Oral presentation 1: TUMOR 1

A-0009

Frameless robot assisted stereotactic biopsy: an effective and minimally invasive technique for pediatric diffuse intrinsic pontine gliomas

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Introduction : Diffuse intrinsic pontine gliomas (DIPGs) are prone to high surgical risks, and they could even lead to death due to their specific sites. To determine the value of frameless robot-assisted stereotactic biopsies of DIPGs, when compared it with microsurgical biopsies.

Material & Methods : We conducted a retrospective study of 71 pediatric patients who underwent biopsies from January 2016 to January 2021. (i) group 1: microsurgical biopsies, and (ii) group 2: frameless robot-assisted stereotactic biopsies. Demographic information, neuroimaging characteristics, pathological diagnoses, operation time, postoperative intensive care unit (ICU)stay time, postoperative hospitalization time, complications, cost, and perioperative mortality rate (POMR) were collected for analyses.

Results : 32 Cases underwent microsurgical biopsies (group 1) and 39 cases underwent frameless robot-assisted stereotactic biopsies (group 2). All cases were accurately diagnosed after surgery. There was no significant difference in gender, age, symptom times and tumor volumes between the two groups ($p > 0.05$); operation time, postoperative ICU, stay time and postoperative hospitalization time were longer in group 1 than in group 2 ($p < 0.001$); the intraoperative bleeding volumes and cost were higher in group 1 than in group 2 ($p< 0.001$). Group 1 patients required more perioperative blood transfusion than group 2 ($p = 0.001$), and the new neurological impairments were more frequent in group 1 than in group 2 ($p = 0.003$). The POMR was 9.38% (3/32) in group 1 and 0 in group 2 ($p = 0.087$).

Conclusions : Frameless robot-assisted stereotactic biopsy was an effective and minimally invasive technique for pediatric DIPGs.

Oral presentation 1: TUMOR 1

A-0065

The risk and benefit of early surgery for infants with congenital brain tumors

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Introduction : Infant brain tumors are known to have different biological features from other pediatric brain tumors. The present study aimed to evaluate the risk of early surgery in congenital brain tumors and analyze the clinical outcomes of infant brain tumors.

Material & Methods : We reviewed the clinical presentations, imaging features, intraoperative findings, and histopathological findings of infant brain tumors at a single institution from January 2008 to June 2023. Outcomes were measured as intraoperative CPR (N = 4) to evaluate the risk of early surgery in congenital brain tumors. Recurrence and overall survival were analyzed in infant brain tumors (N = 69).

Results : Intraoperative CPR occurred in low age and body weight in patients with congenital brain tumors. Cut-off values of age and body weight were found to be 1.3 months and 5.2 kg (AUC of 96.9 (p<0.001) and 95.8 (p<0.001)) for no CPR. Three patients (30%) had intraoperative CPR in the early surgery group, and intraoperative CPR occurred in one patient (7.14%) in the delayed surgery group, whose tumor was excessively enlarged. Younger age at diagnosis (<3 months of age, HR: 10.0, 95% CI: 2.12-46.7, p = 0.004) and leptomeningeal seeding (LMS, HR: 27.0, 95% CI: 3.7-197.0, p=0.001) were significant independent risk factors for high mortality in infant brain tumors.

Conclusions : We suggest delaying surgery until the patient reaches 1.3 months of age and weighs over 5.2 kg with short-term imaging follow-up unless tumors grow rapidly in congenital brain tumors. Younger ages and LMS are independent risk factors for high mortality in infant brain tumors.

Oral presentation 1: TUMOR 1

A-0093

Multimodality in Pediatric Brain Tumor Surgery -endoscope, exoscope, & navigation-

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Introduction : Various types of end-or exoscopic surgery are performed in the field of pediatric neurosurgery. In addition, safer and more reliable surgery utilizing various modalities is required in cases of brain tumors and hydrocephalus. In this report, we describe our experience using multimodality in pediatric brain tumor surgery at our hospital.

Material & Methods : From May 2020 to the present, 27 pediatric brain tumor surgery cases under 20 years of age were included in the study. Sixteen were male and 11 were female, ranging in age from 0 to 20 years (mean 11.8 years). Stealth Station S7 and S8 (Medtronic) navigation systems were used. Endoscopes were Videoscope (Olympus) and 4KEndoArm (Sony& Olympus). Exoscope was ORBEYE (Olympus).

Results : Tumors included 6 gliomas, 4 schwannomas, 4 germ cell tumors, 2 meningiomas, 1 ependymoma, 1 pineoblastoma, 1 rhabdomyosarcoma, and 8 others. Endoscopes were used in 8 cases, and navigation systems were used in 27 cases. Endoscopic biopsy was performed in 4 cases, ETV in 5 cases, and eTSS in 2 cases. Exoscopes were used in 2 cases. The use of intraoperative navigation helped determine the extent of craniotomy and assist in the precise approach to the lesion. The electromagnetic navigation was particularly useful in confirming the location of the tumor in the ventricles and determining the biopsy site.

Conclusions : In the surgical field of pediatric brain tumors, navigation and end-or exoscope made it possible to perform safer and more reliable surgery. Further safe surgery is expected to be possible with the introduction of robotics in the future.

