Intraventricular tanycytic ependymoma

- Synchronous presentation of prolactinoma and supratentorial tanycytic ependymoma
- Minimally Invasive Parafascicular Surgical Approach for the Management of a Pediatric Third Ventricular Ependymoma: Case Report and Review of Literature
- Lack of IDH1 mutation in astroblastomas suggests putative origin from ependymoglial cells?
- Features of intraventricular tanycytic ependymoma: report of a case and review of literature
- intraventricular tanycytic ependymoma: an uncommon fibrillary variant
- A 40-year-old male with an intraventricular tumor. Combined tanycytic ependymoma and subependymoma
- Ventricular and extraventricular ependymal tumors in 18 cats
- Intraventricular tanycytic ependymoma: case report and review of the literature

Case reports

A 29-year-old male was referred from the infertility clinic after serum prolactin levels were found to be elevated. Magnetic resonance imaging (MRI) brain showed an irregular necrotic lesion in the periventricular region of the left parietal lobe which had an intraventricular component and associated perilesional edema. In addition, a sellar mass with suprasellar extension was also found on the MRI. He was started on cabergoline therapy for macroprolactinoma and underwent a left parietal craniotomy, and microsurgical excision of the tumor using intraoperative neurosonographic guidance. Histologically, the tumor showed spindle cytologic features and poorly developed inconspicuous pseudorosettes, with areas of rounded nuclear profiles and perinuclear cytoplasmic clearing. Tumor cells were positive for vimentin, glial fibrillary acidic protein, and S100 and negative for epithelial membrane antigen. Ki67 was <7%. He was diagnosed with tanycytic ependymoma and a coexistent prolactinoma. He received 10 cycles of image-guided radiotherapy. Post-operative imaging showed minimal residual tumor the size of which remained stable at 1-year follow-up scan. The pituitary macroadenoma regressed with cabergoline therapy and he clinically improved. This presentation of synchronous macroprolactinoma and tanycytic ependymoma has not been reported in the literature previously. An exhaustive literature review showed only 18 previously reported cases of supratentorial tanycytic ependymoma [1)].

A 7-year-old boy presented with worsening headache, nausea, vomiting, dizziness, unsteady gait, photophobia, and blind spots with positional changes. Magnetic resonance imaging (MRI) scan revealed a large isointense mass, with areas of hyperintensities suggestive of intratumoral hemorrhage, centered in the posterior segment of the third ventricle with extension into the aqueduct of Sylvius. The superior frontal sulcus was used as an access corridor for the port to the frontal horn of the lateral ventricle en route to the third ventricle. Intraoperative visualization was aided by a 3-dimensional exoscopic system. After cannulation, the tumor was seen within the foramen of Monro and tethered to the thalamostriate vein. The tumor was removed completely, except small residual attached to the thalamostriate vein, which was left intentionally. A flexible endoscope was placed through the port to verify the absence of residual along the superior wall of the third ventricle. An intraoperative MRI scan confirmed the presence of residual, along with normal postoperative changes, including pneumocephalus. A postoperative MRI scan revealed cortical recovery along the sulcal path and resolution of ventriculomegaly.

The patient improved from baseline, with no remaining visual deficits, headaches, or balance issues. Pathology reported a World Health Organization grade II tanycytic ependymoma [2].

A case of tanycytic ependymoma arising from the third ventricle was completed with an immunohistochemical, ultrastructural, and molecular pathology study. The patient was a 44-year-old male who presented with headache, nausea, and visual disturbances of a few months duration. Neuroradiological findings showed a well-defined mass arising from the posterolateral wall of the third ventricle. Histologically the tumor was composed of monotonous spindle cells arranged in fascicles without definitive perivascular rosettes. The tumor cells were diffusely positive for glial fibrillary acidic protein and epithelial membrane antigen, showed faint immunoreactivity for synaptophysin but were negative for neurofilament proteins and Ki-67 was less than 1%. Molecular studies showed the absence of isocitrate dehydrogenase gene 1 and 2 mutation. A diagnosis of tanycytic ependymoma (TE) was made. From the literature review with our current case included, intraventricular tanycytic ependymomas ranged from 1.8 to 4.0 cm. The age of patients ranged from 3.5 to 75 years with a mean age of 37.5 and a male predominance. The tumors occurred as a well-defined, solitary ventricular mass without significant peritumoral edema with or without cystic changes. Histopathology and immunohistochemical profile are rather similar among different tumors. The immediate to short-term outcome is excellent but long-term follow-up data is lacking [3]

1)
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2)
Chakravarthi SS, Lyons L, Bercu M, Singer JA. Minimally Invasive Parafascicular Surgical Approach for the Management of a Pediatric Third Ventricular Ependymoma: Case Report and Review of Literature. World Neurosurg. 2020 Sep;141:311-317. doi: 10.1016/j.wneu.2020.04.201. Epub 2020 May 5. PMID: 32387401.
3)
Agarwal S, Stevenson ME, Sughrue ME, Wartchow EP, Mierau GW, Fung KM. Features of intraventricular tanycytic ependymoma: report of a case and review of literature. Int J Clin Exp Pathol. 2014 May 15;7(6):3399-407. PMID: 25031767; PMCID: PMC4097299.