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BACKGROUND: We integrated clinical, histopathological, and molecular data of central nervous system germ cell tumors to provide insights into their management. METHODS: Data from the Intracranial Germ Cell Tumor Genome Analysis Consortium were reviewed. A total of 190 cases were classified as primary GCTs based on central pathological reviews. RESULTS: All but one of the cases that were bifocal (neurohypophysis and pineal glands) and cases with multiple lesions including neurohypophysis or pineal gland were germinomas (34 of 35). Age was significantly higher in patients with germinoma than other histologies. Comparison between tumor marker and histopathological diagnoses showed that 18.2% of histopathologically diagnosed germinomas were marker-positive and 6.1% of non-germinomatous GCTs were marker-negative, suggesting a limitation in the utility of markers or histopathology alone using small specimens for diagnosis. Comparison between local and central histopathological diagnoses revealed a discordance of 12.7%. Discordance was significantly less frequent in biopsy cases, implying difficulty in detecting all histopathological components of heterogeneous GCTs. Germinomas at the typical sites (neurohypophysis or pineal gland) showed a better PFS than those at atypical sites (p=0.03). A molecular-clinical association study revealed frequent MAPK pathway mutations in males (51.4 vs 14.3 %, p=0.007), and PI3K/mTOR pathway mutations in basal ganglia cases (p=0.004). Basal ganglia cases also had frequent chromosomal losses. Some chromosomal aberrations (2q, 8q gain, 5q, 9p/q, 13q, 15q loss) showed potential prognostic significance.

CONCLUSIONS: These in-depth findings of this study regarding the clinical and molecular heterogeneity will increase our understanding of the pathogenesis of this enigmatic tumor.

CS-05

EXTRA-PARENCHYMAL (PERIPHERAL) ATYPICAL TERATOID / RHABDOID TUMORS

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AT/RT is a malignant embryonal tumor reported by Rorke in 1996. Authors reported first AT/RT in Japan in 1998. This tumor entity was included as new malignant embryonal tumor in WHO 2000, and tumors of Japanese patients has been reported more than 60 cases in the past. This AT/RT is a tumor in the brain parenchyma that a medulloblastoma and PNET and the possibility that it has been misdiagnosed have had pointed out. On the other hand, it is reported that there is the type that we should call peripheral AT/RT which rarely occurs in extra-parenchyma. We want to propose that there is such special tumor group. In the results, age: 6 infants were main (2nd - 14 years old after birth). tumor location: 6 sellar region, 3 civuses, 2 petrous bone or cerebellum and 2 conexities. treatment: duration of survival significantly improved all macroscopic tumor resection by the operation, but, in small pontine part AT/RT, an outcome tended to be poor. On the other hand, in AT/RT which occurred in the sellar region, all cases adult woman tended to have good prognosis. It is necessary for AT/RT (central AT/RT) in the brain to recognize that there is extra-parenchymal AT/RT (peripheral AT/RT) tumor which we reported this time which came to be recognized widely.

CS-06

A CASE OF GLIOBLASTOMA METASTATIC TO THE LUMBAR VERTEBRA

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BACKGROUND: Most cases of glioblastoma recur within one year even under the standard treatment of surgical resection, radiation therapy and chemotherapy. 60–70% of recurrences are local, and in rare cases of metastasis, most are within the CNS. Extradural metastasis is considered exceedingly rare. CASE REPORT: We present a 21-year-old man post total resection of right parietal lobe glioblastoma, diagnosed with lumber metastasis. He originally presented with impaired consciousness and left hemiplegia at the age of 20 and underwent gross total resection of the tumor. Pathology was IDH wild type, H3F3A K34R/V wild-type glioblastoma. Radiotherapy and adjuvant temozolomide per the Stupp regimen as well as infusion of bevacizumab were conducted. 6 months after the resection of tumor, the patient presented with severe back pain. Radiographic studies showed an osteolytic mass on the first lumbar vertebrae, and needle biopsy was consistent with glioblastoma. Posterior spinal fusion, internal decompression and radiotherapy were conducted to relieve the pain. At 3 months after the diagnosis of lumbar metastasis, he is currently treated with temozolomide and bevacizumab, without the enlarging of the tumor. DISCUSSION: As far as we investigated, there has been 30 cases of vertebral metastasis of glioblastoma reported in literature. Considering the biological obstacles that prevent glioblastomas from infiltrating outside of the CNS, it can be speculated that deposition of tumor cells into the blood stream or excision of the dura due to surgical interventions may attribute to extracranial metastasis. Due to the improvement of overall survival of glioblastoma, vertebral metastasis is suspected to be more common. Therefore, investigation of its risk factors and standardization of its treatment is necessary. CONCLUSION: We reported a case of lumbar metastasis of glioblastoma. Extradural metastasis of glioblastoma must be included in differential diagnoses in treating patients with glioblastoma.

CS-07

A CYSTIC LONG-SEGMENT CERVICAL SCHWANNOMA: 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 tensed 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 neurenteric cyst. Contrast enhanced MRI is useful by enhancing the margin of the tumor.

CS-08

A CASE OF CD 57 NEGATIVE OLFACTORY GROOVE SCHWANNOMA IN WHICH SCHWANN / 2 E AND SOX10 WAS USEFUL FOR DISTINGUISHING FROM OEC TUMOR: A CASE REPORT AND REVIEW OF THE LITERATURES

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INTRODUCTION: In the case of the differentiate between olfactory groove schwannomas(OGS) and olfactory ensheathing cell (OEC) tumors, CD57 which is the marker which is specific for Schwann cell is useful. We experienced a case of OGS that was negative for CD57. CASE PRESENTATION: This case was a 13-year-old girl. Medical history: She visited the pediatric department with a chief complaint of headache. On the magnetic resonance image (MRI), a tumorous lesion was found in the anterior skull base and was referred to our department. No dysosmia, visual impairment, or cafe au lait spots were observed. Past history: As a medical history, she has developed acute lymphocytic leukemia at the age of 1 and has achieved complete remission after chemotherapy. At that time, radiation treatment to the head was not performed. Neuroradiological findings: The tumor was strongly enhanced heterogeneously in Gadolinium (Gd) enhanced MRI and the angiography showed hypovascular. Progress after hospitalization: The tumor was clearly demarcated from the surrounding brain surface and adhered strongly to the cribriform plate. Eventually, all tumors were removed and the patient was discharged on the 10th postoperative day. Five years have passed since the operation, and no recurrence of the tumor has been confirmed by MRI. Pathological findings: Antoni A and Antoni B were seen by Hematoxylin & Eosin (H & E) staining. Immunostaining showed S-100 strong positive, Schwann / 2E and Sox10 positive, and CD57 negative. Discussion: In our case, CD57 (Leu7) was negative, but Schwann / 2E and Sox10 were positive, so OGS was diagnosed. CONCLUSION: We experienced a case of OGS that was negative for CD57 (Leu7) but positive for Schwann / 2E and Sox10. For pathological differentiation between OGS and the OEC tumor, Schwann / 2E and Sox10 immunostaining would also be necessary in addition to H & E stain and CD57 (Leu7).