suboccipital craniotomy was performed in the sitting position, a head-up surgery was performed using a 4K / 3D video microscope system (ORBEYE exoscope, Olympus) by the infratentorial supracerebellar approach (ITSCA). The bridging veins and precentral cerebellar vein were dissected to expose the posterior surface of the tumor, and internal decompression was performed. For the complication of air embolism, artificial cerebrospinal fluid was sprayed, and the bilateral internal jugular veins were compressed to confirm the inflow point. There was strong adhesion to the Rt vein of Rosenthal, and the site was removed intracapsularly. Finally, subtotal resection was performed with remaining the upper part of the tumor, a blind lesion behind the Vein of Galen. Vertical gaze palsy occurred after this operation, but it gradually improved over time. Tumor showed pathologically remarkable polymorphism, poor microvascular proliferation and necrosis, but mitotic figures 4–5 / 10HPF, MIB-1 index 10%, GFAP positive, no BRAF V600E mutation. There are few reports of PXA occurring in the pineal gland, and this case is the sixth case. It is also the first report for pineal tumors using ORBEYE through ITSCA in the sitting position.

Key words: PXA | pineal region | sitting position

CS-6

A CASE OF POORLY DIFFERENTIATED CHORDOMA WITH SYSTEMIC METASTASIS

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A case report: The patient was a 32-year-old man with diplopia. He was diagnosed as sphenoid sinusitis on MRI by a local doctor and visited an otolaryngologist. MRI showed extensive extension of neoplastic lesions from the clivus to the sphenoid sinus to the anterior ethmoid sinuses, bilateral cavernous sinuses, and the right medial and lateral pterygoid muscles. The right Lebiere's lymph node was enlarged and thought to be a metastatic site. Based on the rapid growth and extension of the tumor, the patient was referred to the Department of Otolaryngology at our hospital on suspicion of sinonasal carcinoma. The possibility of chordoma could not be denied, so the patient was referred to our department. The patient underwent a joint endoscopic extended transsphenoidal tumor resection. The pathological diagnosis showed mitotic and necrotic features, and the majority of the cells showed highly atypical components without mucous substrate. However, brachyury, a marker for chordoma, was diffusely positive, and there was loss of INI1 (SMARCB1) expression. The final diagnosis was poorly differentiated chordoma. Postoperatively, the tumor in the right cavernous sinus grew rapidly, and the right eye became blind due to obstruction of the superior ophthalmic vein. The patient was treated with Gamma Knife as soon as possible in the hope of local control by high-dose irradiation, and after a total of three irradiations, the residual tumor shrank markedly and symptoms improved, but systemic metastasis occurred in a short period of time and the patient died. The number of cases of poorly differentiated chordoma has been reported rarely (more than 50), and it is more common in children and even rarer in adults. We report this case with a review of the literature.

Key words: poorly differentiated chordoma | systemic metastasis | poor prognosis

CS-7

A CASE OF LYMPHOMATOID GRANULOMATOSIS WITH SKIN, LUNG, AND INTRACRANIAL LESIONS DUE TO MULTICENTRIC DEVELOPMENT

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Introduction: LYG is very rare tumor and composed of large EB-positive B cells and reactive T cells. In this study, we experienced a case of LYG with multiple intracranial, cutaneous, and pulmonary masses. We report the pathogenesis and pathophysiology of LYG, including a discussion of the literature. case: A 69-year-old female presented with a growing lump in her lower back that had been present for several years. Six months later, she was found to have multiple masses in her lungs and intracranial region and underwent surgical removal for diagnostic purposes. Intraoperative findings: The tumor was substantial, reddish to grayish-white in color, and the margins of the tumor were whitish and hard, with some areas that could not be detached. Pathological findings: There were no atypical lymphocytes, and a small number of EBER-positive cells were observed. IgVH PCR: IgVH PCR was performed on the skin lesions and intracranial lesions, and bands of different sizes were detected, suggesting that the IgVH clone was present in the polyclonal region. Finally, we diagnosed LYG grade 1. discussion: EB-associated lymphoproliferative disease can lead to polyclonal reactive growth or monoclonal neoplastic growth depending on the balance between morphology and host immunity. The results of IgVH PCR suggest that the skin lesions did not cause multiple metastases, but rather that the enlargement of the skin lesions triggered intracranial and pulmonary lesions in an allo-centric manner. The results of IgVH PCR suggested that the skin lesions did not cause multiple metastases, but rather that the skin lesions grew to cause intracranial and pulmonary involvement in an other-centric manner.

Key words: Lymphmatoid granulomatosis | EBER | IgVH clonality

CLINICAL OTHERS (COT)

COT-1

CLINICAL QUESTIONS AND ANSWERS ABOUT GLIOMA-RELATED EPILEPSY (GRE): REAL-WORLD DATA IN WAKAYAMA MEDICAL UNIVERSITY HOSPITAL

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Introduction: In glioma patients, epilepsy not infrequently occurred and anti-epileptic drugs (AEDs) are commonly used. In this study, we revealed the real-world data on clinical practice of glioma-related epilepsy in Wakayama Medical University Hospital (WMUH). Methods. We collected clinical and molecular data of glioma patients operated at WMUH from January 1996 to December 2020 and analyzed the data to answer clinical questions as follows: 1) location/histology related GRE, 2) molecular features related GRE, 3) prophylactic AEDs and postoperative seizure, 4) tumor progression and convulsion, 5) GRE and survival. Results. Fifty-five of 113 glioma patients (49%) presented with seizure. CQ1. In tumors located at frontal, temporal and parietal lobe, the occurrence rate of GRE was 27/39 (69%), 13/19 (69%) and 9/14 (64%), respectively. Patients with glioblastoma, astrocytic tumors and oligodendroglial tumors presented with GRE at the rate of 26/54 (48%), 14/30 (47%) and 12/13 (92%), respectively. CQ2. GRE occurred in tumors with IDH mutated (16 cases, 29%), TERT mutated (32 cases, 58%) and MGMT methylated (32 cases, 58%). CQ3. Seizure in peri- or postoperative period occurred in 14 cases (12%); 4 cases in AED(+) group (4/29, 14%) and 10 cases in AED(-) group (10/84, 12%). CQ4. Tumor progression became apparent at the time of seizure in 12 cases (12/55, 22%). CQ5. According to the prognostic IDH/TERT classification of diffuse glioma cases (n = 94), overall survival (OS) times of GRE(+) cases tended to be longer than that of GRE(-) ones, especially in IDH wildtype/TERT mutated group (22.7 months vs. 8.3 months, p = 0.0397). Conclusion. GRE is likely associated with specific clinical and molecular features. Seizure in glioma patients can occur in specific situation regardless of the use of AEDs. Possible better prognosis of GRE(+) cases requires further investigation.

Key words: Epilepsy | Glioma | Clinical questions

COT-3

EXOSOMAL MICRORNA EXPRESSION SIGNATURE IN BLOOD AND CEREBROSPINAL FLUID OF GLIOBLASTOMA PATIENTS

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Analysis of exosomes derived from plasma or cerebrospinal fluid (CSF) has emerged as a promising biomarker platform for therapeutic monitoring in glioblastoma patients. However, the contents of the various subpopulations of exosomes in these clinical specimens remain poorly defined. Here we characterize the relative abundance of miRNA species in exosomes derived from the plasma and CSF of glioblastoma patients. To this end, we first employed