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 12 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 was 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 + CDGP + 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 59.2 ± 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 macrocephaly. Magnetic resonance images showed diffuse hypertensive 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 with medulloblastoma. As 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 3q-syndrome and left retinoblastoma. Left eye enucleation and chemotherapy (modified 98A1) have been performed. PET-MRI in December 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 11%, 22%, IDH1 / 2; WT, TERT C228T mutation positive) and IMRT (39.4Gy) 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 hemispheric 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 its growth 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 (CNHGB) is a rare neoplasm, which predominantly arises 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 (31.8 ± 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 31.0%, p=0.0004), the solid portion rate in the entire lesion was larger in VHL lesions (60.2 vs 6.9%, 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 4.9%, 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 genetic characterization of E-GBM remains elusive. Herein, we report unique patient-derived E-GBM xenograft (PDX) models from 3 E-GBM patients (2 BRAFV600E mutant and 1 BRAFV600E wild-type). Two BRAF mutant E-GBM cells (YM658 and YM839) were originated from adolescent and young adult patients and harbored TERT promoter mutation and CDKN2A homozygous deletion, while 1 BRAFV600E E-GBM cell (YM646) was from elderly patient and had TERT wild-type. YM658 and YM839 could be propagated at multiple passage in vitro, while YM646 could not be maintained. PDX models were established from YM658, YM839, and YM646. 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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Glioblastoma (GBM) predominantly affects patients in the sixth to seventh decade of life, with a median age of 63 years. Patients with glioblastoma (GBM) have a median survival of approximately 14 months after diagnosis. The molecular mechanisms underlying GBM development are complex and involve a combination of genetic and epigenetic alterations. Among the genetic alterations, mutations in the tumor protein p53 (TP53) gene are the most common, occurring in approximately 50% of GBM cases. Other frequently mutated genes include EGFR, IDH1, IDH2, and PIK3CA. In addition to genetic alterations, GBM is characterized by a high degree of heterogeneity, both intratumorally and interpatientally. This heterogeneity is thought to contribute to the complexity of GBM biology and treatment resistance. GBM is considered a highly aggressive tumor, and patients often experience rapid disease progression. Despite advances in treatment modalities, including surgery, radiation therapy, and chemotherapy, the overall survival for GBM remains low, and new therapeutic strategies are urgently needed. In this context, understanding the molecular features of GBM, particularly in young adult patients, is crucial for improving clinical outcomes and developing targeted therapies.
Giant cell tumor of bone is a rare and osteolytic neoplasm that predominantly affects the epiphyses in long bones of the extremities. They seldom occur in the skull, preferentially affecting the sphenoid and temporal bones. Most pathologically benign, and total removal by surgery was regarded as the first treatment, however, it was very difficult in skull lesion. In 2014 the molecular targeted drug anti-RANKL inhibitor was approved in Japan. We report a case in which an anti-RANKL inhibitor was administered to a skull base giant cell tumor that was difficult to remove completely. A 36-year-old man with a sudden right neck pain followed by dysphoria and dysphagia was referred to our hospital. Computed tomography showed 4.4 x 2.0 cm osteolytic lesion involving the right occipital bone and occipital condyle. Magnetic resonance imaging demonstrated an extensive soft-tissue mass occupying. Surgical biopsy was performed and the pathological diagnosis was giant cell tumor. Patient received the anti-RANKL inhibitor (Denosumab®). After 6 weeks, observation was observed, and neurological symptoms were improved after 12 weeks. Patient has been on good course for 5 years without recurrence and is still following-up.

CS-02

CLINICAL AND MOLECULAR ANALYSIS OF ASTROBLASTOMAS
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Astroblastoma is extremely rare brain tumor which mostly arise in cerebellum and cerebellar hemisphere of children and young adult. Limited data exists on its clinical feature and molecular analysis. We recently experienced two female patients with astroblastoma in the cerebrum.

Case 1 is three-year-old girl. She developed left hemiparesis. CT and MRI revealed large supratentorial mass with cystic component and calcification. Gross total removal was achieved. She is well without recurrence on MRI one year after surgery. Case 2 is 42-year-old lady. She developed partial seizure. CT and MRI revealed a mass with ring-enhancement in the left temporal lobe. Gross total removal was achieved under awake craniotomy. She is well without recurrence on MRI six months after surgery.

Pathologic examination of both patients showed pseudosarcomatous formation of tumor cells around vasculature. Molecular analysis revealed rearrangement of MN-1 in case 1 but not in case 2. Case 2 showed BRAF V600E mutation and loss of CDKN2A/B. Both patients received no adjuvant therapy.

Prognosis of astroblastoma varies and standard of treatment is not established. Gross total resection is associated with increased survival, but the role of adjuvant chemotherapy and radiation therapy are controversial. Advances in molecular analysis will lead to establish molecular classification and risk-adapted treatment strategy.

CS-03

LARGE CYSTIC INTRADURAL SCHWANNOMA IN CEREBRAL REGION: A CASE REPORT
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Schwannomas are the most common intradural extramedullary spinal tumors. However, they are usually solid tumors, and totally cystic changes are rare. Herein, we report a case of a 46-year-old male presenting with numbness of right limbs, right hemiplegia, and posterior neck pain for one year. MRI revealed a well-defined cystic long-segment, from C1 to C6, intradural extramedullary mass. The lesion showed hypointense on T1WI, hyperintense on T2WI, hyperintense on DWI, and it was marginally enhanced on the contrast image with Gd-DTPA. C1 laminectomy and hemi-laminectomy from C2 to C6 was performed for tumor resection. The tumor was found to be totally cystic and tesed with a jelly-like content. It was completely resected with the attachment of the C3 dorsal root. Histopathological examination confirmed it to be a schwannoma. The mechanism of cyst formation in schwannoma is considered as results of ischemic necrosis associated with tumor growth, or cystosis due to degeneration of Antoni-type B region. The long segment, totally cystic intradural cervical schwannoma is rare, but it should be included in the differential diagnosis of a cystic mass in the spinal region. It can be difficult to distinguish cystic spinal schwannomas from other cystic lesions like arachnoid cyst, epidermoid cyst, and neurinomatous cyst. Contrast enhanced MRI is useful by enhancing the margin of the tumor.

CS-04

INTEGRATED CLINICAL, HISTOPATHOLOGICAL, AND MOLECULAR DATA ANALYSIS OF 190 CENTRAL NERVOUS SYSTEM GERM CELL TUMORS FROM THE JGCT CONSORTIUM
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