Intracranial ependymoma outcome

Relapsed intracranial ependymoma has a poor prognosis despite multimodal therapy. Novel therapeutic strategies are desperately needed for this disease [1].

Factors affecting prognosis are poorly understood.

PubMed was searched to identify studies that reported clinical outcomes in adult patients with intracranial ependymoma. Data were extracted for patient and tumor characteristics, extent of resection, progression-free survival (PFS), and overall survival (OS). Tumors were categorized as supratentorial or infratentorial and extraventricular or intraventricular. Presenting clinical features and tumor characteristics were tabulated. Kaplan-Meier and multivariate Cox regression survival analyses were performed to determine PFS and OS by tumor location. Extent of resection was also analyzed by tumor location. A total of 183 patients were included in the meta-analysis. Patients presented at a mean of 8.2months with a myriad of clinical features. The mean tumor size was 3.38 cm, and 19.3% of tumors were cystic. Supratentorial tumors were most commonly located in the frontal and parietal lobes, and infratentorial tumors in the fourth ventricle. Supratentorial tumors demonstrated significantly poorer PFS ($p<0.001$) and OS ($p=0.003$) than infratentorial tumors, despite a higher rate of gross total resection (GTR) for the supratentorial tumors (72.6% versus 42.1%). Extraventricular ependymomas displayed significantly poorer PFS than intraventricular ependymomas ($p=0.009$). In summary, supratentorial ependymomas have significantly poorer PFS and OS than their infratentorial counterparts, despite being more conducive to GTR, suggesting increased clinical aggressiveness. Extraventricular location is also associated with significantly poorer PFS than intraventricular location [2].

Recurrence occurs in almost 50% of patients with intracranial ependymoma, and their outcome following recurrence is poor.

In 22 patients with intracranial ependymoma and subsequent relapse(s) (59 recurrences) treated at Children's Hospital Los Angeles or New York University between January 1997 and December 2012, the median duration of follow-up was 52 months (7-171 months). Median age at initial diagnosis was 4 years (0.3-19 years) with 8 patients younger than 3 years at presentation. Eleven patients had anaplastic and 11 cellular pathologies. Eighteen patients had infratentorial tumors at diagnosis and 3 (all infratentorial) had metastatic spinal cord involvement at presentation. Cerebrospinal fluid involvement was not identified at diagnosis or relapse. Median time to first recurrence was 16 months (1.3 to 115 months). The number of recurrences in each patient ranged from 1 to 9 (median = 2). Thirty-seven recurrences (63%) were detected asymptomatically by surveillance imaging. Fifteen recurrences (26%) arose outside the initial tumor site. Recurrences were treated by surgical resection (45), with irradiation (30), and with various oral chemotherapies (23) with (7) or without (16) conventional chemotherapy. The 5 and 10 year overall survival rates from first recurrence were 0.37 ± 0.14 and 0.25 ± 0.14.

Prolonged (5-10 year) survival from first relapse was noted in over one-quarter of our patients. It remains unclear whether early radiographic diagnosis, differing treatment modalities beyond radical surgical resection or possibly unrecognized biological differences contributed towards this prolonged survival [3].

Data show that local radiation therapy may have long-term effects on patients' QoL. Since in the incompletely resected Grade II tumors local irradiation did not lead to a benefit in PFS in this retrospective study, prospective randomized studies are necessary. In addition to age, supratentorial

tumor location is associated with a worse prognosis in adult ependymoma patients [4].

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Tsai JW, Manoharan N, Alexandrescu S, Zimmerman MA, Scully J, Chordas C, Clymer J, Wright KD, Filbin M, Ullrich NJ, Marcus KJ, Haas-Kogan D, Chi SN, Bandopadhayay P, Yeo KK. Outcomes after first relapse of childhood intracranial ependymoma. Pediatr Blood Cancer. 2021 Feb 9:e28930. doi: 10.1002/pbc.28930. Epub ahead of print. PMID: 33565268.
2)
Sayegh ET, Aranda D, Kim JM, Oh T, Parsa AT, Oh MC. Prognosis by tumor location in adults with intracranial ependymomas. J Clin Neurosci. 2014 Dec;21(12):2096-101. doi: 10.1016/j.jocn.2014.05.011. Epub 2014 Jul 15. PubMed PMID: 25037313.
3)
Antony R, Wong KE, Patel M, Olch AJ, McComb G, Krieger M, Gilles F, Sposto R, Erdreich-Epstein A, Dhall G, Gardner S, Finlay JL. A retrospective analysis of recurrent intracranial ependymoma. Pediatr Blood Cancer. 2014 Feb 24. doi: 10.1002/pbc.24996. [Epub ahead of print] PubMed PMID: 24615997.
4)
Dützmann S, Schatlo B, Lobrinus A, Murek M, Wostrack M, Weiss C, Schaller K, Raabe A, Meyer B, Goldbrunner R, Franz K, Seifert V, Senft C. A multi-center retrospective analysis of treatment effects and quality of life in adult patients with cranial ependymomas. J Neurooncol. 2013 Sep;114(3):319-27. doi: 10.1007/s11060-013-1187-2. Epub 2013 Jun 29. PubMed PMID: 23813228.