

A case of pediatric infratentorial oligodendroglioma

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Abstract

Pediatric posterior fossa tumors most commonly are pilocytic astrocytomas (PA), ependymomas, medulloblastomas, and atypical teratoid rhabdoid tumor (ATRT). MRI has some special radiological findings which can be used in the differential diagnosis. We report on the radiologic images and the clinical course of a 3 year-old-girl with oligodendroglial tumor arising from the cerebellum. There were no focal deficits in the neurological examination. MRI revealed a left cerebellar lesion extending into the left cerebellopontine region causing a pressure and shift in the fourth ventricle, pons and mesencephalon. She underwent a left occipital craniectomy and total tumor decompression. In the pathological examination, mainly oligodendroglial cells were seen in the tumor and they had round nucleus and perinuclear halos. No recurrence was detected at MRI and CT at 6 months postoperatively. Except for the following most common tumors as PA, ependymoma, medulloblastoma and AT/RT in the posterior fossa, the oligodendroglioma must be considered in the differential diagnosis.

Keywords: Radiology; Oligodendroglioma; Pediatric.

INTRODUCTION

Central nervous system (CNS) tumors are the second most frequent tumor group in childhood (1). Almost half of the pediatric intracranial tumors occur at the posterior fossa. More than 90% of these tumors are ependymoma, pilocytic astrocytoma (PA), atypical teratoid rhabdoid tumor (ATRT), and medulloblastoma (2). MRI has some special radiological findings which can be used in the differential diagnosis. Here, we report on the radiologic images and the clinical course of a 3 years old girl with oligodendroglial tumor arising from the cerebellum.

CASE REPORT

A 3 year-old-girl presented with vomiting after falling from a 30-40 cm chair. The patient was in a good state of health before this accident. On physical examination vital signs were stable. There were no focal deficits in the neurological examination. Laboratory studies were all within normal limits. Computed tomography (CT) revealed a 4 × 3.5 cm sized hypodense, left cerebellar hemispheric lesion with millimetric calcifications. The lesion was extending into the left cerebellar vermis and had a mass effect on the fourth ventricle (Figure 1).



Figure 1. Non-contrast Axial CT : Hypodense left cerebellar hemispheric lesion with millimetric calcifications

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MRI revealed a left cerebellar lesion extending into the left cerebellopontine region causing a pressure and shift in the fourth ventricle, pons and mesencephalon. The lesion was hypointense in T1 and heterogeneous hyperintense in T2 sequence (Figure 2).

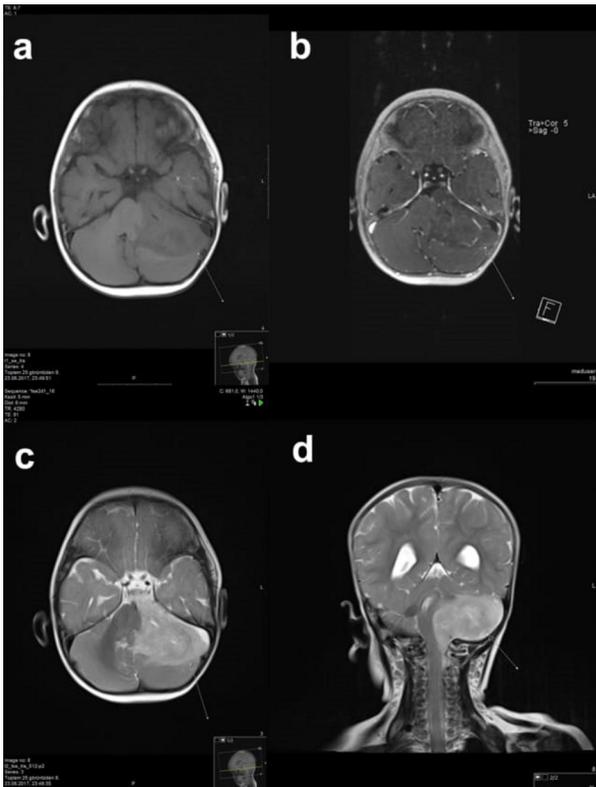


Figure 2. MRI Images a, Non-contrast axial T1 weighted image. b, postcontrast axial T1 weighted image. c, non-contrast axial T2 weighted image. d, Non-contrast coronal T2 weighted image

The lesion showed minimally contrast enhancement and moderately restricted in diffusion. A region of interest (ROI) measurement was obtained from the tumor lesion with the lowest signal on apparent diffusion coefficient (ADC) map. The ADC value of the lesion was $0,9 \times 10^{-3} \text{ mm}^2/\text{s}$ (Figure 3).