Oral presentation 1: TUMOR 1

A-0121

ANALYZING THE EXTENT OF TUMOR RESECTION IN INTRAOPERATIVE ULTRASOUND GUIDED SURGERIES FOR PEDIATRIC POSTERIOR FOSSA TUMORS : A SINGLE CENTRE EXPERIENCE

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Introduction : Malignant central nervous system (CNS) tumors are the second most common pediatric malignancy, following hematological malignancies. The extent of tumor resection plays a crucial role in the management of CNS tumors. Intraoperative ultrasound (IOUS) has emerged as a valuable neuroimaging tool for real-time surgical guidance, offering accessibility, minimal risk, minimal additional operative time, and lower economic investment. The objective of this study was to analyze the extent of tumor resection in intraoperative ultrasound-guided surgeries with early post operative MRI.

Material & Methods : We conducted a prospective observational study at Hospital Tunku Azizah, Kuala Lumpur, including patients under 12 years of age who were admitted between December 1, 2019, and November 30, 2021, with a preoperative MRI diagnosis of posterior fossa tumors. Intraoperative ultrasound (IOUS) was performed during the surgical procedure, and postoperative MRI scans were conducted within 24-72 hours after surgery.

Results : A total of 13 pediatric patients who met the inclusion criteria were analyzed. Concordance analysis between IOUS evaluation and MRI scans demonstrated good agreement. Multivariate logistic regression analysis was performed to identify significant factors of discordance, but none of the variables were found to be significant.

Conclusions : Intraoperative ultrasound-guided surgeries can aid in analyzing the extent of tumor resection for pediatric posterior fossa tumors. The findings of this study highlight the potential of IOUS as a real-time navigation tool to improve surgical outcomes in the management of pediatric CNS tumors.

Oral presentation 1: TUMOR 1

A-0151

Neuroendoscopic surgery for deep brain tumors using a small-diameter ultrasonic aspirator

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Introduction : As with cancers of other organs, minimally invasive surgery for brain tumors is increasingly being performed using endoscopy. In particular, endoscopic surgery for brain tumors requires careful use of tools because the surgical field is narrow. The small-diameter ultrasonic aspirator we currently use is useful, and we will introduce neuroendoscopic surgery for deep brain tumors using the small-diameter ultrasonic aspirator.

Material & Methods : Methods : A port is inserted from the brain surface to the tumor using navigation through a linear incision and small craniotomy. The route to reach this time is determined by considering safe and maximum extraction from the non-eloquent area. ViewSite 21x15mm ports are often used to secure the endoscope and perform two-handed surgery. Small tumors may be removed with tumor forceps, but larger tumors are suctioned out using a thin ultrasonic aspirator. This small-diameter ultrasonic aspirator can also be used for transnasal surgery. Bleeding is stopped using a small diameter bipolar device. Patients : Infantile huge bilateral intraventrivular AT/RT case, pediatric right thalamic GBM case, skull base highly calcified chondromyxoid fibroma case.

Results : In all of the cases presented, we were able to perform subtotal resection or higher.

Conclusions : The small-diameter ultrasonic aspirator has a TissueSelet function that selectively disrupts tissue while leaving blood vessels intact, making it possible to perform ultrasonic aspiration of tumors while preserving deep normal blood vessels. Even in highly calcified tumors, it is possible to crush calcified lesions by maximizing the output. Tumor aspiration using a small-diameter ultrasonic aspirator is considered to be a useful surgical technique.

Oral presentation 1: TUMOR 1

A-0197

Two-staged endoscopic surgery for craniopharyngioma with obstructive hydrocephalus

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Introduction : Pediatric craniopharyngiomas are difficult to treat because radical tumor resection can lead to high postoperative complication rate, whereas incomplete resection can lead to high tumor recurrence rate. We present herein a girl who underwent two-staged endoscopic surgery for craniopharyngioma with acute obstructive hydrocephalus.

Material & Methods : A 9-year-old girl presented gradually worsening headache, polyuria, memory disturbance and visual disturbance, and visited to nearby hospital. Head computed tomography showed a suprasellar tumor with calcification and severe obstructive hydrocephalus. She was referred to our hospital for treatment with preoperative diagnosis of craniopharyngioma. Magnetic resonance imaging showed a cystic suprasellar lesion, filling the third ventricle.

Results : Emergency cyst fenestration was performed via right lateral ventricle with a flexible endoscope, and cystic components of the tumor were radically fenestrated. Her preoperative symptoms were released just after the surgery. One month after the surgery, tumor resection was performed via endoscopic extended trans-sphenoidal approach. The tumor was completely removed with preservation of the pituitary stalk. The postoperative course was uneventful. Anterior pituitary functions were partially deteriorated, therefore hormonal replacement therapy has been continued. No tumor recurrence was observed at the last follow-up.

Conclusions : Endoscopic cyst fenestration is essential for craniopharyngioma with acute obstructive hydrocephalus not only to release the life-threatening comorbidity, but also to induce intermediate-term tumor shrinkage by natural intracranial pressure. Two-staged endoscopic surgery can be one of the most effective treatment options for craniopharyngioma with obstructive hydrocephalus.

Oral presentation 1: TUMOR 1

A-0196

Usefullness of 4K3D-exoscopic removal for pediatric fourth ventricular tumors

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Introduction : Pediatric brain tumors often occur in the fourth ventricle, and the standard approach of surgical removal is considered to be the cerebellomedullary fissure approach, but this approach and concorde landing position is tough to both patients and the operators. We discuss the safeness and effectiveness of 4K3D-exoscopic surgery for removal of fourth ventricular tumors in comparison with microsurgery, based on experience in our institution.

