# Posterior fossa ependymoma

More than 70% of intracranial ependymomas occur in the infratentorial region, usually arising from the fourth ventricle floor in the midline.

Recurrent ACVR1 mutations in posterior fossa ependymoma<sup>1)</sup>

# Classification

Posterior Fossa Ependymoma Classification.

## **Clinical features**

Posterior fossa tumor clinical features.

# Diagnosis

Posterior fossa ependymoma diagnosis

## Treatment

Posterior fossa ependymoma treatment.

#### Outcome

Posterior fossa ependymoma outcome.

#### **Case series**

All surgical procedures for posterior fossa ependymomas performed since 2001 in the Department of Neurosurgery, University Medicine Greifswald, were analysed.

8 patients (2f, 6m, mean age 41.9 years, range from 29 to 56 years) underwent surgery for posterior fossa ependymoma. All tumors were WHO°II. Tumor adherence was found to be in the caudal rhomboid fossa (between the obex and striae medullare, but below the facial colliculus) in all patients. The four-hand suction-irrigation technique led to gross total resection in all patients (100%) without significant permanent neurological deficits after surgery. None of the patients got further treatment (chemotherapy, radiation therapy, second surgery). In none of these patients, tumor recurrence was

seen on MRI after a mean follow-up of 102 months (ranging from 14 to 181 months).

Long-term progression free survival in adult patients suffering from posterior fossa ependymoma is possible by gross total resection without adjuvant radio- or chemotherapy. By careful bimanual microsurgical dissection using the four-hand suction-irrigation technique and avoidance of bipolar coagulation on the floor of the fourth ventricle, the risk for permanent neurological deficits is low <sup>2</sup>.

## **Case reports**

A case of a 32-year-old male patient complained of headaches for three months. Pure tone audiometry showed a slight decrease in bilateral hearing. Auditory evoked potential indicated that the hearing on the left was slightly weaker than that on the right. Magnetic resonance imaging (MRI) revealed a primary tumor arising within the fourth ventricle and metastasizing to bilateral cerebellopontine angle (CPA), the third ventricle, the left lateral ventricle, T1, L1-2 and L5. A gross total resection (GTR) was performed on the lesion located in the left CPA. The histological examination showed a papillary ependymoma (WHO grade II). Immunohistochemical staining for H3K27me3 showed that nuclear positivity in more than 80% of cells. No NF2 mutation was observed. No progression was found during a 24-month follow-up.

This data indicate that preoperative multiple metastases in adult PF-EPN are extremely rare. This kind of disease usually has a low WHO grade and a favorable prognosis. GTR should be achieved when feasible and patients need a long-term follow-up with MRI<sup>3</sup>.

The development of secondary neoplasms following therapeutic cranial irradiation is rare and quite often lethal. Meningiomas, sarcomas, and high-grade gliomas are the most common tumors that manifest as a result of radiation therapy. A 11-year-old child who presented with symptoms of supratentorial space-occupying lesion 7 years after curative surgery and cranial irradiation for a posterior fossa ependymoma. Magnetic resonance imaging of the brain revealed a right-sided temporoparietal dural-based contrast-enhancing lesion with evidence of overlying bone and skin involvement. The histological report of ependymoma from the previous surgery led us to suspect that we were dealing with a recurrence until the histopathology of the second surgery revealed highly malignant osteosarcoma. The child recovered fully and underwent chemotherapy, but ultimately succumbed to the disease <sup>4)</sup>.

A 13-year-old boy with severe clinical symptoms and signs underwent surgery for a posterior fossa ependymoma in 1954. The tumor was adjacent to the floor of the fourth ventricle, and surgery was complicated by profound bleeding. Therefore, only a partial resection was performed. Postoperative radiotherapy was given to the posterior fossa. The recovery was uneventful, and he has been in full-time work until the age of 62 years and is now 74 years old. Repeated MRI scans demonstrate a stable residual fourth ventricular tumor <sup>5)</sup>.

## References

3)

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