Central neurocytoma case series

Kang et al. identified 61 patients with pathologically diagnosed CN treated between 1996 and 2016, of which 24 met inclusion criteria. Patient, tumor, and treatment characteristics are reported in context of progression-free survival and treatment-related adverse events.

Of 24 patients identified, median age at diagnosis was 21 years (range, 14-60). Median maximal tumor diameter was 4.5 cm (range, 1.4-6.8). Eighteen (75%) patients underwent upfront surgery alone. Sixteen (67%) patients received adjuvant or salvage PBT at a median dose of 54 Gy (relative biological effectiveness). Median follow-up was 56 months. Median progression-free survival (PFS) was 61 months. Eleven patients had disease progression with median time to progression of 22 months. Of the 5 patients with gross total resection, 4 experienced local recurrence and had MIB-1 >4% (range, 4.5-30). There was improved PFS with addition of PBT to definitive surgery (log-rank, P = .06); there was no disease progression to date. In patients who experienced disease recurrence/progression, MIB-1 <4% was associated with improved PFS (log-rank, P = .007). All patients tolerated PBT well with toxicities typical for cranial irradiation and with no grade 3+ toxicities.

In this cohort, CN with elevated MIB-1 index were at increased risk for disease progression. However, adjuvant radiation therapy appears to effectively prevent failure. PBT toxicities appear to be comparable to if not less than published photon experiences [1)].

Byun et al. classified a retrospective study cohort as 21 (52.5%) typical and 19 (47.5%) atypical CN cases. No significant differences were found in terms of sex, mean age, mean tumor size or tumor location between these groups. Recurrences occurred in 2 (9.5%) typical and 6 (33.3%) atypical cases. The typical CN 2-,3- and 5-year PFS rates were 100%, 100%, 92.3%, and those for the atypical group were 93.8%, 78.1%, 65.1%, respectively ($p = 0.02$). The PFS rates did not statistically differ by treatment modality (gross total resection alone, subtotal resection (STR) alone and STR plus radiation therapy (RT) or radiosurgery (RS)) either in the whole cohort ($p = 0.75$) or in the typical CN and atypical CN subgroups ($p = 0.45$ and 0.98, respectively). An atypical histology was the only prognostic indicator of recurrence by univariate analysis (hazard ratio: 5.40, $p = 0.04$).

An atypical lesion (MIB-LI > 2%) is an important prognostic indicator in CN. The clinical implications of the extent of resection for CN patients are still debatable. The use of STR plus RT or RS may be a viable treatment strategy for CN but different therapeutic and follow-up approaches for atypical CN will be needed [2)].

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868 patients were diagnosed with biopsy-proven neurocytoma and analyzed (0.4% or approximately 75 patients annually). Median age at diagnosis was 31 years and median tumor size was 4-5 cm. Diagnosis was similar between male (49.5%) and female (50.5%). Regarding location, 622 (72%) tumors were intraventricular, 168 (19%) were extra-ventricular, and 78 (9%) overlapping or unspecified. Five-year overall survival among all patients was 89%. On multivariable analysis tumor location, extent of resection, and use of radiation, were not predictive for improved survival (each $p > 0.05$); however, patient age ($p < 0.001$), WHO grade ($p < 0.001$), and medical comorbidity scores ($p = 0.002$) were independently associated with overall survival.

Patients with central neurocytoma often present as young adults with sizable tumor burden and are well managed with surgery alone. Considering their favorable survival, efforts to improve tumor control should be carefully weighed against the long-term risks associated with adjuvant therapy like radiation. [3)]

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A total of 28 patients (15 males, 13 females) were treated between 1995 and 2014, with a median age at diagnosis of 26 years (range 5-61). Median follow-up was 62.2 months and 3 patients were lost to follow-up postoperatively. Thirteen patients experienced recurrent/progressive disease and 2-year PFS was 75 % (95 % CI 53-88 %). Two-year PFS was 48 % for MIB-1 labeling >4 % versus 90 % for ≤4 % (HR 5.4, CI 2.2-27.8, p = 0.0026). Nine patients (32 %) had gross total resections (GTR) and 19 (68 %) had subtotal resections (STR). PFS for >80 % resection was 83 versus 67 % for ≤80 % resection (HR 0.67, CI 0.23-2.0, p = 0.47). Three STR patients (16 %) received adjuvant radiation which significantly improved overall PFS (p = 0.049). Estimated 5-year PFS was 67 % for STR with radiotherapy versus 53 % for STR without radiotherapy. Salvage therapy regimens were diverse and resulted in stable disease for 54 % of patients and additional progression for 38 %. Two patients with neuropathology-confirmed atypical neurocytomas died at 4.3 and 113.4 months after initial surgery. For central neurocytomas, MIB-1 labeling index >4 % is predictive of poorer outcome and our data suggest that adjuvant radiotherapy after STR may improve PFS. Most patients requiring salvage therapy will be stabilized and multiple modalities can be effectively utilized [4)].

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203 patients, with a median age of 31 years; 46% of patients were male, and 80% of patients were white. GTR was performed in 47% of patients, with the remainder receiving STR. Adjuvant RT was delivered to 15% of patients, including 9% after GTR and 20% after STR (P = .018). Age, sex, race, diagnosis year, World Health Organization (WHO) grade, and tumor size were not significantly associated with GTR or RT utilization. OS was 87% at 5 years for all patients, with a median follow-up of 39 months. There was no significant OS benefit with GTR compared with STR (89% vs 86%; P = .82). There was also no significant OS benefit with RT after either GTR (86% with RT vs 90% without RT at 5 years; P = .76) or STR (79% with RT vs 89% without RT at 5 years; P = .53) [5)].

Xiong et al. described and analyzed the clinical, radiological, operational and outcome data of 13 cases of huge intraventricular CNs, and discussed the treatment strategies in this study. All huge CNs (n=13) in the study were located in bilateral lateral ventricle with diameter ≥5.0 cm and had a broad-based attachment to at least one side of the ventricle wall. All patients received craniotomy to remove the tumor through transcallosal or transcortical approach and CNs were of typical histologic and immunohistochemical features. Adjuvant therapies including conventional radiation therapy (RT) or gamma knife radiosurgery (GKRS) were also performed postoperatively. Transcallosal and transcortical approaches were used in 8 and 5 patients, respectively. Two patients died within one month after operation and 3 patients with gross total resection (GTR) were additionally given a decompressive craniectomy (DC) and/or ventriculoperitoneal shunt (VPS) as the salvage therapy. Six patients received GTR(+RT) and 7 patients received subtotal resection (STR)(+GKRS). Eight patients suffered serious complications such as hydrocephalus, paralysis and seizure after operation, and patients who underwent GTR showed worse functional outcome [less Karnofsky performance scale (KPS) scores] than those having STR(+GKRS) during the follow-up period. The clinical outcome of

huge CNs seemed not to be favorable as that described in previous reports. Surgical resection for huge CNs should be meticulously considered to guarantee the maximum safety. Better results were achieved in STR(+GKRS) compared with GTR(+RT) for huge CNs, suggesting that STR(+GKRS) may be a better treatment choice. The recurrent or residual tumor can be treated with GKRS effectively [6].

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