Material & Methods : The cerebellomedullary fissure approach for pediatric fourth ventricular tumors was performed in 16 cases at our hospital between August 2015 and January 2022. Cases with lesions including invasion into the cerebellopontine angle were excluded. 9 medulloblastoma cases, 6 ependymoma cases, and a case of AT/RT were enrolled. Eight cases (group A) were removed by 4K3D-exoscope, and others (group B) were removed using a microscope. There were no apparent statistical differences between the two groups in terms of age, maximum tumor diameter, tumor volume, hydrocephalus, and lateral extension.

Results : Surgery using microscopy or 4K3D-exoscopy was successfully completed in all cases. No significant differences were observed between the two groups in terms of blood loss (group A average: 305 ml, group B average: 258 ml), surgical time (group A average: 587 minutes, group B average: 535 minutes) and the extent of removal. The usefulness of 4K3D-exoscopic surgery was more exellent in terms of the size of the working space, the brightness of intraoperative fluorescence diagnosis, and the burden on the surgeon during surgery.

Conclusions : 4K3D endoscopic removal for the fourth ventricular tumors can be safe and useful.

Oral presentation 1: TUMOR 1

A-0182

Preoperative radiological diagnosis of pediatric posterior fossa tumors

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Introduction : In pediatric primary posterior fossa tumors such as medulloblastoma, ependymoma, and pilocytic astrocytoma, accurate preoperative diagnosis through radiological imaging is important for determining treatment strategies. The aim of this study is to compare the diagnostic accuracy between our preoperative diagnosis and the retrospective diagnostic algorithm, and to make a better algorithm.

Material & Methods : We conducted a retrospective analysis of <18-year-old patients with primary posterior fossa tumors who underwent surgery and received pathological diagnoses at our institution between April 2013 and August 2023. Cases with brain stem tumors and insufficient MRI data were excluded. We investigated the preoperative imaging diagnosis made by neurosurgeons and radiologists, the retrospective diagnosis by reported algorithm based on apparent diffusion coefficient, contrast enhancement, and tumor localization, and the pathological diagnosis.

Results : A total of 37 cases were included, with mean age was 8.1±4.5 years, and 11 patients were female. The concordance rate between our preoperative diagnosis and retrospective diagnostic algorithm was 81.1%. Based on the pathological results, the accuracy of our preoperative diagnosis and retrospective diagnostic algorithm were 86.5% and 81.1%, respectively. In addition to the algorithm's criteria, we used cerebral blood volume to distinguished pilocytic astrocytoma from hemangioblastoma, and age to distinguished medulloblastoma from AT/RT. With the inclusion of these, diagnostic accuracy improved to 89.2%.

Conclusions : Improving the accuracy of preoperative imaging diagnosis aid in decision-making during surgery and treatment planning. The diagnostic algorithm also serves as a communication tool when receiving referred patients from other facilities. Further development of the radiological diagnosis including molecular characteristics is desired.

Oral presentation 1: TUMOR 1

A-0184

Is surgical resection the first line for left hippocampal tumor in pediatric case? : a case report and literature review

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Introduction : Surgical treatment for mesial temporal lobe epilepsy is standard treatment and has been established in its efficacy and safety. However, the hippocampal tumor is a quite rare entity and there are controversies about treatment options. Herein, we present a pediatric case of a left hippocampal tumor treated with surgical resection.

Material & Methods : A right-handed 8-year-old healthy boy was transferred to a hospital with bilateral tonic-clonic seizure. CT scan showed a calcified mass at the left hippocampus, and the lesion showed high T2 and FLAIR signals with less enhancement on MRI. The tumor was located in the left hippocampus and parahippocampal gyrus, and 24 x 50 mm in size. Physical and neurological examination were normal except for focal epilepsy which has been well controlled using antiseizure medication.

Results : "Selective" tumor resection through the transsylvian approach was planned because there was no lesion at the lateral temporal lobe. The tumor was grayish and elastic hard. Lower-grade glioma was suspected in intraoperative rapid diagnosis. Near total removal was achieved except for the posterior side of the hippocampal tail. Currently, a histopathological diagnosis has not been finished yet. Postoperatively, there were no neurological deficits including memory disturbance and obvious visual field defects.

Conclusions : We present a pediatric case with a hippocampal tumor successfully treated with surgical treatment. Surgical treatment is invasive and challenging with a potential risk of memory disturbance, however, we believe the maximum tumor resection can improve progression-free and overall survival in pediatric patients with hippocampal lower-grade glioma.

Oral presentation 2: TUMOR 2

A-0101

MICROSURGICAL RESECTION FOR PEDIATRIC PINEAL REGION TUMORS: EXPERIENCES FROM VIETNAM

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Introduction : The purpose of this study is to evaluate the role of microsurgery for pineal region tumors at Children's Hospital 2, Southern Vietnam

Material & Methods : This retrospective case-series of 45 consecutive patients were diagnosed with pineal region tumors admitted at Children's hospital 2 from April 2010 to April 2023

Results : All three surgical approaches included transcallosal, occipito-transtentorial and infratentorial-supracerebellar routes were used for the pineal region lesions. Total and nearly total resection wereachieved in 85%. The transcallosal and occipito-transtentorial approaches were used mostly in 89%. The ratio of motality in 5.5% and morbidity in 16% of all patients. One patient had temporary vision loss and two others had minor subdural CSF effusion after resection of tumours without requiring any further procedures.

