OTHER BRAIN TUMORS (BT)

BT-02

MULTIDISCIPLINARY TREATMENT FOR EPENDYMOMA

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BACKGROUND: In intracranial ependymoma, the effectiveness of chemotherapy and radiation therapy is unclear, and the degree of tumor removal contributes to the improvement of life prognosis. Methods: We examined ependymoma cases treated in our institution from July 1998 to March 2017. RESULTS: There were 18 boys and 7 girls. The average age at the time of surgery is 5.3 ± 3.6 years. The pathological diagnosis was Grade II for 8 cases and Grade III for 17 cases. Genetic analysis was performed in 16/25 cases (64%). Of the infratentorial cases, 10/11 cases (90.1%) were PFA and PFB were one case. Of the supratentorial cases, 3/5 cases (60%) were positive for RELA fusion. As chemotherapy, 19 patients were VCR + VP-16 + CDDP + CPA. Irradiation was performed in all cases, local irradiation (50.4–55.8Gy) in 22 cases (88%), and craniospinal irradiation in 2 cases (8%). The 7-year OS was 74.6 ± 9% and the 7-year PFS was 59.7 ± 10.5%. Grade III showed a short OS ($p = 0.053$). GTR and NTR were obtained in the first excision in 14 cases (56%), and OS and PFS were not significantly different from those in the STR group ($p = 0.219$, $p = 0.248$). GTR and NTR including 2nd-look surgery were obtained in 18 cases (72%), and significant improvement of OS was observed compared with STR group ($p = 0.02$). In patients with hydrocephalus preoperatively, OS tended to be short ($p = 0.057$), especially in cases requiring VP shunt placement, OS was significantly shortened ($p = 0.017$). CONCLUSION: Even if it is not GTR or NTR at the first operation, improvement of OS is expected by total excision after chemotherapy. The importance of chemotherapy was suggested to be suppression of tumor growth until reoperation and reduction of blood loss during surgery.

BT-03

A CASE OF ADULT-ONSET MEDULLOBLASTOMA PRESENTING WITH ATYPICAL CLINICAL COURSE AND MAGNETIC RESONANCE IMAGING

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An 18-year-old woman gradually developed speech and gait disturbance for seven months. On admission, she presented with cerebellar ataxia and tetraparesis. Magnetic resonance images showed diffuse hyperintense lesions around the fourth ventricle in FLAIR, in addition to lesions of nodular diffusion restriction with enhancement. Heavily T2-weighted images revealed small cystic appearance within this lesion. We diagnosed her as classical medulloblastoma by open brain biopsy. We should consider medulloblastoma as a differential diagnosis of these characteristics around the fourth ventricle, even if magnetic resonance findings are atypical in point of no mass effect and heterogeneous enhancement.

BT-05

A CASE OF GLIOMATOSIS CEREBRI WITH TOTAL SPINAL CORD EXTENSION

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INTRODUCTION: Although gliomatosis cerebri is no longer a pathological diagnostic name, it is still an important disease state as a gliomatosis cerebri growth pattern. Here we report a case of gliomatosis cerebri originating from the left cerebral hemisphere that developed whole spinal cord. CASE: A 10-year-old boy. He has a history of 13q-syndrome and left retinoblastoma. Left eye enucleation and chemotherapy (modified 98A1) have been performed. MRI in July 2017 showed no abnormalities, but in September 2018, he developed epilepsy. MRI revealed a gliomatosis cerebri that spreads extensively in the left cerebral hemisphere. Biopsy revealed anaplastic astrocytoma (MIB-1 LI; 22%, IDH1 / 2; WT, TERT C228T mutation positive) and IMRT (59.4Gy) and temozolomide (Stupp regimen) were performed in December 2018. In June 2019, neck pain developed. Head and neck MRI revealed that the tumor in the head increased lightly, and there was no suspicion of tumor growth in the brainstem, but the tumor progressed to the entire spinal cord. Therefore, radiation therapy was started from the lower brainstem that had not been irradiated last time to the entire spinal cord, and administration of bevacizumab was started. DISCUSSION: Based on the single cell origin theory, the left hemisphere tumor and spinal cord tumor should be continuous. Since gliomatosis cerebri is visualized on MRI only after the tumor volume has increased and edema has occurred, it may appear as if there is no tumor in between. The spinal cord MRI was not taken, so it is only speculation, but it seems that tumor cells had probably infiltrated the spinal cord from the beginning, and it seems that it gradually increased because it was not irradiated. Considering the possibility of remote invasion as in this case, it is necessary to consider taking MRI of spinal cord at the first occurrence.