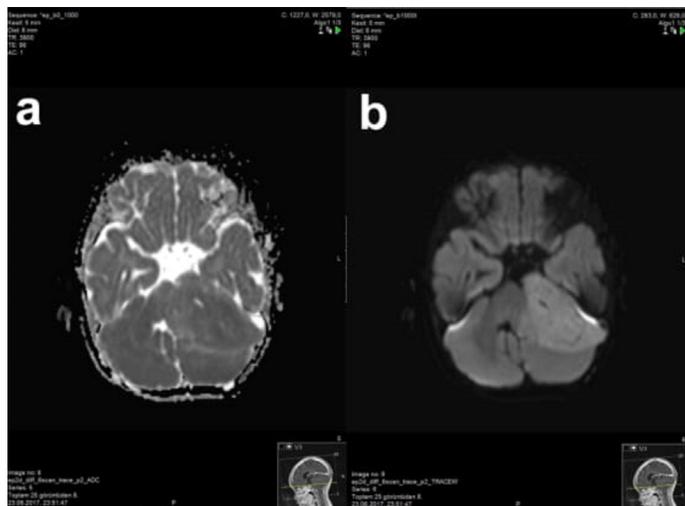


Figure 3. Diffusion weighted MRI images. a, ADC image. b, diffusion weighted image (DWI)

ADC ratio, 1.2 in our case, was obtained by dividing mean ADC value of lesion by ADC value of normal cerebellum parenchyma. She underwent a left occipital craniectomy and total tumor decompression. In the pathological examination, mainly oligodendroglia cells were seen in the tumor and they had round nucleus and perinuclear halos. Immunohistochemical staining showed that the tumor was positive for GFAP and olig2. There was no positivity for synaptophysin stain (Figure 4).

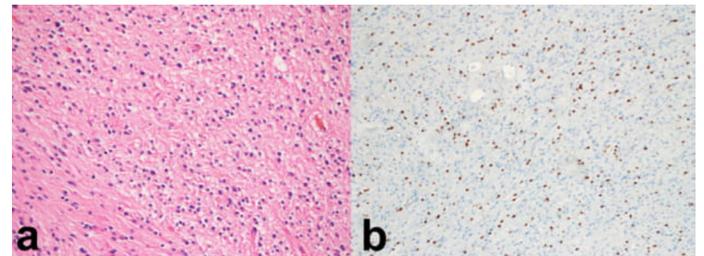


Figure 4 a. Oligodendroglial cells with perinuclear halos H&E x 100. **b.** positive staining of oligodendroglial cells with Olig2 antibody Olig2 Ab x 100

The Ki-67 proliferation index was $< 5\%$. Since genetic tests for IDH mutation and 1p19q codeletion could not be done because of the technical deficiencies, the tumor type according to the WHO classification could not be determined. Postoperatively, she did not receive local radiotherapy or chemotherapy because of the age of the patient and the low grade of the totally excised tumor. A follow-up MRI and CT at 6 months showed no recurrence of the lesion.

DISCUSSION

Pediatric posterior fossa tumors most commonly are pilocytic astrocytomas (PA), ependymomas, medulloblastomas, and atypical teratoid rhabdoid tumor (ATRT).

Classical imaging appearance of PA is large cystic lesion with a small mural nodule. Also, the solid portion of PA typically has diffusion characteristics of greater than $1,3 \times 10^{-3} \text{ mm}^2/\text{s}$ (3). The most common tumor seen in the posterior fossa is medulloblastoma. The tumor is usually in the vermis at the midline and grows into the fourth ventricle (4). Even in the absence of calcifications, a high-density appearance is seen on the CT because of the high nuclear to cytoplasmic ratio. The appearance on T2W imaging is relatively hypointense. The ADC values are low, typically lower than $0,8 \times 10^{-3} \text{ mm}^2/\text{s}$ (3). ATRT is malignant CNS neoplasm occurring mostly in children younger than 2 years (5). On imaging, this tumor has many similarities with medulloblastoma. Since ATRT has high cellularity, the ADC values of this tumor are low, typically lower than $0,7 \times 10^{-3} \text{ mm}^2/\text{s}$. Posterior fossa ependymomas have origin either at midline or at lateral. The most common form is midline tumors filling the fourth ventricle. Lateral ependymomas which can project into the medullary and cerebellopontine angles arise from the inferior margin of the middle cerebellar peduncle (6). Signs of calcifications are seen in half of the lesions. Also, small internal cystic

components are common. Because of the anatomic localization, the imaging findings like calcifications in the lesion and the ADC values, our preoperative radiologic diagnosis was ependymoma.

Pediatric oligodendroglial tumors are rare and are most commonly located in the cerebral hemispheres. However, infrequently, they can be located infratentorial. Although the typical radiological findings of oligodendrogliomas include areas of calcification with heterogeneous contrast enhancement (7), degree of enhancement is extremely variable due to the tumor grade, ranging from no enhancement to strikingly vivid enhancement. High grade tumors do more commonly show contrast enhancement, as well as necrosis, hemorrhage and peritumoral edema on MRI. However none of these findings are reliable features of high tumor grade (8).

CONCLUSION

In conclusion, except for the following most common tumors as PA, ependymoma, medulloblastoma and AT/RT in the posterior fossa, the oligodendroglioma must be considered in the differential diagnosis if the following imaging characteristics as areas of calcifications with heterogeneous contrast enhancement are present.

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