Conclusions : The multimodalities of treatments for pineal region tumors are recommended commonly. The microsurgical resection of tumours is too complex and risky but only neccessary in 2/3 of all cases. Numerous surgical approaches are noticed worldwide. The neurosurgeons can use one of them according to some specific categories and belonging to their own experiences.

Oral presentation 2: TUMOR 2

A-0123

The diagnostic accuracy of intracranial germinomas: tumor markers versus tumor biopsy

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Introduction : Germinoma treatment does not typically rely on surgical resection, emphasizing the importance of accurate diagnosis before chemotherapy and radiotherapy. This study compares the diagnostic accuracy between tumor markers and tumor biopsy.

Material & Methods : The medical record of 117 patients with intracranial germ cell tumor between 1987 and 2022 were reviewed. Patients diagnosed as germinoma by pathological examination were included. The pathology reports and serum/cerebrospinal fluid tumor markers, including alpha fetoprotein (AFP) and beta human chorionic gonadotropin (beta-HCG), and outcome were collected in these patients for analysis.

Results : Seventy-one cases, aged 3 to 64 years were enrolled. Twenty-four cases had elevated beta-HCG, with 17 between 5 to 50 IU/L, 2 between 50 to 100 IU/L, and 5 over 100 IU/L. Histologically, syncytiotrophoblastic cells were found in five of the 71 cases. One of the 5 cases with initial beta-HCG level of 38.3 IU/L had recurrence as embryonal carcinoma and yolk sac tumor 10 months later. Two of the 5 cases with beta-HCG level of 2856 and 26110 IU/L, respectively, also had concomitant elevation of AFP, had early recurrence and poor outcomes. Metastases and beta-HCG over 100 IU/L with elevated AFP at diagnosis were associated with poor outcomes.

Conclusions : Tumor markers may provide a more accurate diagnosis than histological examination, even when obtained through craniotomy. Sampling errors during tumor biopsy underscores the need to consider tumor markers in treatment planning to enhance clinical outcomes of patients with intracranial germinoma.

Oral presentation 2: TUMOR 2

A-0113

Long-term outcome of central nervous system germ cell tumors: Reduced-dose radiotherapy and intensified chemotherapy improves clinical outcome and quality of life of long-term survivors

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Introduction : Chemoradiotherapy is required for treatment against central nervous system germ cell tumors (CNSGCT), but optimal treatment intensity should be selected from the viewpoint of both improvement of clinical outcome and avoidance of late complications. We introduced a protocol with reduced-dose radiotherapy and intensified chemotherapy for CNSGCT. We retrospectively analysed the prognosis and long-term outcome of late complications, enrollment and employment, as indicators of quality of life (QOL).

Material & Methods : 38 children and young adults with CNSGCTs treated in our institution from 1997 to 2013 were enrolled in this study. They consisted of 26 germinomas including HCG-producing cases and 12 non-germinomatous GCTs (NGGCT). Local irradiation was selected for all patients, and the dose of irradiation was 23.4-54 Gy. The whole-brain irradiation was made in patients who had intracranial dissemination. For NGGCT, high-dose chemotherapy and peripheral blood stem cell transplantation (PBSCT) were introduced. Second-look surgeries were performed for cases with residual tumors after induction chemotherapies.

Results : In germinoma group and NGGCT group, 10-year progression-free survival was 86% and 84%, 10-year overall survival was 93% and 91%, respectively. About late complications, endocrinological replacement (39%), cerebrovascular disease such as cavernous hemangioma and arterial stenosis (18%), secondary neoplasm (2.6%) were observed. Enrollment and return to school rate was 92% and employment rate was 89%, which were influenced by hemipararesis associated with basal ganglia lesion, intractable epilepsy and whole-brain irradiation.

Conclusions : Reduced-dose radiotherapy and intensified chemotherapy for CNSGCT, especially NGGCT, improved the clinical outcome and QOL of long-term survivors, suppressing late complications. Further comprehensive follow-up and analysis are needed.

Oral presentation 2: TUMOR 2

A-0028

Survival and malignant transformation of pineal parenchymal tumors: A 30-year retrospective analysis in a single-institution

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Introduction : This study aims to elucidate clinical features, therapeutic strategies, and prognosis of pineal parenchymal tumors (PPT) by analyzing a 30-year dataset of a single institution.

Material & Methods : We reviewed data from 43 patients of all ages who were diagnosed with PPT at Seoul National University Hospital between 1990 and 2020. We performed survival analyses and assessed prognostic factors.

Results : The cohort included 10 patients with pineocytoma (PC), 13 with pineal parenchymal tumor of intermediate differentiation (PPTID), and 20 with pineoblastoma (PB). Most patients presented with hydrocephalus at diagnosis. Most patients underwent an endoscopic third ventriculostomy and biopsy, with some undergoing additional resection after diagnosis confirmation. Radiotherapy was administered with a high prevalence of gamma knife radiosurgery for PC and PPTID, and craniospinal irradiation for PB. Chemotherapy was essential in the treatment of grade 3 PPTID and PB. The 5-year progression-free survival rates for PC, grade 2 PPTID, grade 3 PPTID, and PB were 100%, 83.3%, 0%, and 40%, respectively, and the 5-year overall survival rates were 100%, 100%, 40%, and 55%, respectively. High-grade tumor histology was associated with lower survival rates. Significant prognostic factors varied among tumor types, with WHO grade and leptomeningeal seeding (LMS) for PPTID, and the extent of resection and LMS for PB. Three patients experienced malignant transformations.

