mostly sensory at lower limbs; NCS disclosed an axonal neuropathy; management and outcome were variable. Nine patients (dabrafenib-trametinib, n=5, encorafenib-binimetinib, n=3, and vemurafenib-cobimetinib, n=1) developed a demyelinating polyradiculoneuropathy; symptoms affected the four limbs and included hypoesthesia, weakness, and ataxia; cranial nerves were involved in four; NCS showed predominantly demyelinating features; most patients received intravenous immunoglobulins (n=6) or glucocorticoids (n=5). The outcome was variable; one patient was rechallenged with a different BRAF/MEK inhibitor with a rapid relapse. CONCLUSION: Patients under treatment with BRAF/MEK inhibitors may develop treatment-induced PN. Two main phenotypes are seen: a symmetric, axonal, length-dependent polyneuropathy, and a demyelinating polyradiculoneuropathy.

P11.53.B. EFFICACY AND TOLERABILITY OF REGORAFENIB IN PRETREATED PATIENTS WITH PROGRESSIVE CNS GRADE 3 OR 4 GLIOMAS
J. Werner1, L. Wolf2, G. Tischer2,1, E. K. Bauer1, M. Wollring2,1, G. Geccen3, M. Deckert4,1, A. Brunn1, R. Pappesch5, R. Goldbrunner6, G. R. Fink6, N. Galiakdas1,2, Dept. of Neurology, Faculty of Medicine and University Hospital Cologne, University of Cologne, Cologne, Germany, 1Inst. of Neuroscience and Medicine (INM-3), Research Center Juelich, Juelich, Germany, 2Inst. of Neuroradiology, Faculty of Medicine and University Hospital Cologne, University of Cologne, Cologne, Germany, 3Dept. of Internal Medicine (IOL), University of Aachen, Cologne, and Düsseldorf, Cologne, Germany, 4Inst. of Pathology, Faculty of Medicine and University Hospital Cologne, University of Cologne, Cologne, Germany, 5Dept. of General Neurosurgery, Faculty of Medicine and University Hospital Cologne, University of Cologne, Cologne, Germany.

BACKGROUND: The phase 2 REGOMA trial suggested an encouraging overall survival benefit in glioblastoma patients at first relapse treated with the multikinase inhibitor regorafenib. Here, we evaluated the efficacy and side effects of regorafenib in a real-life setting. MATERIAL AND METHODS: From 2018-2021, 30 patients with progressive WHO CNS grade 3 or 4, after first-line treatment with regorafenib (160 mg/d; first 3 weeks on, 2 weeks off) with individual dose adjustment depending on toxicity were retrospectively identified. Side effects were evaluated according to the Common Terminology Criteria for Adverse Events (version 3.0). MRI was obtained at baseline and after every second cycle. Tumor progression was assessed according to RANO criteria. After regorafenib initiation, the median progression-free survival (PFS) and overall survival (OS) were calculated. RESULTS: The median number of treatment lines before regorafenib was 2 (range, 1-4). The majority of patients (73%) had two or more pretreatment lines. At first relapse, 27% of patients received regorafenib. A total of 94 regorafenib cycles were administered (median number of cycles, 2; range, 1-9 cycles). Grade 3 and 4 side effects were observed in 47% and 7% of patients, respectively, and were not significantly increased in patients with two or more pretreatment lines (P=0.03). The most frequent side effects were laboratory abnormalities (62%). PFS was 2.6 months (range, 0.8-8.2 months), and the OS was 6.2 months (range, 0.9-24 months). CONCLUSION: In patients with progressive WHO grade 3 or 4 gliomas, treatment with regorafenib was effective despite considerable grade 3 or 4 side effects.

P11.54.A. FACTORS AFFECTING EARLY DIAGNOSIS IN NEUROLYMPHOMATOSIS
A. Egert1, P. Karschmitz MD, Dsc2, A. Huttner MD3, J. M. Baehring MD, Dsc,3, L. D. Kaulen MD, Dsc3,1, Department of Neurology, Yale School of Medicine, New Haven, CT, United States, 3Department of Neurology, Heidelberg University Hospital, Heidelberg, Germany, 2Clinical Cooperation Unit Neuro-Oncology, German Consortium for Translational Cancer Research (DKTK), German Cancer Research Center (DKFZ), Heidelberg, Germany.

