Proton magnetic resonance spectroscopy of neurocytoma outside the ventricular region – case report and review of the literature

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Abstract Central neurocytoma is classically considered as an intraventricular benign tumour, largely based on data from small retrospective series and single case reports. We report on a 16-year-old girl who suffered from a large parietooccipital tumour that was diagnosed histologically as central neurocytoma. The features of CT, MRI and proton MR spectroscopy studies are discussed. This is the first report on spectroscopic findings in a case of extraventricular neurocytoma. As well as elevated choline (Cho), the tumour spectrum showed strongly decreased N-acetylaspartate (NAA). NAA is assumed to be produced in mature neurons, and we therefore expected to find high amounts of NAA in this well-differentiated tumour, which was histologically composed of mature neuronal tissue. This observation leads to the conclusion that even the highly differentiated cells of neurocytomases are too immature to produce NAA.

Keywords Extraventricular neurocytoma · Proton magnetic resonance spectroscopy · Magnetic resonance imaging · Computed tomography

Introduction

Central neurocytoma is a rare neuronal tumour of the central nervous system (CNS) accounting for 0.25% to 0.5% of all CNS tumours [1]. It typically appears in the ventricular and periventricular regions, close to the septum pellucidum and the foramen of Monro, but since the first description of this tumour by Hassoun in 1982 [2], neurocytomas were observed increasingly in extraventricular locations such as spinal cord, cerebellum and different lobes of cerebral hemispheres [3, 4, 5]. They occur typically in children and young adults, but also affect older patients; according to a recently published review of the literature [6], the age at clinical manifestation ranges from 8 to 67 years (mean 29 years). More than 70% of neurocytomas are diagnosed in patients between the ages of 20 and 40 years [1]. On the basis of histological examination, neurocytomas may resemble ependymomas, oligodendrogliomas, pilocytic astrocytomas and glioneuronal lesions such as DNTs [5, 7], but their morphological appearance on electron microscopy and their immunohistological features are clearly defined, with the immunostaining for synaptophysin as one of the hallmarks of this entity [6]. Characteristic imaging findings include sharp demarcation of the tumour, intratumoral calcifications, cysts, and contrast enhancement both on CT and MRI [7, 8]. Proton magnetic resonance spectroscopy (1H-MRS) has given new insights into metabolic and cellular characteristics of intracranial tumours, and it has been proven to be a useful tool for differentiating gliomas and other neoplastic and non-neoplastic intracranial mass lesions.
and rim-like calcification and intratumoral cysts. On T1-weighted MR images (Fig. 2a, b) the solid tumour part showed signal intensities equal to normal brain, with focal signal loss due to calcification as detected on CT. The solid tumour part revealed strong and somewhat inhomogeneous contrast enhancement. There was obvious peritumoral oedema and severe mass effect on the occipital horn of the right ventricle (Fig. 2c). On T2-weighted images, the tumour was isointense to grey matter, with small hyperintense foci in the cystic areas and hypointense foci in regions with calcifications (Fig. 2c, d).

Proton spectroscopy
Serial proton MR spectroscopy (single voxel point resolved spectroscopy (PRESS) with a TR/TE of 1,500/135 ms, VOI: 8 ml, n = 256 acquisitions), which was done in the solid tumour part and in the contralateral parietooccipital white matter (Fig. 3), revealed a nearly complete absence of NAA, a slight increase in creatine/ phosphocreatine (Cr) and a strong increase in choline (Cho). There were no resonances from lactate or lipids. Identification of resonances detected on 1H-spectra was based on findings reported in the literature: NAA, 2.0 ppm; Cr, 3.0 ppm; lactate (Lac), dephased peak at 1.3 ppm; lipid, 1.3 and 0.9 ppm [13, 14]. Spectral intensities were defined as the peak integrals determined from flat baselines on phase- and baseline-corrected spectra. Metabolite ratios were calculated as percent signal intensities relative to the signal intensities of the corresponding metabolites of contralateral white matter (SI in %) and as NAA to Cr and Cho to Cr ratios (Table 1). In addition, the data were compared with reference values of brain tumours previously studied by the authors (Table 1) [10, 15]. Due to absent NAA-peaks the preoperative differential diagnosis included primary or secondary extra-axial tumours rather than primary brain tumour.

Histological findings
Staining with hematoxylin-eosin (H&E) revealed a tumour extensively composed of uniform round cells with round or oval nuclei finely speckled with chromatin. Nucleoli were rarely observed. Clear mitotic activity could not be found. The cytoplasm was swollen and clear with a honeycomb appearance. Between the cells there was a conspicuous capillary network partly arranged in an arborizing pattern. In one area there was an extensive perivascular infiltration of lymphocytes. Additionally, we found some haemorrhage with haemosiderin and haematoxidin deposits. Furthermore, calcifications could be observed, predominantly in perivascular location in the centre of the tumour. Conspicuous nucleus-free areas of neuropil were not seen.

After incubation with antibodies against the neuronal markers synaptophysin and neuron-specific enolase, there was strong labelling in most of the tumour cells, but a lack of neurofilament and cytokeratin. After being immunostained with antibodies against the glial fibrillary acid protein (GFAP) only some astrocytes in the peripheral area of the tumour were marked, whereas the tumour cells did not express GFAP. The proliferation potential of the tumour, measured by use of MIB-1 antibody, revealed a KI67 labelling index below 1%.

On the basis of the H&E staining and the immunoreactions, a central neurocytoma, corresponding histologically to WHO grade II but with some unusual features, could be diagnosed.

Discussion
Although most neurocytomas have been observed in the lateral ventricles, cases at extraventricular sites have

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**Case report**

**History**

A 16-year-old Indian girl presented with episodes of cramping right frontal headaches that had started 2 months before admission. An EEG examination performed 1 week before admission showed slow waves in the right hemisphere. When she was seen in our department, neurological examination yielded normal findings. After 4 days of dexamethasone p.o., a right parietal craniotomy and transsulcal access to the lesion were performed using the assistance of an image-guided surgery system. Macroscopically complete extirpation of the lesion was achieved.

Even though the postoperative neurological status was normal, the postoperative course was complicated by a subgaleal CSF colletion, requiring lumbar CSF drainage for 11 days. The subsequent postoperative course was uneventful, and the girl was discharged on the 24th postoperative day without any neurological deficit.

**Neuroradiological findings**

Native CT (Fig. 1) showed a well-delineated hyperdense mass in the parietooccipital region of the right hemisphere with punctate

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**Fig. 1.** Native CT shows a well-delineated hyperdense right parietal mass with rim-like calcifications and intratumoral cysts [9, 10]. Because neurocytomas are tumours of neuronal origin, tumour spectra are expected clearly to demonstrate N-acetylaspartate (NAA), a neuronal marker usually lacking in extra-axial neoplasms [11]. Only two papers in the literature have focussed on 1H-MRS findings in central neurocytomas located in the ventricular region [11, 12]. To the best of our knowledge, this is the first report documenting proton spectroscopy findings in a so-called central neurocytoma, i.e. a neurocytoma outside the ventricular region.