

## Pediatric cerebellar cystic oligodendroglioma: case report and literature review

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**SUMMARY:** Baysefer A, Düz B, Erdoğan E, Deveci MS. Pediatric cerebellar cystic oligodendroglioma: case report and literature review. Turk J Pediatr 2004; 46: 95-97.

Oligodendrogliomas rarely occur in the posterior fossa of childhood and constitute approximately 1% of pediatric brain tumors. Only six pediatric posterior fossa oligodendroglioma cases have been reported to date and none of them were cystic. The authors present a seven-year-old girl with cystic, cerebellar midline localized tumor. A standard suboccipital craniectomy was performed and the tumor was histologically confirmed as oligodendroglioma. After operation the patient underwent radiation therapy and at one the-year follow-up, no recurrence of the tumor was observed.

**Key words:** pediatric oligodendroglioma, cyst, astrocytoma.

Oligodendrogliomas constitute approximately 1% of pediatric brain tumors<sup>1,2</sup>. The most common localizations of oligodendrogliomas in the pediatric population are supratentorial, with the majority of them in the frontal lobe. Involvement of the frontal and parietal lobes was reported by Razack et al.<sup>3</sup> as 42.1% and 36.6%, respectively. Infratentorial oligodendrogliomas have been reported in six pediatric cases in the literature to date.

The authors report the first cystic oligodendroglioma localized in the posterior fossa in the midline in a seven-year-old girl.

### Case Report

A seven-year-old girl admitted to our department with a one-month history of vomiting and the slight loss of tandem gait. Brain magnetic resonance imaging (MRI) revealed a cystic lesion in the cerebellum localized in the posterior fossa in the midline. Contrast enhancements were seen in the T1-weighted images. There was hydrocephalus. The brain stem and the cerebral aqueduct were minimally compressed by the tumor. MRI findings resembled cerebellar cystic astrocytoma (Figs. 1, 2). We preoperatively examined the cerebrospinal fluid (CSF) for malignant cells but the result was negative.

The patient underwent surgery. A standard suboccipital craniectomy was performed. The cyst of the lesion was first aspirated, and then the

capsule of the tumor was totally excised under operation microscope. There was mural nodule other than the tumor capsule. The fourth ventricle was not opened in the operation. There was no complication after the surgery. Pathological examination revealed oligodendroglioma (Fig. 3). The tumor was histologically determined and then the patient underwent radiation therapy. One year later follow-up MRI showed no recurrence of the tumor.

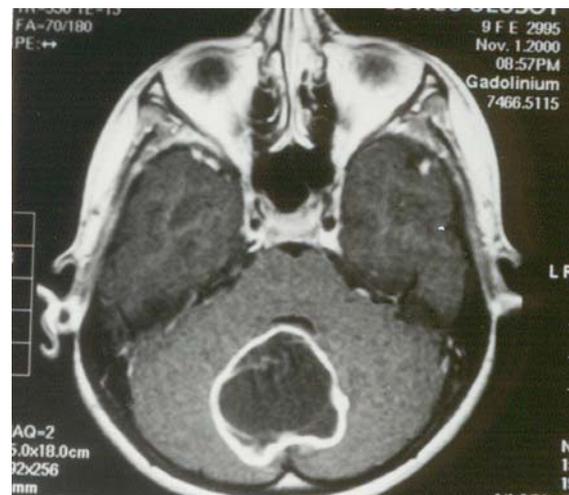


Fig. 1. The T1-weighted, axial magnetic resonance image after gadolinium enhancement. The fourth ventricle is compressed.



Fig. 2. The T1-weighted, sagittal magnetic resonance image.

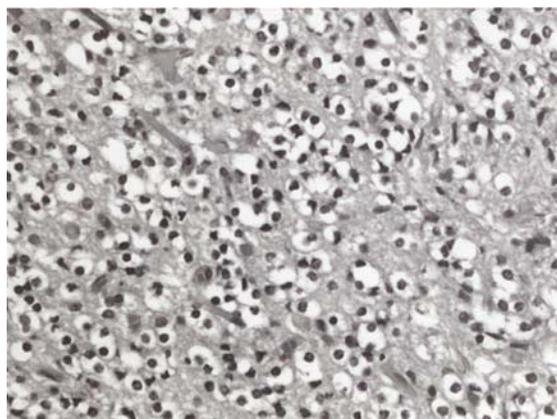


Fig. 3. Fibrillary and vascularized background of oligodendroglioma: many calcium spherulites were seen among oligodendrocytes (H&E, x200).

