MR Spectroscopic Profile of an Angiocentric Glioma

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Abstract. *A 2-year-old female with focal motor seizures was referred to our Center for brain MRI. Imaging revealed a non-enhancing, demarcated, cortically-based mass lesion in the right superior frontal gyrus and cingulate gyrus. Single voxel MR spectroscopy through the tumor showed markedly elevated myoinositol and/or glycine, mildly elevated choline, and moderately decreased N-acetyl aspartate. Histological assessment of the lesion following gross total resection was diagnostic of an angiocentric glioma. Herein, we present the first illustration of MR spectroscopic findings from a typical angiocentric glioma.*

An angiocentric glioma is a rare primary brain tumor first described in 2005, with histological features of both astrocytic and ependymal lineages (1). Because of its benign behavior and histologic features, it has been classified as low-grade (WHO grade I) (2). While CT and MR characteristics of typical angiocentric gliomas have been previously described, MR spectroscopic findings are sparse (3). The MR spectroscopy profile of a typical angiocentric glioma has not been previously illustrated. We present MRI and MRS features from a 2 year-old patient with a histologically-confirmed WHO grade I angiocentric glioma.

Case Report

An otherwise healthy 2-year-old female was referred to our imaging service for brain MR after a recent history of a focal motor seizure characterized by acute, rapid left arm movements. During the episodic motor dysfunction, her personality changed; she was unusually aggressive, but did not lose consciousness. Past medical history was unremarkable. She was the product of an uncomplicated pregnancy, born at-term. Her neurological examination was normal.

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Brain imaging was performed on a 1.5T MR (Signa HDxt Optima edition, General Electric, Milwaukee, WI, USA). Prescribed pulse sequences included: Sagittal T1WI SPGR (Spoiled Gradient Echo), axial T1WI, axial T2WI, axialT2 FLAIR (Fluid Attenuation Inversion Recovery), axial diffusion weighted images, and axial gradient echo images. Post-contrast axial and coronal T1WI were obtained after injection of 0.2 mmol/Kg IV gadolinium. Single voxel point resolved MR spectroscopy (PRESS) was also performed through the tumor and contralateral left superior frontal gyrus (TE 35, TR 1500); voxel sizes were approximately 2×2×2 cm.

Axial T2WI shows a relatively demarcated, expansile, hyperintense lesion centered in the right superior frontal gyrus extending posteriorly to the paracentral lobule (Figure 1). Facilitated diffusion is present on axial diffusion-weighted images (Figure 2). Gradient-echo images show no abnormal susceptibility to suggest blood products or mineralization. Axial T1WI through the mass before and after IV gadolinium administration demonstrate no abnormal lesional enhancement (Figure 3).

Single voxel PRESS (point resolved spectroscopy) performed with voxel of interest over the mass reveals a dominant metabolic peak at 3.55 ppm representing elevated myoinositol and/or glycine, mildly increased Cho:Cr, and moderately decreased NAA:Cr; no significant lactate is present (Figure 4). Single voxel MR spectroscopy (MRS) through the contralateral, structurally normal superior frontal gyrus shows normal metabolic ratios for comparison (Figure 5).

The patient underwent craniotomy and gross-total tumor resection. Histology was diagnostic of an angiocentric glioma (WHO grade I). Follow-up post-resection brain MR exams over the course of 3 years have shown no evidence of residual or recurrent neoplastic disease. The patient is now seizure-free without major neurological deficit apart from mild left lower extremity weakness and hypertonia.

Discussion

Angiocentric glioma is a recently described and classified low-grade (WHO I) primary brain tumor (1, 2). It is a rare glioma sub-type of debatable origin with histological characteristics of both astrocytomas and ependymomas (3, 4). Including our patient, only 70 cases have been reported in the

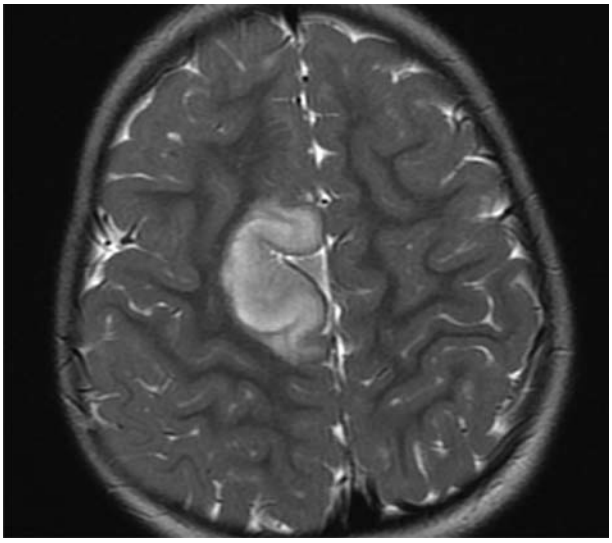

Figure 1. *Axial T2WI (repetition time msec/echo time msec, 3517/101) showing a relatively demarcated, hyperintense mass lesion in the right superior frontal gyrus extending posteriorly to the paracentral lobule.*

