2025/06/30 06:42 1/2 1987

Shi et al. analyzed adult and pediatric patients with newly diagnosed or recurrent intracranial ependymoma or spinal ependymomas treated with SRS in Stanford. Following SRS, local failure (LF) was defined as failure within or adjacent to the SRS target volume, while distant failure (DF) was defined as failure outside of the SRS target volume. Time to LF and DF was analyzed using competing risk analysis with death as a competing risk. Overall survival (OS) was calculated from the date of first SRS to the date of death or censored at the date of last follow-up using the Kaplan-Meier method.

Twenty-one patients underwent SRS to 40 intracranial (n = 30) or spinal (n = 10) ependymoma lesions between 2007 and 2018, most commonly with 18 or 20 Gy in 1 fraction. Median follow-up for all patients after first SRS treatment was 54 months (range 2-157). The 1-year, 2-year, and 5-year rates of survival among patients with initial intracranial ependymoma were 86, 74, and 52%, respectively. The 2-year cumulative incidences of LF and DF after SRS among intracranial ependymoma patients were 25% (95% Cl 11-43) and 42% (95% Cl 22-60), respectively. No spinal ependymoma patient experienced LF, DF, or death within 2 years of SRS. Three patients had adverse radiation effects.

SRS is a viable treatment option for intracranial ependymoma and spinal ependymoma with excellent local control and acceptable toxicity ¹⁾.

Intracranial ependymomas (n = 146) from children treated on the RT1 trial at St. Jude Children's Research Hospital were evaluated for the status of multiple pathological features. Interphase FISH (iFISH) defined the status of loci on chromosomes 1q (EXO1), 6q (LATS1) and 9, including 9p21 (CDKN2A). Data relating to these clinicopathological and cytogenetic variables were compared with survival data in order to model disease risk groups. Extent of surgical resection was a significant determinant of outcome in both supratentorial and infratentorial compartments. Tumor cell density and mitotic count were associated with outcome among children with posterior fossa ependymomas (n = 119). Among pathologic features, only brain invasion was associated with outcome in children with supratentorial ependymomas (n = 27). For posterior fossa tumors, gain of 1q was independently associated with outcome and in combination with clinicopathological variables defined both a two-tier and three-tier system of disease risk. Among children developing posterior fossa ependymomas treated with maximal surgical resection and conformal radiotherapy, key clinicopathological variables and chromosome 1q status can be used to define tiers of disease risk. In contrast, risk factors for pediatric supratentorial tumors are limited to sub-total resection and brain invasion 2 .

1987

In 33 patients undergoing operation and postoperative irradiation for intracranial ependymomas between January 1963 and December 1983, the tumor was grade 1 or 2 in 26 (79%) patients and grade 3 or 4 in 7 (21%). Operation consisted of only biopsy in 1 (3%), subtotal removal of tumor in 28 (85%), and gross total resection in 4 (12%). All patients received brain irradiation with a median dose of 4800 cGy. Seventeen (52%) patients also received spinal axis irradiation (median dose, 3000 cGy) which included 5 with high-grade tumors and 12 with low-grade infratentorial tumors. The relapse-free and overall survival rates at 5 years were 61% and 62%, respectively. Prognostic factors analyzed for statistically significant survival differences included age, sex, hydrocephalus, site, grade, extent of operation, extent of brain field, spinal axis irradiation, and brain dose. Grade was the only significant factor found: the 5-year survival of patients with low-grade ependymomas, 71%, was significantly better (p less than 0.04) than that of patients with high-grade ependymomas, 29%. Among the 31 patients evaluable for patterns of failure, treatment failed in 12 (39%) (10 only in the brain, 1 in the brain and spinal cord, and 1 only in the spine). All but one of the brain failures were at

02:58

the site of the original primary lesion. Treatment failed in 4 of the 6 (67%) patients with high-grade tumor but in only 8 of the 25 (32%) with low grade tumor. Among the 7 low-grade infratentorial ependymomas treated with brain irradiation only, there was 1 treatment failure (in the spine; salvaged with further irradiation). Among the 12 patients with low-grade infratentorial tumors who received spinal axis irradiation, treatment failed in 1 (8%) (in the spine and also in the brain; patient subsequently died of disease). Nineteen (58%) patients remain alive; all but 2 of the patients who had recurrence died of their disease. This retrospective study suggests that: (a) patients with high-grade tumors have significantly poorer survival compared with those with low-grade tumors; (b) the main cause of death in ependymoma patients is intracranial failure at the primary site; and craniospinal axis irradiation may not be necessary for patients with low-grade infratentorial ependymoma (localized irradiation alone may be adequate) ³⁾.

1)

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