Conclusions : This study underscores the prognostic significance of WHO grades in PPT. It is necessary to provide specific treatment according to tumor grade. Grade 3 PPTID showed a poor prognosis. Potential LMS and malignant transformations necessitate aggressive multimodal treatment and close-interval screening.

Oral presentation 2: TUMOR 2

A-0039

A case series and literature review on 98 pediatric patients of germ cell tumor developing growing teratoma syndrome

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Introduction : Malignant germ cell tumors (MGCTs) can develop either extrac- ranially or intracranially. Growing teratoma syndrome (GTS) may develop in these patients following chemotherapy. Reports on the clinical characteristics and outcomes of GTS in children with MGCTs are limited.

Material & Methods : We retrospectively collected the data, including the clinical character- istics and outcomes of five patients in our series and 93 pediatric patients selected through a literature review of MGCTs. This study aimed to analyze survival out- comes and risk factors for subsequent events in pediatric patients with MGCTs developing GTS.

Results : The sex ratio was 1.09 (male/female). In total, 52 patients (53.1%) had intracranial MGCTs. Compared with patients with extracranial GCTs, those with intracranial GCTs were younger, predominantly boys, had shorter intervals be- tween MGCT and GTS, and had GTS mostly occurring over the initial site (all $p<0.001$). Ninety-five patients (96.9%) were alive. However, GTS recurrence ($n=14$), GTS progression ($n=9$), and MGCT recurrence ($n=19$) caused a sub- stantial decrease in event-free survival (EFS). Multivariate analyses showed that the only significant risk factors for these events were incomplete GTS resection and different locations of GCT and GTS. Patients without any risk had a 5-year EFS of 78.8% ± 7.8%, whereas those with either risk had 41.7% ± 10.2% ($p < 0.001$).

Conclusions : For patients with high-risk features, every effort should be made to closely monitor, completely remove, and pathologically prove any newly de- veloped mass to guide relevant treatment. Further studies incorporating the risk factors into treatment strategies may be required to optimize adjuvant therapy.

Oral presentation 2: TUMOR 2

A-0128

Treatment standard and controversies on the pediatric intracranial NGGCT

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Introduction : Widespread controversy surrounds the standardized treatment of pediatric intracranial NGGCT (non-germinomatous germ cell tumors), with significant limitations observed in the existing cohort studies. Additionally, there is a lack of corresponding research in the high-incidence Asian region. To address this gap, we conducted a single-center retrospective study to explore viable treatment approaches.

Material & Methods : We conducted a retrospective analysis of 49 cases of pediatric intracranial NGGCT over the past 3 years. We employed Kaplan-Meier survival analysis to assess the three-year overall survival rate (OS) and progression-free survival rate (PFS), which were then compared with publicly available data.

Results : Fifty-one percent of patients underwent direct surgery followed by adjuvant radiotherapy and chemotherapy. Forty-nine percent of patients initially received chemotherapy, followed by surgical intervention after a two-cycle assessment (wherein 10% experienced tumor reduction, and 90% showed no tumor reduction). Subsequent to surgery, routine postoperative radiotherapy and chemotherapy were administered. The complete response rate after surgery in our cohort was 100%, whereas achieving a complete response rate solely through chemotherapy was 0%. The three-year OS rate was 86.3%, and the PFS rate was 82.1%.

Conclusions : Surgical intervention is nearly always necessary for NGGCT patients, with the timing of surgery being crucial. For NGGCT cases characterized by smaller tumor size, greater surgical feasibility, and the presence of teratoma components, we recommend direct surgical excision followed by adjuvant radiotherapy and chemotherapy. This approach can significantly enhance treatment outcomes and efficiency.

Oral presentation 2: TUMOR 2

A-0185

A retrospective analysis of clinical course of pediatric ependymoma cases in a single institute

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Introduction : Maximum tumor removal is the most useful treatment for ependymoma, however, standard strategy of adjuvant therapy has not been established yet, because the number of cases is limited. Therefore, it is quite useful to share clinical course of experienced cases.

Material & Methods : We reviewed the clinical course of 12 consecutive cases of pediatric ependymoma treated at our institute, retrospectively.

Results : The median age was 2.5 (0-15) years. 10 cases had posterior fossa ependymoma and 2 had supratentorial ependymoma. Gross total resection (GTR) was achieved in 9 cases (75%). 5 cases were diagnosed with ependymoma, Grade II, and 7 with anaplastic ependymoma, Grade III (WHO classification in 2007 and 2016). Post-operative adjuvant therapy was performed in 8 cases. Among 6 relapsed cases, 4 revealed local recurrence and 2 did dissemination. 2- and 5-year progression-free survival (PFS) rates were both 60%, and 2- and 5-year overall survival rates were 100% and 66.6%, respectively. For PFS, there was a better trend for GTR (HR = 0.18, 95% CI: 0.025 - 1.28) and supratentorial lesion (HR = 0.18, 95% CI: 0.030 - 1.11), but no difference for WHO grade (HR = 1.022, 95% CI: 0.20 - 5.24).

Conclusions : As previous studies revealed, WHO grade did not reflect prognosis. The latest WHO classification described that Posterior Fossa group A (PFA) ependymoma reveals unfavorable prognosis compared with PFB and supratentorial ependymomas. In near future, determination of treatment strategy may be needed for each subgroup. We also compare the clinical course between PFA, PFB and supratentorial tumors in our analyzed cases.

Oral presentation 2: TUMOR 2

A-0195

Multidisciplinary treatment in ependymoma

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Introduction : 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.