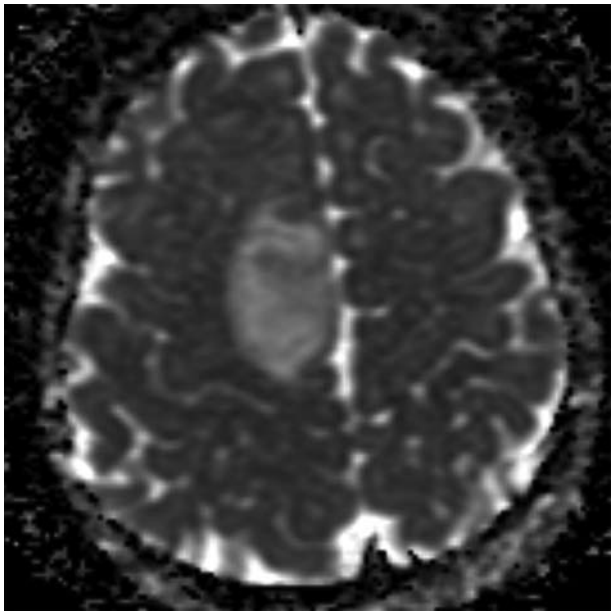

Figure 2. *Axial apparent diffusion coefficient image (ADC) (repetition time msec/echo time msec, 8875/113) showing hyperintense signal associated with the lesion in the right superior frontal gyrus compatible with facilitated diffusion.*

literature to date. It most often presents in childhood, mean age 16+/-14 years (range=2-70) years at the time of surgery (4). Clinically, nearly 90% of patients harboring an angiocentric glioma present with seizures. In keeping with its benign nature, gross total resection is curative (5, 6).

From an imaging perspective, angiocentric gliomas manifest as superficial cerebral hemispheric mass lesions, and thus, should appear in the differential diagnosis of a cortically based mass lesion in a pediatric patient with seizures. They tend to involve the cortex and subjacent white matter, expanding the corresponding gyrus. On CT, attenuation characteristics are variable, ranging from hypodense to hyperdense; calcifications are rare but may be present (7). On MR, lesional T2 prolongation with facilitated diffusion is the rule; lesional signal on T1WI may be hypointense or hyperintense (8, 9). Contrast enhancement is rare (4). Our case exhibits classic MR imaging features: T1 and T2 prolongation, facilitated diffusion, and no contrast enhancement. The focal motor seizures involving the left extremities that brought the patient to imaging and the post-operative left lower extremity deficit relate to the location of the original tumor: it involved the right paracentral lobule and supplemental motor area.

MR spectroscopy findings of histologically-typical angiocentric gliomas are scarcely mentioned in the literature. Grajkowska and colleagues reported MRS abnormalities in a 14 year-old with a parieto-occipital angiocentric glioma; they noted slightly elevated choline and slightly depressed NAA in the tumor (3). A mesial temporal angiocentric glioma in a 4-year-old was found to have a markedly reduced NAA to Cr ratio but no significant aberration of the choline to creatine ratio (10). MRS findings associated with an unusual high-grade angiocentric glioma have demonstrated markedly elevated choline, decreased NAA, and abnormal lactate (11). However, in that case the brain MR showed findings that would be atypical for a WHO grade I angiocentric glioma, namely, extensive contrast enhancement and restricted diffusion (11).

In our patient, MRS revealed a dominant metabolic peak at 3.55 ppm compatible with myoinositol and/or glycine. An osmolyte and marker for astrocytes, myoinositol can be elevated in a number of pathological brain processes and in some tumors, including astrocytomas (12). It metabolizes the membrane phospholipid phosphatidyl inositol; therefore, it may be increased with membrane break down or changes in metabolism (12). Short echo-time MRS is required to assess myoinositol because it has a relatively short T2 value. In neonates, it is one of the dominant metabolic peaks identified on short echo time MRS; the concentration of myoiniositol declines quickly within the first few years of life. At short echo times, it overlaps and is inseparable from glycine at 3.55 ppm. Intermediate to long echo times are necessary to distinguish myoinositol from glycine as glycine has a longer T2. Glycine is a synaptic amino acid that may be a neuromodulator or antioxidant (12, 13). Glycine elevation has been reported in astrocytomas, oligodendrogliomas, medulloblastomas, and central neurocytomas (12, 13).

