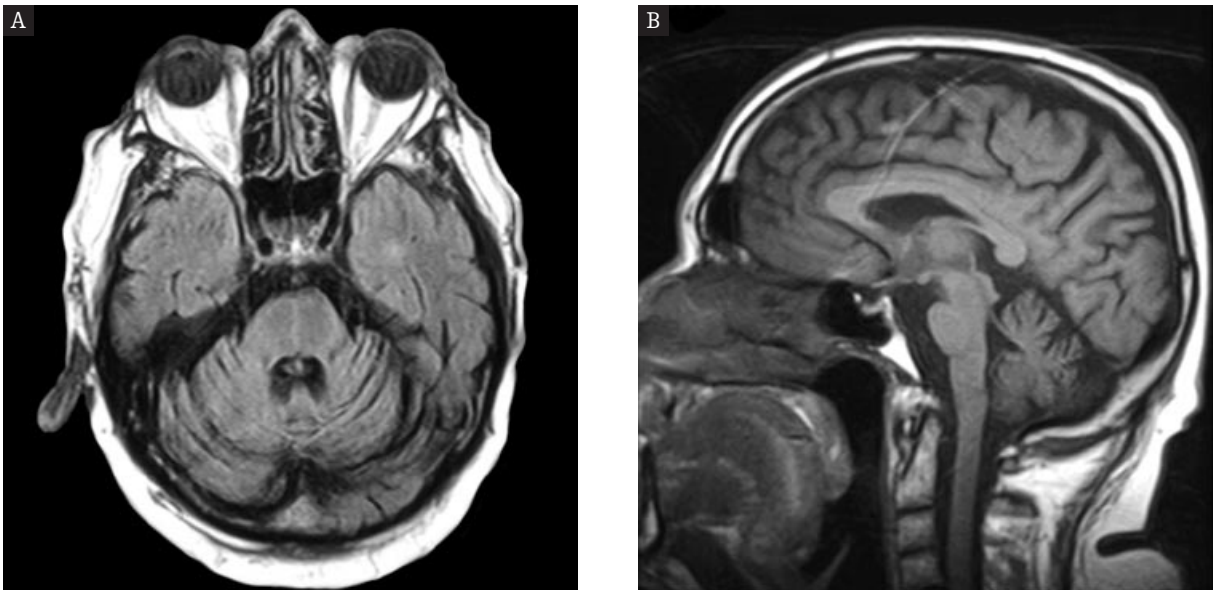

Figure 1. Axial T-1W and sagittal T-1W images show severe atrophy of the pons and cerebellum. The supratentorial structures show a mild degree of atrophy.