BT-06

CENTRAL NERVOUS SYSTEM HEMANGIOBLASTOMA; DIFFERENCES IN CLINICAL PICTURE OF SPORADIC CASES AND VON-HIPPEL LINDAU DISEASE IN 184 CASES

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INTRODUCTION: Central nervous system hemangioblastoma (CNS HGB) is a rare neoplasm, which predominantly arise in the posterior fossa and spinal cord. The etiology is divided into sporadic and von-Hippel Lindau (VHL) disease. The difference in clinical picture of these 2 types of HGB and differentiation of treatment have not been extensively unraveled yet. METHODS: Retrospective analysis of consecutive, neurosurgically managed CNS HGB at Mayo Clinic, 1988–2018. RESULTS: 117 sporadic and 67 VHL HGBs were treated by Mayo Clinic. No significant difference in sex was observed. Compared with sporadic cases, VHL cases were younger (51.8 vs 36.0 years old, $p<0.0001$), had more frequent family history (0.0 vs 41.5 %, $p<0.0001$), and higher frequency of germline alteration (0.0 vs 84.2 %, $p<0.0001$). Regarding imaging findings, VHL cases had multiple lesions at presentation more frequently (3.4 vs 82.1 %, $p<0.0001$), it was more common for sporadic lesions to contain cysts (72.2 vs 51.0 %, $p=0.0004$), the solid portion rate in the entire lesion was larger in VHL lesions (60.2 vs 69.5 %, $p=0.02$), and the volume was larger in sporadic cases (15.1 vs 6.6 cc, $p<0.0001$). Regarding treatment, 131 and 123 surgeries were performed for sporadic and VHL cases, respectively, among which the indication of surgery was preventative in 8.4 and 47.3 %, respectively ($p<0.0001$). VHL cases had higher number of treatments per case in the follow-up (1.3 vs 2.1, $p<0.0001$). Recurrence-free survival of sporadic cases was significantly longer than that of VHL cases ($p=0.007$) and overall survival was longer in sporadic cases than VHL, but not significant ($p=0.07$). CONCLUSION: Clinical presentation and tumor appearance on imaging are highly dependent on the etiology. Differences in clinical manifestations require further study, but may reflect contrasting tumor biology that are tied to genetic differences.

BT-07

PATIENT DERIVED XENOGRAFT MODELS OF EPITHELIOID GLIOBLASTOMA AND THERAPEUTIC VULNERABILITY IN MOLECULAR TARGET THERAPY

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Epithelioid glioblastoma (E-GBM) predominantly arises at younger age and promotes dismal prognosis. Because of its rare etiology, pathological and genetical characterization of E-GBM remains elusive. Herein, we report unique patient-derived E-GBM xenograft (PDX) models from 3 E-GBM patients (2 $BRAF^{V600E}$ mutant and 1 $BRAF^{V600E}$ wild-type). Two BRAF mutant E-GBM cells (YMG62 and YMG89) were originated from adolescent and young adult patients and harbored TERT promoter mutation and CDKN2A homozygous deletion, while 1 $BRAF^{V600E}$ E-GBM cell (YMG64) was from elderly patient and had TERT wild-type. YMG62 and YMG89 could be propagated at multiple passage in vitro, while YMG64 could not be maintained. PDX models were established from YMG62, YMG89, and YMG64. All PDX tumors were preferentially disseminated and negative expression of GFAP, which were recapitulated to the patient characteristics. BRAF and MEK inhibitor mildly suppressed cell viability in vitro. Collectively, E-GBM PDX models recapitulate patient characteristics, which may be helpful to elucidate tumor biology and establish novel therapeutic target in E-GBM.

BT-09

CLINICAL AND MOLECULAR GENETIC FEATURE OF CEREBELLAR GLIOBLASTOMA

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