Proton MR spectroscopy in gliomatosis cerebri

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J. Pyhtinen
Department of Diagnostic Radiology,
University of Oulu, Kajaanintie 50,
90220 Oulu, Finland

Abstract Two cases of gliomatosis cerebri are presented in which there was markedly decreased N-acetyl aspartate and an elevated lactate-lipid area in the MR proton spectra.

Key words Gliomatosis cerebri - Magnetic resonance imaging - Magnetic resonance spectroscopy

Introduction
Gliomatosis cerebri has been classified as a discrete entity but there are also opinions that it is only an infiltrative glioma [1, 2]. There are no earlier reports of MR spectroscopy in gliomatosis cerebri. Two cases are presented here.

Case reports
Case 1
A previously healthy 12-year-old girl gained 26 kg in weight in a year and had also been a little tired. One morning she vomited, had a convulsive attack and her level of consciousness became lowered. Mild strabismus was seen. Laboratory findings were normal.

CT revealed a large low-density tumour in the right temporal lobe. MRI showed diffuse infiltration of the right temporal and frontal lobes, extending from the hypothalamus to the left thalamus and basal ganglia. Mild contrast enhancement was seen in the right hypothalamus (Fig. 1 a). The cerebellar tonsils projected below the foramen magnum, and the fourth ventricle was very small. A diagnosis of gliomatosis cerebri was made, and this was confirmed by biopsy. Proton MR spectroscopy of the right temporal lobe demonstrated markedly reduced N-acetylaspartate (NAA) and an elevated lactate-lipids area, choline (Cho) and myoinositol (MI) being normal; NAA/Cho was 0.60 and Cho/Cr 0.92 (Fig. 1 b). The spectra were obtained at 1.5 T using the PRESS method and 8 cm³ voxel size.

Case 2
A previously healthy 75-year-old woman had right facial twitching and difficulties in speaking. She was sometimes a little disoriented, had small epileptic-like attacks and became euphoric. Her clinical state, normal laboratory findings and MRI, with follow-up, indicated a diagnosis of gliomatosis cerebri, confirmed by biopsy. Proton spectroscopy, using the same technique as in case 1, revealed markedly decreased NAA, elevated lactate-lipids area, MI at the upper limit of normal (within two SD), and normal Cho; NAA/Cho was 0.73 and Cho/Cr 0.86 (Fig. 2).

Discussion
Gliomatosis cerebri has been thought to be very rare, but there is accumulating evidence that this is not so [1–5]. This is probably due to better diagnosis, based on MRI; it may be that the incidence really has increased. In these cases the symptoms and signs were typical. As a rule, the symptoms and signs are fluctuating and mild, given the extensive lesions in the brain seen on MRI [3, 4, 6–8]. Obesity in the first case was probably due to hypothalamic-thalamic infiltration.

MRI typically shows large, often fairly symmetrical, paramedian lesions, with low signal on T1- and uniform high signal on T2-weighted images. Abnormalities are always present in the basal ganglia, thalamus and hy-
Fig. 1 a T2-weighted image shows a large, homogeneous high-signal, predominantly right-sided mass. Spectra of the right temporal lobe (b) revealed markedly decreased NAA and increased lipid-lactate (c). L lipid-lactate, NAA N-acetyl-aspartate, Cr creatine, Cho choline, mi myoinositol

Abnormal spectroscopic findings in these present cases were low NAA and elevated lactate-lipids area, Cho and MI being normal. In the second case MI was, however, at the upper limit of normal, which might be due to insufficient water suppression. The presence of lactate is not confirmed by turning the peak down with an echo time of 135 ms, but the lipid-lactate peak is too high. The spectra also contain noise. They spectra are not characteristic of high-grade gliomas, whose characteristic features are high Cho, low or absent NAA and creatine (Cr), along with lipid and/or lactate. NAA/Cho is lower and Cho/Cr higher in high-grade than in low-grade gliomas [9, 10]. The spectra are also not characteristic for low-grade gliomas: low NAA and creatine, high Cho and presence of lactate alone. The spectra had features of both low- and high-grade gliomas [9–10]. They were very similar in both cases and differed clearly from normal spectra (Fig. 3). These two cases do not, however, allow us to conclude whether gliomatosis cerebri has specific MR proton spectra. The diagnosis of gliomatosis cerebri is still difficult and needs biopsy confirmation, but can now be suspected with high probability on the clinical and MRI findings. More experience with MRS is needed to ascertain whether it is also characteristic.