Only mildly increased choline at 3.2 ppm and moderately decreased NAA at 2.1 ppm on MRS in our patient compared to

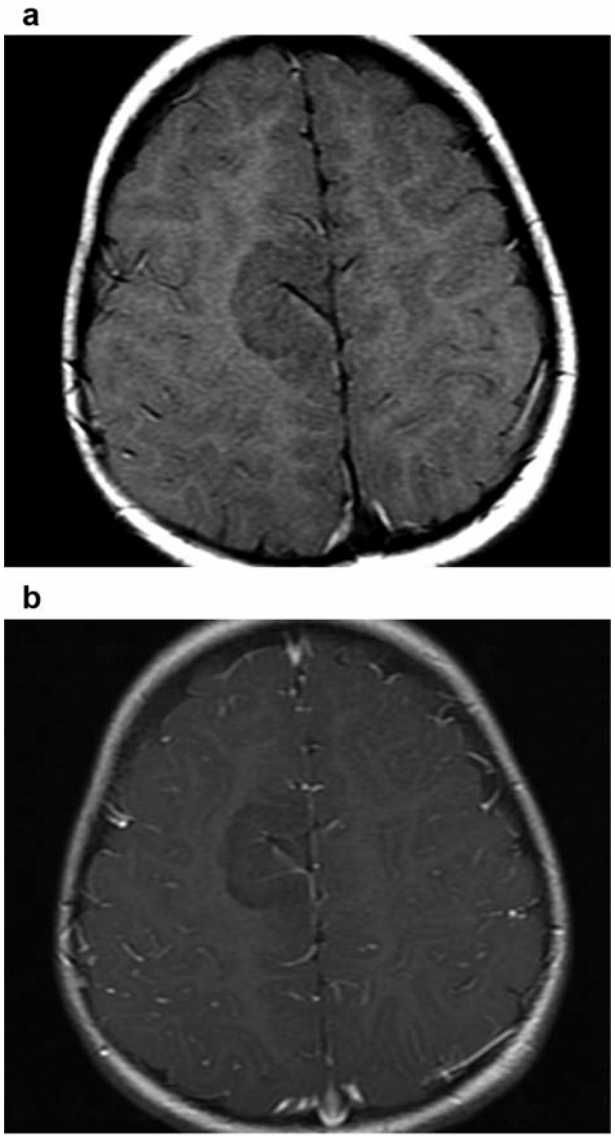

Figure 3. *Pre (a) and post-contrast (b) axial T1WI (repetition time msec/echo time msec, 517/ 21) depict T1 prolongation and no associated lesional contrast enhancement.*

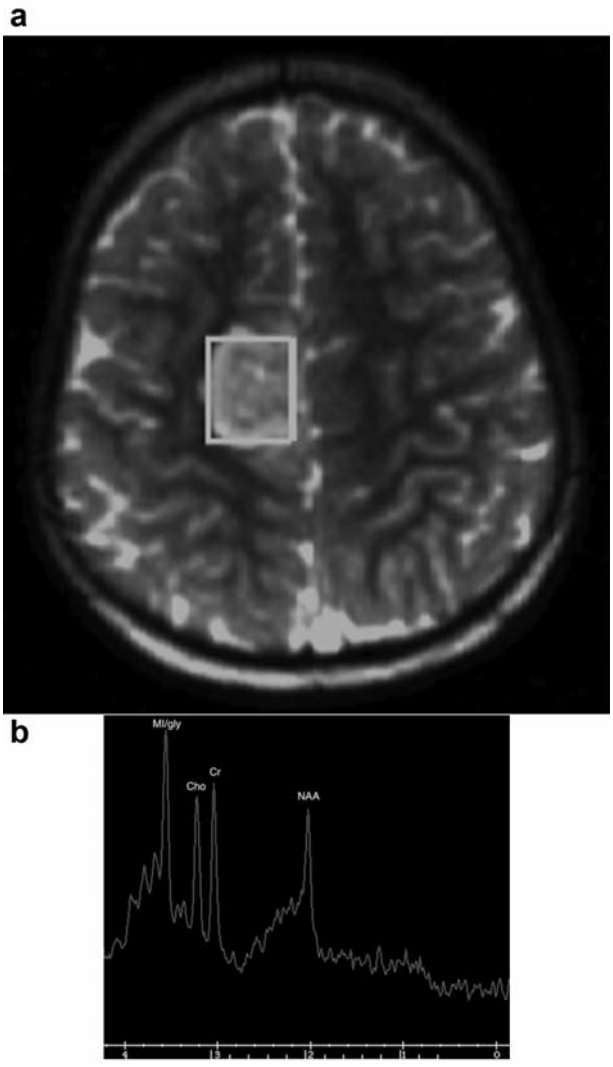

Figure 4. *a: Axial localizer T2WI demonstrating single voxel placement over the right frontal mass lesion. b:Single voxel point resolved MR spectroscopy through the right frontal mass (35 msec, TR 1500 msec)reveals a dominant metabolic peak at 3.55 ppm representing elevated myoinositol and/or glycine, mildly increased Cho:Cr, and moderately decreased NAA:Cr; no significant lactate is present. NAA, n-acetylaspartate; Cho, choline,; Cr, creatine; MI, myoinositol; gly, glycine.*

the corresponding contralateral brain parenchyma is probably a testament to the low-grade nature of the tumor. NAA is a marker of mature healthy neurons and reflects structural integrity of the neuronal-axonal unit (12). Choline elevation reflected increased cell membrane turnover or increased axonal density.

