analyzed using iPlan® Flow software (Brainlab AG, Munich, Germany) for volumetric measurements. Target and catheter coordinates as well as radial, depth, and absolute error in MRI space were calculated with the Clearpoint imaging software (Clearpoint®, MRI Interventions Inc. Irvine, USA). RESULTS: Seven patients underwent two or more sequential CED infusions. No patients experienced deficits Clinical Terminology Criteria for Adverse Events (CTCAE) grade 3 or greater. One patient had persistent grade 2 cranial nerve deficit after a second infusion. No patients experienced hemorrhage or stroke post-operatively. There was a statistically significant decrease in radial error (p= 0.005) and absolute tip error (p=0.008) for infusion two compared to the initial infusion. Sequential infusions did not result in significantly different distribution capacity between the first and second infusion (Vd:Vi ratio: 2.66 ± 0.35 versus 2.42 ± 0.75; p=0.45). CONCLU-SIONS: This series demonstrates the ability to safely perform sequential CED infusions into the pediatric brainstem. Past treatments did not negatively influence the procedural work flow, technical application of the targeting interface, or distribution capacity. This limited experience provides a foundation for using repeat CED for oncologic purposes.

### SURG-25. EFFECT OF VENTRICULAR ENTRY DURING GLIOBLASTOMA RESECTION ON PATIENT OUTCOMES

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BACKGROUND: Tumor proximity to the ventricle and ventricular entry (VE) during surgery have both been associated with poorer prognoses; however, the interaction between these two factors is poorly understood. METHODS: The UCSF tumor registry was searched for patients with newly diagnosed and recurrent supratentorial glioblastoma who underwent surgical resection with the senior author between 2013 - 2018. Tumor location with respect to the subventricular zone (SVZ), size, VE, and extent of resection were assessed using pre and postoperative imaging. RE-SULTS: In the 200-patient cohort of newly diagnosed and recurrent glioblastoma, 26.5% had VE. Comparing patients with VE to those without VE, there was no difference in postoperative hydrocephalus (1.9% vs. 4.8%, p = 0.36), ventriculoperitoneal shunting (0% vs. 3.4%, p = 0.17), pseudomeningoceles (7.5% vs. 5.4%, p = 0.58), or subdural hematomas (11.3% vs. 3.4%, p = 0.07). Importantly, rates of leptomeningeal disease (7.5% in VE vs. 10.2% w/o VE, p = 0.57) and distant parenchymal recurrence (17.9% in VE vs. 23.1% w/o VE, p = 0.35) were not different between the groups. There was no effect of VE on EOR when controlling for SVZ type. Newly diagnosed patients with tumors contacting the SVZ (Type 1 or 2) had worse survival than patients with tumors that did not contact the SVZ (Type 3 or 4) (1.27 vs 1.84 years, p = 0.014, HR 1.8, CI 1.08 – 3.03), but VE was not associated with worse survival in these patients with high risk SVZ Type 1 and 2 tumors (1.15 vs 1.68 years, p = 0.151, HR 0.59, CI 0.26 – 1.34). DISCUSSION: VE was well tolerated with complications being rare events. There was no increase in leptomeningeal spread or distant parenchymal recurrence in patients with VE. Finally, VE did not change survival for patients with tumors contacting the ventricle.

#### SURG-27. TREATING HYDROCEPHALUS IN DIFFUSE MIDLINE GLIOMAS WITH AN H3 K27M MUTATION

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BACKGROUND: Diffuse midline gliomas (DMG) are a subset of malignant gliomas that share a characteristic Histone H3K27M mutation. These tumors are centrally located and may cause hydrocephalus on initial presentation. DMG lack characteristic imaging that distinguish from other primary brain tumors in the midline. We conducted this retrospective chart review of 43 consecutive patients presenting with midline tumors to determine: how many had a DMG; whether DMG patients with hydrocephalus were candidates for resection; and what the outcomes of endoscopic third ventriculostomy (ETV) versus ventriculoperitoneal shunt (VPS) placement were, as compared to wild type (WT) tumors. METHODS: We conducted an IRB approved retrospective chart review of patients presenting with midline tumors from 9/2016-3/2020 to determine H3K27M mutation status, hydrocephalus, and neurosurgery intervention. RESULTS: The median age of all midline tumor patients was 19.1 years (range 1.1-80.1). 26% (11/43) of midline tumors presented with H3K27M mutation, with a higher rate of hydrocephalus compared to patients without mutation [7/11 (65%) for DMG vs. 6/32 (19%) for WT, p< 0.05]. Of the seven H3K27M patients presenting with hydrocephalus, none were candidates for resection, 5 underwent ETV, and 2 underwent VPS placement as initial management. 4 out of these 5 ETVs failed within an average of 24 days (6-42 days). 2 patients then underwent VP shunt placement; the other 2 underwent secondary ETV

but both failed and required VP shunting as well. All 6 WT tumor patients had one procedure (1 ETV, 5 resection) to relieve hydrocephalus, and no patients had recurrent hydrocephalus. CONCLUSIONS: Both pediatric and adult patients may present with DMG associated with a higher rate of unresectable tumors and hydrocephalus on presentation. Furthermore, these data suggest that neuroendoscopic third ventriculostomy and septum pellucidum fenestration for the management of obstructive hydrocephalus in patients with DMG may be less robust than shunting.

