# Supratentorial ependymoma

## Epidemiology

Supratentorial intracranial ependymomas are rare, especially in the third ventricle.

Approximately half of the supratentorial ependymomas arise from the wall of the ventricles, whereas the remainder seem to arise from the brain parenchyma itself.

## Classification

Lateral ventricle ependymoma.

Third ventricular ependymoma.

Pediatric supratentorial ependymoma.

Supratentorial hemispheric ependymoma.

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Supratentorial ependymoma ZFTA fusion-positive

Supratentorial ependymoma YAP1 fusion-positive

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The majority of supratentorial ependymomas (ST-ependymomas) have few mutations but frequently display chromothripsis of chromosome 11q that generates a fusion between C11orf95 and RELA (RELAFUS). Neural stem cells transduced with RELAFUS ex vivo form ependymomas when implanted in the brain. These tumors display enhanced NF-κB signaling, suggesting that this aberrant signal is the principal mechanism of oncogenesis. However, it is not known whether RELAFUS is sufficient to drive de novo ependymoma tumorigenesis in the brain and, if so, whether these tumors also arise from neural stem cells. Ozawa et al., show that RELAFUS drives ST-ependymoma formation from periventricular neural stem cells in mice and that RELAFUS-induced tumorigenesis is likely dependent on a series of cell signaling pathways in addition to NF-κB [1)]

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### Case series

Supratentorial ependymoma is usually characterized by NOTCH-1 mutation and p75 expression. TNC mutation, no hypermethylation of RASSF1A, and GFAP/NeuN expression may be diagnostic clues of posterior fossa ependymoma. Although MEN1, TP53, and PTEN mutations are rarely reported in ependymoma, they may be related to a poor prognosis, such as recurrence or metastasis. Spinal ependymoma has been found to be quite different from intracranial ependymoma in genetic studies,

and the favorable prognosis in spinal ependymoma may be the result of the genetic differences. A more detailed understanding of these various genetic aberrations may enable the identification of more specific prognostic markers as well as the development of customized targeted therapies [2].

### 2013

Fourteen of the 23 supratentorial ependymomas were in the region of the third ventricle and the remainder were located in the hemispheres. Resections were gross total in 12 patients, subtotal in 8, and biopsy in 3. A single pathologist reviewed all slides and quantitated the deoxyribonucleic acid. The mean follow-up duration was 95 months (+/-75 mo).

All of the malignant ependymomas were hemispheric (n = 4). Mortality occurred only in patients with third ventricular tumors; two patients died as a result of surgical complications and three as a result of tumor progression. Kaplan-Meier estimates of 5- and 10-year survival rates were 100% for hemispheric and 72.5% for third ventricular tumors (62.5% including the two perioperative deaths). The median time to recurrence was 53 months, with a 10-year progression-free survival rate of 27%. Univariate analysis revealed that recurrence was associated with malignant histology, including mitoses, cellularity, and aneuploidy. For nonmalignant ependymomas, recurrence was associated with subtotal resection and metastases. S-phase fraction did not correlate with recurrence. Only malignant histology correlated with recurrence on multivariate analysis.

Although the numbers are too small to draw any definite conclusions, treatment of ependymomas that arise in the supratentorial compartment in adult patients results in excellent outcomes despite frequent recurrences. Association with the third ventricle and metastases seem to have a negative impact on survival, whereas malignant histology, subtotal resection, and metastases may be predictors of recurrence [3].

## Case reports

### 2017

A case of aggressive anaplastic ependymoma arising in the right frontoparietal lobe, which had genetically 1q25 gain, CDKN2A homozygous deletion, and L1CAM overexpression. The patient was a 10-year-old boy who underwent four times of tumor removal and seven times of Gamma knife radiosurgery. Metastatic loci were scalp and temporalis muscle overlying primary operation site, lung, liver, buttock, bone, and mediastinal lymph nodes. He had the malignancy for 10 years and died. This tumor is a representative case of Ependymoma RELA fusion positive, showing aggressive behavior [4].

1)
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2)
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4)
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