Statistically speaking, the primary differential diagnosis for a cortically-based lesion in a child primarily includes malformations of cortical development (focal cortical dysplasias, migrational and organizational gray matter abnormalities), glial tumors (astrocytomas, ependymomas, oligodendrogliomas), neuronal and glial/neuronal tumors, DNET (dysembryoplastic neuroepithelial tumor), primitive neuroectodermal tumor, and rare tumors such as astroblastoma and angiocentric glioma as in this case. The imaging spectrum of these lesions overlaps considerably; however, higher-grade tumors characteristically manifest solid lesional components that can demonstrate reduced diffusion and malignant MRS profiles: marked choline elevation and NAA diminution with associated lactate. Ultimately, histological evaluation may be required to distinguish lesions with atypical or non-diagnostic imaging features.

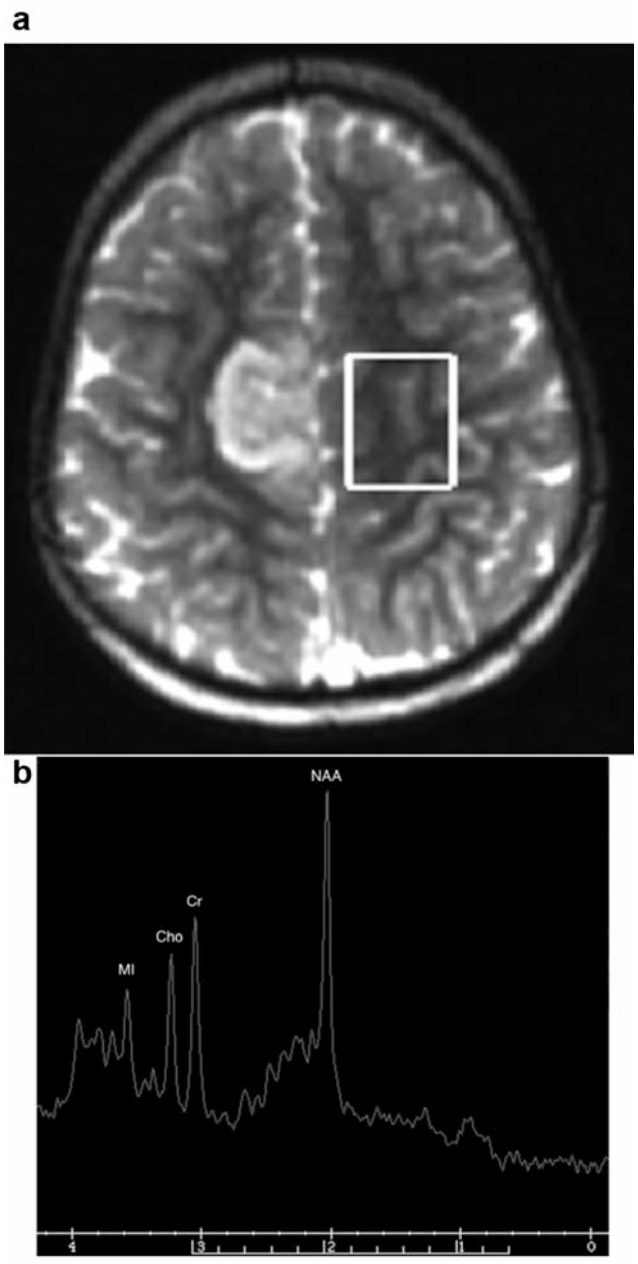

Figure 5*a: Axial localizer T2WI demonstrating single voxel placement over the structurally normal left superior frontal gyrus. b: Single voxel point resolved MR spectroscopy through the normal left superior frontal gyrus shows normal metabolic ratios for comparison. NAA=n-acetylaspartate; Cho, choline; Cr, creatine; MI, myoinositol; gly, glycine.*

Conclusion

We present an MR spectroscopy profile of an angiocentric glioma from a 2 year-old presenting with focal motor seizures. A dominant metabolic peak at 3.55 ppm may correspond to myoinositol and/or glycine elevation.

Conflicts of Interest

The Authors declare that there exist no conflicts of interest regarding the publication of this study.

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