#### SURG-28. PRESENTATION OF A POSTERIOR FOSSA TUMOR AT A HOSPITAL IN WESTERN JAMAICA

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The posterior cranial fossa is part of the cranial cavity, located between the foramen magnum and tentorium cerebelli that houses the cerebellum. pons and medulla oblongata. Commonly, tumors arising from this region in adults are cerebellar metastases or schwannomas of the vestibular nerve - the incidence of primary neoplasms is uncommon and more reserved for the pediatric population. A 28 year old female was in her usual state of health until last year when she started experiencing recurrent headaches with associated vomiting and intermittent loss of consciousness. A CT brain was done upon presentation to the hospital that revealed a 4th ventricle mass with obstructive hydrocephalus. A ventriculoperitoneal (VP) shunt was done thereafter to decompress the ventricular system, in anticipation for further surgical intervention for mass. Occipital craniotomy and resection of tumor was done and patient managed in a multidisciplinary manner in the intensive care unit. Post-operative course was marked by occipital pseudomeningocele with an associated CSF leak; a lumbar drain was placed in situ until complete resolution of leak. Histological analysis showed WHO Grade II Astrocytoma. Adult primary posterior fossa tumors are rare and can present with a constellation of symptoms. Although patient presented with findings in keeping with the diagnosis of an ependymoma, close clinical follow up will be required henceforth due to the refractory nature of such a low grade astrocytoma post-resection. Radiotherapy can also be considered in further management of case.

## SURG-29. CLINICAL FACTORS ASSOCIATED WITH RECURRENCE IN ATYPICAL MENINGIOMA: RETROSPECTIVE ANALYSIS OF 99 PATIENTS IN TWO INSTITUTES

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OBJECTIVE: The objectives of this study were to examine the local control (LC) rate of atypical meningioma after surgical resection with or without adjuvant treatment, to identify risk factors for the recurrence of atypical meningioma, and to compare our results to known factors from the literature. METHODS: Clinical and radiological records of patients with atypical meningiomas diagnosed at two institutes from January 2000 to December 2018 were reviewed retrospectively. Histopathological features were also reviewed using formalin-fixed paraffin embedded samples from pathological archives. RESULTS: Of the 99 atypical meningiomas eligible for analysis, 36 (36.4%) recurred during the follow-up period (mean 83.3 months, range 12-232 months). The rate of 3-year LC and 5-year LC was 80.8% and 74.7% respectively. The mean time-to-recurrence was 49.4 months (range 12-150 months). Multivariate analysis using Cox proportional-hazard regression model showed that the extent of resection (Hazard ratio [HR] 4.761, p=0.013), Ki67 index (HR 8.541, p=0.004), mitotic index (HR 3.275, p=0.044), tumor size (HR 3.228, p=0.041), and radiotherapy (HR 3.816, p=0.029) were independently associated with 3-year LC. These factors was also statistically associated with recurrence-free survival. In terms of radiotherapy after surgical resection, the recurrence was not prevented by immediate radiotherapy because of the strong effect of proliferative index on recurrence. Three cases of malignant transformation to WHO grade III meningioma were histopathologically confirmed after repeated surgery. Two out of these three patients succumbed to malignant transformation. The mean Ki67 proliferative index increased for recurrent cases in 18 patients (58.1%) from 7.55% (range 4-16) to 11.81% (range 5-24). CONCLUSION: The present study suggests that the extent of resection, proliferative index (according to Ki67 expression) and mitotic index, tumor size, and radiotherapy are associated with recurrence of atypical meningiomas. However, our results should be further validated through prospective and randomized clinical trials.