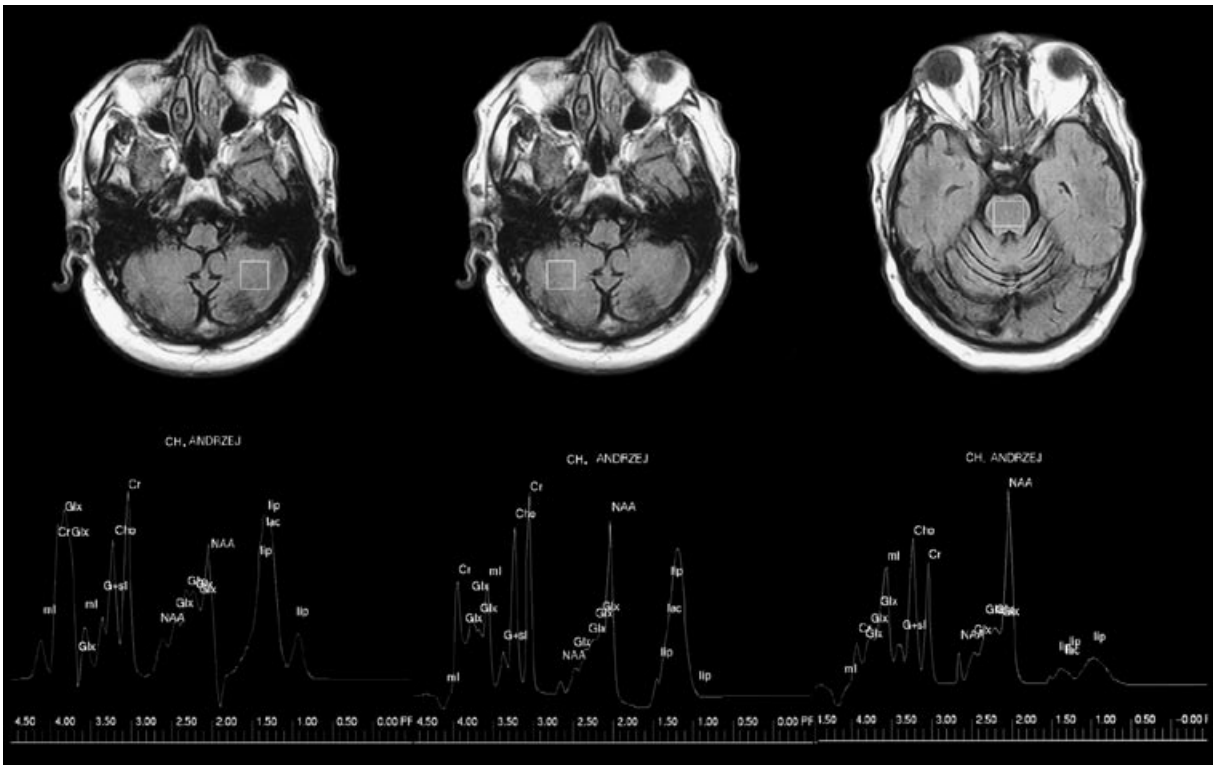

Figure 2. Single-voxel proton MR spectroscopy of cerebellar hemispheres and pons. Pontine and cerebellar NAA/Cr and Cho/Cr ratios are significantly decreased. Expression of Cho, ml and Lip. Lesions poorly marked within the stem.

massive symmetric cortical-subcortical atrophy of cerebellar hemispheres and vermis with enlarged sulci, adjacent subarachnoid cisterns and the 4th ventricle widened. Discrete signs of pons atrophy were also visible (fig. 1a, 1b).

The MR examination was extended with spectroscopic single voxel H1MRS. The H1MRS spectrum was registered from an 8 cm3 single voxel using PRESS sequence, with following parameters: TR=1.5 s, TE=35 ms. The examinations were performed with the use of head coil and the spectrum recordings were preceded with field homogeneity improvement within the whole head. Spectra were approximated in frequency domain with the producer – supplied software.

Measuring fields were placed in the pons and symmetrically in the cerebellar hemispheres. The results were

referred to the values of mean indicators (metabolite/Cr) obtained in two institutes which use identical spectrometers (control group comprised 12 patients with no clinical symptoms of cerebellar injury). It was arbitrarily decided that due to the lack of proper study group values higher or lower than the mean 15% will be threshold for the normal values.

The H1 MRS examination revealed a decrease of NAA-concentration, mainly in both cerebellar hemispheres where the NAA/Cr proportion was about twice lower than the standard, and a significant increase of the Cho/Cr indicator (fig. 2). In addition, proportion of these metabolites was also altered in the pons, but to a smaller extent. However, high concentration of Glx, Cho and Lip was observed (high lipid bands were also found in the control group). Morphologic image and chemical analysis were typical for atrophy corresponding to the olivopontocerebellar atrophy [4, 5, 6].

Conclusions

The olivopontocerebellar atrophy can be recognized based on clinical symptoms and characteristic morphologic image.

Extending the diagnostics with H1 MRS provides additional data which characterize metabolic lesions revealed in the morphologic examination [1, 2] and enables ruling out other neurodegenerative diseases with similar neuropathological-clinical image, such as the Friedrich diseases, SLA, or Parkinson.

Moreover, it allows monitoring the course of disease and reaction to the applied treatment. According to hitherto experiences the morphologic image of olivopontocerebellar syndrome corresponds to the level of clinical advancement of the disease. Precise analysis of metabolic profile (1HMRS) and clinical image (based on appropriate representative material) can have significant therapeutic implications.

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