## Discussion

Oligodendrogliomas of the cerebrum account for 1% of pediatric brain tumors<sup>1,2</sup>. Posterior fossa localized oligodendrogliomas have been reported randomly in the literature. There are only six cases in the literature reported to date and none was cystic. The first case of cerebellar oligodendroglioma in the literature was reported by Holliday et al.<sup>4</sup> in 1980. Packer et al.<sup>5</sup> reported four cases of oligodendroglioma of the posterior fossa in childhood. According to Packer et al, posterior fossa origin is relatively more common in childhood. Bhatoe<sup>6</sup> from India reported the “childhood cerebellar vermian oligodendroglioma” in 1999 (Table I).

The differential diagnosis among pilocytic astrocytoma and other astrocytomas is important, because the pilocytic astrocytoma has a better prognosis than diffuse astrocytoma counterparts, especially when it occurs in the cerebellum<sup>7</sup>. Rosenthal fibers assist in distinguishing the pilocytic astrocytoma from other variants. These fibers, typical structures for pilocytic astrocytomas, are highly eosinophilic, hyaline structures. They are round, oval, or beaded, with slightly irregular margins<sup>8</sup>. In addition, eosinophilic droplets of protein are sometimes found intracellularly in association with Rosenthal fibers. Their bright pink color, similar to Rosenthal fibers, distinguishes them from mucoid degeneration in

Table I. Cases of Cerebellar Oligodendrogliomas in Childhood

Authors and year	Patients	Mean age	Pathology
Holliday et al. <sup>4</sup> , 1980	1	--	Oligodendroglioma
Packer et al. <sup>12</sup> , 1985	4	7.5	Malignant oligodendroglioma <sup>3</sup> Oligodendroglioma <sup>1</sup>
Bhatoe et al. <sup>6</sup> , 1999	1	--	Oligodendroglioma

--: not available.

The diagnosis of oligodendrogliomas or ependymomas is suggested preoperatively when intratumoral or peritumoral calcification is noted on neuroimaging studies. In this case neither MRI nor cranial computed tomography revealed any calcification. The tumor was cystic and contrast enhancement was seen in the T1-weighted images. Preoperative diagnostic studies indicated cerebellar cystic astrocytoma. The diagnosis is achieved histologically by pathological examination.

oligodendrogliomas. Neither Rosenthal fibers nor eosinophilic droplets were present in this cerebellar tumor.

The cellular features of ependymomas vary between fibrillar and epithelial, posing special problems of differentiation, not only from other gliomas. Presence of rosettes would confirm suspicion of an ependymoma, but some samples of ependymoma also lack rosettes. Clear cell ependymomas, uncommon gliomas, resemble mixtures of oligodendroglioma and ependymoma.

They usually include perivascular rosettes and cilia, which are absent in oligodendrogliomas. A pure oligodendroglioma is less fibrillar than clear cell ependymoma<sup>9</sup>. The pure oligodendroglioma differs from other gliomas, except for a few ependymomas, in having an epithelioid rather than a fibrillar appearance. This appearance is most evident within the central portion of the neoplasm, which is mostly crowded with neoplastic cells. Well-differentiated oligodendrogliomas usually have sheets of monotonous cells with uniform central nuclei, surrounded by a perinuclear clear halo of cytoplasm, therefore resembling fried eggs<sup>10</sup>. Perinuclear halos were also an important feature of the biopsy specimens of our case. There is a delicate network of capillaries, and it contains numerous foci of calcification. However, microcalcospherites are also commonly seen in other glial tumors such as ependymomas and pilocytic astrocytomas.

Dysembryoplastic neuroepithelial tumor (DNT) has been described in the posterior fossa<sup>11</sup>, and may show an oligodendroglioma-like hypercellularity. The nodules of DNT are composed of cells more uniform in size with nuclear chromasia than those of oligodendroglioma. Unlike the nodules occasionally seen, nodules of DNT are more discrete and patterned. In DNT, particularly paranodular and internodular cortices include mucin accumulation and isolation of neurons within mucous pools ("floating neurons") rather than satellitosis. Intra nodular cells are often arranged in clusters or intricate patterns, unlike the cells of oligodendroglioma.

Subarachnoid metastatic disease (SAMD) secondary to a dissemination of childhood primary central nervous system neoplasms is well documented<sup>12,13</sup>. SAMD is frequently a thin coating on the cord, nerve roots, or meningeal surface<sup>14,15</sup>. Sensitive evaluation for SAMD requires cytopathologic evaluation of the cerebrospinal fluid<sup>13</sup>, but several series have documented that even multiple samples of CSF may not yield an accurate diagnosis<sup>16</sup>. In our case, preoperative examination of CSF was not positive for malignant cells. On the basis of their experience, Packer et al.<sup>5</sup> suggested local and presymptomatic craniospinal radiation for all children with oligodendroglioma of the posterior fossa. But because of the rarity of posterior fossa localized oligodendrogliomas,

there is no evidence that radiation therapy prolongs survival in these cases. Only local radiation was performed in this case.

Finally, although cerebellar oligodendrogliomas are very rare lesions, they should be kept in mind in pediatric patients.

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