## SURG-30. SURGICAL RESECTION OF PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA: IMPACT OF PATIENT SELECTION ON OVERALL SURVIVAL

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OBJECTIVE: A possible prolonged survival after surgical resection for primary central nervous system lymphoma (PCNSL) lesions in selected patients has been suggested, but selection criteria for surgery, especially for solitary lesions, have never been established. METHODS: We retrospectively searched our patient database for records of adult patients (≥18 years) who were diagnosed and treated for a solitary lesion of PCNSL between 2005 and 2019. Patients were divided into groups according to whether they underwent surgical resection or needle biopsy. Statistical analyses were performed in an attempt to identify variables affecting outcome and possible survival advantage and to characterize subgroups of patients who would benefit from resection of their tumor compared to undergoing biopsy only. RESULTS: 113 patients with a solitary lesion of PCNSL were identified, 36 underwent surgical resection and 77 a diagnostic stereotaxic biopsy only. Pre-operative risk factors were found to include age > 70 years ([HR] 9.61, 95% [CI] 2.42-38.11, p=0.001) and deep seated lesions (adjusted HR 3.33, 95% CI 1.13-9.84, p=0.030). Having a postoperative Karnofsky Performance Scale (KPS) under 80 (adjusted HR 3.21, 95% CI 1.05-9.77, p=0.040) or surgical-site infections (adjusted HR 4.27, 95% CI 1.18-15.47, p=0.027) were significant postoperative risk factors. In a subgroup analysis, patients with a superficial tumor who underwent surgical resection had significantly longer survival times compared with those who underwent needle biopsy (median survival 34.3 months versus 8.9 months, p=0.014). Patients under 70 years who had a superficial tumor and underwent surgical resection had significantly prolonged survival, with a median survival of 35.0 months versus 8.9 months in patients from the same group who underwent needle biopsy (p=0.007). CONCLUSION: Specific subgroups of patients with a solitary PCNSL lesion might gain a survival benefit from surgical resection compared to undergoing only a diagnostic biopsy.

#### SURG-31. SURGICAL OUTCOME ANALYSES ON 244 CENTRAL NERVOUS SYSTEM HEMANGIOBLASTOMA

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BACKGROUND: Surgical resection of the central nervous system hemangioblastoma is often challenging, with vascular-rich nature and surrounding critical structures. von Hippel-Lindau (VHL) disease cases make the management more difficult with repeated surgeries required for multiple lesions. Here we investigated the surgical outcome of CNS HGBs on a large cohort to clarify the clinical and radiological parameters pertaining to the surgical success. METHODS: Retrospective analysis of consecutive, neurosurgically managed CNS HGB at Mayo Clinic, 1988-2018. RESULTS: Total 244 surgeries were performed for 172 patients. Gross total resection (GTR) was achieved in 90.0%, which was lower in cases brainstem lesions (78.3%) and was associated with the tumor, especially solid-portion volume (p=0.017). Intraoperative blood loss correlated with the size of solid portions, and the transfusion was performed in 7.5%. Postoperative complication was observed in 52.2%, including new/ worsening neurological deficit in 45.4%, wound complication in 9.1% and systemic complication in 4.0%. Postoperative rehabilitation was introduced in 61.7% of the patients, which was statistically associated with age, non-GTR and tumor location (brainstem and spine) on multivariate analysis (p=0.0031, 0.027 and 0.0066, respectively). Treatment-free survival was longer in VHL (vs sporadic) cases and GTR (vs non-GTR) cases, and multivariate analysis showed GTR was the only factor associated with treatment-free survival (p=0.0015). CONCLUSIONS: Surgery for CNS HGBs was shown to be challenging, with abundant intraoperative bleeding and high risk of postoperative complications necessitating rehabilitation. GTR of the lesion is of utmost importance, which increases the chance of long treatment-free survival as well as favorable direct postoperative clinical course.

## SURG-32. DIFFERENCES IN SURGICAL OUTCOME BETWEEN VON HIPPEL-LINDAU DISEASE RELATED AND SPORADIC HEMANGIOBLASTOMAS IN THE CENTRAL NERVOUS SYSTEM Shohei Nambu<sup>1</sup>, Shunsaku Takayanagi<sup>1</sup>, Hirokazu Takami<sup>2</sup>, Taichi Kin<sup>1</sup>, Shota Tanaka<sup>1</sup>. Hirofumi Nakatomi<sup>1</sup>, and Nobubito Saito<sup>1</sup>; <sup>1</sup>Department of