Material & 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 ($p = 0.02$).

Conclusions : Even if it is not GTR or NTR at the first operation, improvement of OS is expected by total excision after chemotherapy.

Oral presentation 2: TUMOR 2

A-0112

Direct evidence of the development of a myxopapillary ependymoma of the filum terminale into a cauda equina tumor and its clinical implication

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Introduction : Myxopapillary ependymoma (MPE) is a variant of ependymomas with predominant location at filum terminale (FT), conus medullaris, and cauda equina. MPEs are assumed to arise from ependymocytes normally present in conus medullaris or ectopically in FT. Our single case aims to present direct evidence of development of a MPE of FT into a huge cauda equina ependymoma.

Material & Methods : A 17-year-old girl presented with back pain for 3 months. Magnetic resonance imaging of lumbosacral spine showed a cauda equina tumor from middle L1 to L4 levels with a seeding at L5 region. There's no other neuraxis lesions. The surgical and pathological findings of the case and our proposition will be presented.

Results : Upon durotomy, nerves of cauda equina were all encased by the encapsulated tumor, which compressed conus medullaris without involvement of it. After peeling off the tumor from each nerve, an enlarged FT was noted, which was en bloc removed. The pathology showed a MPE. The central canal of FT was filled with tumor cells though both ends were free. Some tumors penetrated the filum and connected with the main tumor in cauda equina space. The tumor at L5 level was discrete from the main tumor. The CSF cytology was negative for tumor cells. After total excision, there was no recurrence under 2-year follow-up.

Conclusions : This case explained the pathogenesis of ependymomas of cauda equina, where there is no ependymocyte. The only "seeding" at the caudal portion of thecal sac further supports the tumor came from cells splitted from the FT ependymoma.

Oral presentation 3: TUMOR 3

A-0148

Utility of cerebrospinal fluid cell free DNA analysis for the diagnosis of H3K27-altered diffuse midline gliomas

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Introduction : Detection of Histone H3K27M mutations is critical for the diagnosis of diffuse midline gliomas H3K27-altered. However, especially when the tumor is located in the brain stem, it is not always easy to perform a biopsy. Therefore, we assessed the feasibility and utility of H3K27M mutation detection from cerebrospinal fluid (CSF) cell free DNA (cfDNA).

Material & Methods : CSF was obtained from 18 clinically or histologically diagnosed diffuse midline glioma patients between , 2019 and January, 2023 at Niigata University Hospital. CSF cfDNA was extracted and *H3F3A* K27M mutations were detected by droplet digital PCR.

Results : Median patient age was 19 (range 5-75) and 7 (39%) patients were female. CSF was obtained by lumbar puncture in 10 patients, intraventricular route during or after surgery in 7 patients and from a cyst in 1 patient. H3K27M-mutant droplets were detected from 11 (61%) patients, including more than 3 droplets in 3 (33%) patients. Median variant allele frequency was 7.5% (range 0-87.5%).

Conclusions : Detection of H3K27M mutant droplets from CSF cfDNA is feasible in diffuse midline gliomas. Further efforts to improve diagnostic accuracy is necessary.

Oral presentation 3: TUMOR 3

A-0189

Rapid gene analyses in pediatric central nervous system tumor using quantitative PCR device

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Introduction : Recent comprehensive molecular analyses of central nervous system (CNS) tumors identified several diagnostic or prognostic molecular markers. In pediatric CNS tumors, *BRAF* p.V600E mutation in pediatric-type diffuse low-grade glioma or circumscribed astrocytic glioma and histone H3 gene mutations in pediatric-type diffuse high-grade gliomas are quite important diagnostic markers. *CDKN2A* homozygous deletion (HD) is also an important marker in high-grade astrocytoma with piloid features (HGAP). Rapid identification of these molecular alterations could be effective for decision-making of tumor removal strategy.

Material & Methods : In this study, we established rapid gene analysis method using rapid quantitative PCR device, GeneSoC, by which we could obtain genotype of these important genes for 20 minutes.

Results : Using GeneSoC, we could perform 50 cycles of quantitative PCR for approximately 12 minutes. We established rapid gene analyses for mutations of H3 K27M, *BRAF* p.V600E and homozygous deletion of *CDKN2A* for pediatric CNS tumors. For elution of DNA, we incubated at least 1mg of tumor tissue at 95°for 5 minutes. Combined this boiling method with GeneSoC, we could obtain those genetic alterations for 20 minutes after collection of tumor tissue. For diffuse midline glioma, H3 K27-altered, pilocytic astrocytoma and HGAP cases, we could obtain accurate genetic alterations (H3 K27M, *BRAF* p.V600E *or CDKN2A* HD), using this method within 20 minutes, respectively.

Conclusions : In this study, we could obtain the genotype of important molecular markers rapidly, using GeneSoC device. This rapid genetic analysis might be effective not only for intra-operative molecular diagnosis, but also for immediate introduction of post-operative adjuvant therapy.

Oral presentation 3: TUMOR 3

A-0030

Since the Introduction of mTOR Inhibitors, Surgical Resection of SEGA Has Decreased, But Surgical Skills Remain an Essential Technique

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Introduction : Tuberous sclerosis complex (TSC) is a rare genetic disorder that can lead to the growth of benign tumors, including Subependymal Giant Cell Astrocytoma (SEGA). The introduction of mammalian target of rapamycin inhibitors (mTORi) has offered a pharmacological approach to manage SEGA in TSC patients.