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Central nervous system hemangioblastoma (CNS HB) is a benign but highly vascular tumor, which accounts for 1.7% of all intracranial tumors. CNS HBs predominantly arise in the cerebellum, brainstem, and spinal cord. Approximately 20% of CNS HB is related to von-Hippel-Lindau (VHL) disease and repeated surgical resections are often required in VHL patients due to the presence of multiple CNS HBs. To explore the differences of surgical effect between VHL disease related and sporadic CNS HBs, outcome

analyses were performed based on the tumor locations. PATIENT AND METHODS: A total 83 cases that underwent surgical resections of CNS HBs at Tokyo University in 1996-2019 were included. Clinical information including age, sex, number of past surgical resections, pre- and post-operative modified Rankin Scale(mRS) were examined. RESULTS: Regarding the etiology, 46 VHL cases(cerebellum:30, medulla oblongata:6, spinal cord:10) and 37 sporadic cases (cerebellum: 28, medulla oblongata:4, spinal cord:5) were enrolled. The average changes in the postoperative mRS minus preoperative mRS were cerebellum -0.9/medulla oblongata -0.5/spinal cord -0.8 for VHL cases, and cerebellum -0.75/medulla oblongata -1/spinal cord -1 for sporadic cases. Fisher's exact tests were used for between-group comparison. DISCUSSION: The study showed no statistically significant difference between cerebellar and spinal cord cases regarding the perioperative change in the patient capability. Because of multiple lesions and repeated surgeries, VHL patients have a possibility to present with worse neurology after surgery. Further analysis is needed to clarify differences in surgical outcome for VHL disease related and sporadic related CNS HBs.

# SURG-33. ROLE OF SURGERY IN TREATING EPSTEIN-BARR VIRUS-ASSOCIATED SMOOTH MUSCLE TUMOR (EBV-SMT) WITH CENTRAL NERVOUS SYSTEM INVASION: A SYSTEMIC REVIEW Ko-Ting Chen<sup>1</sup>, Ka-Wei Lau<sup>2</sup>, Yu-Wei Hsu<sup>2</sup>, and Yin-Ting Lin<sup>2</sup>; <sup>1</sup>Chang Gung Memorial Hospital at Linkou, Taipei, Taiwan (Republic of China), <sup>2</sup>Chang Gung University, Taoyuan, Taiwan (Republic of China)

Epstein-Barr virus-associated smooth muscle tumor (EBV-SMT) is a rare mesenchymal tumor that almost exclusively occurs in immunocompromised hosts. The term unknown malignant potential once used interchangeably to describe EBV-SMT and the less invasive clinical behavior than leiomyosarcoma indicate an indolent growth pattern of EBV-SMT. Nevertheless, an EBV-SMT with CNS invasion would result in neurological deficits which may jeopardize patients' survival. We provide a systemic review of literature under PRISMA guideline on the clinical features, treatment modalities, role of surgery intervention and outcomes of all 65 reported EBV- SMTs with central nervous system (CNS) invasion. Over 95% of the reported cases were immunocompromised, while human immunodeficiency virus infection and post- organ transplantation being the most commonly associated underlying causes (near 90%). A 76.0% 1-year survival rate and 59.6% 5-year survival rate confirms the indolent and non-deadly nature of EBV-SMT even with CNS invasion. An immune survey and reconstruction should be conducted for every patient with CNS EBV-SMT. Surgical resection is mostly adopted as primary treatment to acquire diagnosis and relieve compressive effect. A total resection of tumor may be beneficial if tumor was symptomatic and had intracranial invasion

## SURG-34. MULTIDISCIPLINARY MANAGEMENT OF A PATIENT WITH OPTIC PATHWAY-HYPOTHALAMIC LANGERHANS CELL HISTIOCYTOSIS: A CASE REPORT

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Langerhans cell histiocytosis (LCH) of the Central Nervous System (CNS) is rare. Isolated involvement of the hypothalamic region is much more extremely rare with only 0.04 to 0.6% of the cases. We report a case of a 33 year-old female who presented with a one-year history of amenorrhea and a five-month history of intermittent headache, memory lapse, and somnolence. The patient was of normal body mass index with normal visual acuity and intact visual fields. Laboratory examinations revealed panhypopituitarism with central diabetes insipidus. Cranial magnetic resonance imaging showed a large lobulated mass measuring 1.9 x 2.2 x 2 cm in the suprasellar region which extended to the pituitary infundibulum, hypothalamus and retrochiasmatic region, with surrounding edema. The patient underwent right orbitozygomatic craniotomy and subtotal excision of the mass through subfrontal and transsylvian approaches. Histopathological examination of langerhans cells were observed with positive immunohistochemical stain for CD1a and S100 protein antigen markers establishing a diagnosis of CNS LCH. Thoracoabdominal computed tomography scan and bone scan were done postoperatively and showed no evidence of extracranial lesions. The patient had been receiving prednisone and vinblastine based chemotherapy regimen. She remains to be asymptomatic and on close surveillance. To date, there is no standardized treatment strategy for CNS LCH in the adult population. An accurate histopathologic diagnosis and a specialized multidisciplinary team approach especially involving Oncology, Neurosurgery, Ophthalmology and Endocrinology are critical to optimally tailor possible effective treatment options for patients with this similar disease. Long-term follow-up is crucial due to the increased risk of local recurrence and multisystemic involvement.