Material & Methods : This study retrospectively reviewed patient charts from August 1979 to July 2020. Patients were divided into a pre-mTORi era group (Pre-group), treated before November 2012, and a post-mTORi era group (Post-group), treated from November 2012 onward when mTORi became available for SEGA in Japan. Treatment modalities, SEGA size, acute hydrocephalus rate, recurrence, malignant transformation, and mTORi adverse effects were compared between groups.

Results : Of 120 TSC patients, 24 had SEGA. Surgical resection was significantly more frequent in the Pre-group (6 of 7 patients, 86%) than in the Post-group (2 of 17 patients, 12%; $p = 0.001$). No malignant transformation of SEGA was observed. Patients treated with mTORi had significantly smaller SEGA sizes compared to those under a wait-and-see policy ($p = 0.012$). Adverse effects were identified in 7 of 11 patients on mTORi (64%).

Conclusions : Since the introduction of mTORi, the frequency of surgical resection for SEGA in TSC patients at our facility has significantly decreased. However, despite the effectiveness of mTORi, surgical skills remain an essential technique, especially for cases not responding to mTORi or experiencing significant adverse effects. The recent application of neuroendoscopic resection has emerged as a promising, minimally invasive surgical option for selected patients with SEGA associated with TSC.

Oral presentation 3: TUMOR 3

A-0055

Pediatric lower grade glioma surgery in the era of genomic medicine

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Introduction : In the WHO Brain Tumor Classification 2021, pediatric-type gliomas are classified separately from adult-type tumors. Additionally, with the advancement of cancer genomics, treatment options have expanded. Here we would like to consider surgery for pediatric low-grade gliomas.

Material & Methods : We retrospectively analyzed 41 cases of low-grade gliomas or glioneuronal tumors in pediatric patients who underwent surgery at our institute from 2002 to 2023. We discussed surgical strategies based on molecular classification.

Results : Among the cases, there were 34 gliomas and 7 glioneuronal tumors. According to the new WHO classification, they corresponded to 1) Adult-type diffuse gliomas, 2) Pediatric-type diffuse low-grade gliomas, 3) Circumscribed astrocytic tumors, or 4) Glioneuronal and neuronal tumors. Cases without molecular diagnosis were also considered in light of the new WHO classification. Among molecularly diagnosed astrocytomas involving the cerebral hemisphere, brain stem, spinal cord, there were no cases of 1), and 2) 3) were the main cases. It is important in surgical selection to note that these cases generally have a more favorable prognosis compared to the adult type and have usefulness in diagnosing MAPK pathway abnormalities leading to the selection of molecularly targeted drugs. 3) includes pilocytic astrocytoma, subependymal giant cell astrocytoma, and pleomorphic xanthoastrocytoma. Among these, in the treatment of pilocytic astrocytoma of the visual tract and hypothalamus, biopsy is essential for molecular diagnosis and therapeutic agent selection. 4) is mainly an epilepsy-related tumor, and epilepsy control by resection is expected.

Conclusions : Based on the new WHO classification, a reevaluation of treatment strategies for pediatric brain tumors is necessary.

Oral presentation 3: TUMOR 3

A-0058

Clinical characteristics of three patients with deficient DNA mismatch repair glioblastoma

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Introduction : Primary brain tumors associated with deficient mismatch repair (dMMR) are rare, accounting for less than 2% of primary brain tumors. dMMR is associated with microsatellite instability (MSI) and tumor mutational burden (TMB), subsequently leading to tumorigenesis. However, the findings in glioblastoma are still limited. We experienced three cases of dMMR-associated glioblastoma.

Material & Methods : All three patients were young adult males with left temporal IDH-wildtype glioblastoma having common distinctive feature of giant multinucleated cells. At the molecular level, Case1 had TMB-high and *MSH6* mutation, Case 2 had TMB-high, MSI-high, and *MLH1* mutation, and Case3 had TMB-high.

Results : Case 1 presented distant recurrence 14 months postoperatively. Case 2 was treated with Pembrolizumab, an immune-checkpoint inhibitor, but presented distant recurrence 15 months postoperatively. Case 3 presented good tumor control after Stupp regimen but presented a distant recurrence 48 months after the initial surgery.

Conclusions : Giant multinucleated cells in young patients with glioblastoma can suggest a dMMR-associated tumor. Therefore, genetic testing is highly recommended if we encounter such clinical features. All three cases presented distant recurrences, indicating that follow-up MRI needs to be extra cautious not to overlook distant lesions including the spinal cord. Effectiveness of an immune-checkpoint inhibitor for a dMMR-associated glioblastoma should be investigated in a prospective study.

Oral presentation 3: TUMOR 3

A-0078

Genetic diagnosis of pediatric brain tumors by liquid biopsy

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Introduction : For genetic diagnosis of brain tumors, it is necessary to directly harvest tumor cells by surgery. In this study, we performed genetic diagnosis by liquid biopsy (LB) using cell free DNA (cf DNA) extracted from plasma and cerebrospinal fluid (CSF) in children with brain tumors.

Material & Methods : The study included children who underwent surgical resection or biopsy for brain tumors between January 2019 and December 2020. Tumor-specific mutations were identified using whole exome sequencing on genomic DNA extracted from both tumor samples obtained through biopsy or resection procedures and blood cells. CSF (1-2 ml) collected intraoperatively and plasma (500-700 µl) obtained during surgery were used to analyze tumor-specific mutations through droplet digital PCR.

Results : Tumor-specific mutations were detected in 6 of 13 (46.2%) CSF samples. Two of the 6 samples had Class II or lower CSF cytology, and genetic mutations were also detected in one case of low-grade pilocytic astrocytoma. The median variant allele frequency in CSF was 16.0% (0.6-86.1%). On the other hand, no genetic mutations were detected in plasma in all patients.

Conclusions : In certain pediatric brain tumors, genetic diagnosis using cfDNA extracted from CSF, which can be repeatedly obtained with minimal invasiveness, is feasible. This approach has the potential to diagnose cases even with negative cytology or low-grade tumors.

Oral presentation 3: TUMOR 3

A-0094

Molecular pathological analysis of radiation-induced glioblastoma after pediatric cerebellar medulloblastoma treatment

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Introduction : The number of long-term survivors of pediatric medulloblastoma has been increasing due to improved treatment options. However, secondary cancer can be an issue for long-term survivors. We report the molecular pathological evaluation of two cases of radiation-induced glioblastoma.

Material & Methods : Of the 27 patients with cerebellar medulloblastoma treated at our institution, two developed cerebellar glioblastomas after the initial treatment at five years and 11 months and 22 years and eight months, respectively. These two cerebellar glioblastomas were clinically diagnosed as radiation-induced secondary cancer.

Results : Both of the two cases we clinically diagnosed as radiation-induced glioblastoma had negative IDH-1, EGFR expression and positive p53 expression in immunohistochemistry.

Conclusions : It has been reported that EGFR expression is negative as a molecular feature of radiation-induced glioblastoma, and the two cases we experienced were consistent with radiation-induced secondary cancers. In general, primary glioblastoma is positive for EGFR expression in the supratentorial region and negative in the infratentorial region. Therefore, in the case of supratentorial glioblastoma, it is useful to evaluate EGFR expression to distinguish primary glioblastoma from secondary glioblastoma.

Oral presentation 3: TUMOR 3

A-0186

Efficacy and safety of bevacizumab, irinotecan, and temozolomide combination for relapsed or refractory pediatric central nervous system embryonal tumor

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Introduction : Multidisciplinary treatment, which combines surgical treatment, various types of chemotherapy, and radiation therapy, is becoming standard in the treatment of pediatric brain tumors. However, there is a lack of evidence regarding treatments for treatment-resistant, recurrent, and disseminated cases. The purpose of this study was to evaluate the efficacy and safety of combination therapy of bevacizumab (Bev), irinotecan (CPT-11), and temozolomide (TMZ) in patients with relapsed/refractory central nervous system embryonal tumors.

Material & Methods : We retrospectively examined 13 pediatric patients with relapsed or refractory CNS embryonal tumors who received combination therapy. Specifically, 9 patients had medulloblastoma, 3 had atypical teratoid/rhabdoid tumor (AT/RT), and 1 had CNS embryonal tumor with rhabdoid features. Of the 9 medulloblastoma cases, 2 were categorized in the Sonic hedgehog subgroup and 6 in molecular subgroup 3 for medulloblastoma.

Results : The complete and partial objective response rates were 66.6% in patients with medulloblastoma and 75.0% in patients with AT/RT or CNS embryonal tumors with rhabdoid features. Furthermore, the 12- and 24-month progression-free survival rates were 69.2% and 51.9% for all patients. In contrast, the 12- and 24-month overall survival rates were 67.1% and 58.7% for all patients. We observed grade 3 neutropenia, thrombocytopenia, proteinuria, hypertension, diarrhea, and constipation in 23.1%, 7.7%, 23.1%, 7.7%, 7.7%, and 7.7% of patients, respectively. Furthermore, grade 4 neutropenia was observed in 7.1% of patients. Nonhematological adverse effects were mild and controlled with standard antiemetics.

Conclusions : Our findings suggest favorable survival outcomes and safety of combination therapy of TMZ, Bev, and CPT-11 in patients with relapsed/refractory embryonal tumors.

Oral presentation 3: TUMOR 3

A-0085

Pediatric brain stem gliomas: the past, present and future

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Introduction : Tumors arising in the brainstem can be broadly classified as; pattern of progression (exophytic or intrinsic), location (midbrain, pons, medulla oblongata, or upper cervical cord) and type of extension (diffuse or focal). we retrospectively reviewed the imaging, histopathology, and course of treatment in patients with and without surgical treatment.

Material & Methods : Twenty-one cases of brain stem tumors under the age of 20 years old between 2006 to 2023 were eligible for this retrospective review. Location; Four cases at midbrain, 15 cases at pons (including diffuse intrinsic pontine glioma), and one case at medulla oblongata.

Results : Headache, diplopia, and facial nerve palsy were the most common initial symptoms. Histological diagnosis was performed in 14/21 (67%); 9 cases of pediatric type high grade glioma; 5 cases of p-low grade glioma (p-LGG). Histological diagnosis: stereotactic biopsy in 1 case; open biopsy/extraction in 13 cases. Prognosis: morality rate 18/21 (86%); mean survival 13.8 months (8-27 months).

Conclusions : The prognosis of brain stem tumors is still devastating and surgical intervention can rarely be candidates for treatment. With the development of surgical instruments and the increasing availability of gene panel tests in recent years, histological diagnosis of brainstem lesions, especially for DIPG, has become increasingly important not only for diagnosis but also treatment. Besides, the evaluation of cranial nerve is necessary for assess the clinical course of brain stem tumors.