BACKGROUND: Neurolymphomatosis is defined as infiltration of the peripheral nervous system by non-Hodgkin lymphoma (NHL). Given its rarity, correct diagnosis is often delayed. Here we assessed diagnostic work-up and factors affecting timely diagnosis at a tertiary referral center. MATERIAL AND METHODS: The quality control database of the Section of Neuro-Oncology at Yale Cancer Center was searched for neurolymphomatosis cases seen between 2000 and 2021. Clinical, radiological and pathological findings were collected. Factors affecting time to diagnosis were investigated in univariate analysis using SPSS version 27. P-values <0.05 were considered statistically significant. RESULTS: We identified 22 cases of neurolymphomatosis (primary n=7, secondary n=15) with secondary disease diagnosed a median of 16 months (range: 4-144) after NHL. Patients presented with painful polyneuropathy (n=16), painless polyneuropathy (n=4), cranial neuropathy (n=3), and autonomic neuropathy (n=1). Diagnosis was nerve biopsy-based (n=11) or resulted from integration of imaging findings (n=11) with NHL history (n=3) or detection of NHL in the cerebrospinal fluid (n=3). Most patients received regorafenib (n=16), peripheral nerves (n=12), plexus (n=10), cranial (n=3) or autonomic (n=1) nerves. Histologically, most cases were classified as diffuse-large B-cell lymphoma (n=18). Median interval from symptom onset to diagnosis was 3 months (range, 1-12). Secondary neurolymphomatosis was recognized before diagnosis in five cases. Conclusions of PET imaging (p<0.01) were associated with an earlier diagnosis. CONCLUSION: Diagnosis of neurolymphomatosis requires a high degree of clinical suspicion with subsequent integration of clinical, radiological and pathological findings. FDG-PET, the imaging gold standard, and a known NHL history correlated with an earlier diagnosis.

P11.55.B. POSTOPERATIVE MRI IS ABLE TO DETECT AN UNEXPECTED RESIDUAL TUMOR AFTER SURGERY OF BRAIN METASTASES: EXPERIENCE FROM 5 SPECIALIZED CENTERS
K. B. Kiesel1, J. Kerschaumser1, R. Prihoda1, M. Borkovec1, S. Thakur1, F. Merca2, D. Feucht3, A. Steindl1, A. S. Bergboff4, J. Furten5, J. Leitner3, A. Romagna3, C. Schweitz3, H. Stefania3, E. Marhold8, T. Roter1, M. Preussler1, C. Freyenschlag1, G. Widhalm3; 1Medical University Vienna, Vienna, Austria, 2Medical University Innsbruck, Innsbruck, Austria, 3Universitätsklinikum St. Pölten, St. Pölten, Austria, 4University Hospital Salzburg, Salzburg, Austria, 5Kepler Universitätsklinikum, Linz, Austria.

BACKGROUND: Brain metastases (BM) constitute the most common central nervous system tumors. The treatment options of BM consist of surgery, chemotherapy, radiosurgery, and immunotherapy. Regarding surgery in BM, the extent of resection (EOR) represents a crucial factor for patient prognosis. However, first studies using postoperative MRI demonstrated that an unexpected residual tumor after surgery of BM was not uncommon despite cone to core biopsy. The aim of this study was thus to investigate in a large cohort including multiple neurosurgical centers the EOR following BM resection, potential risk factors for incomplete resection and postsurgical follow-up data. MATERIAL AND METHODS: In the current retrospective study conducted at 5 specialized neurosurgical centers in Austria, we included patients with BM resection and available postoperative MRI. The EOR following BM resection was determined by postoperative MRI (complete vs incomplete resection). Additionally, the data on the intraoperative judgment of the EOR of the performing neurosurgeon were collected. Moreover, potential factors for incomplete resection including tumor localization, tumor volume, primary tumor, pattern of contrast media enhancement on imaging and tumor eloquence were investigated. Finally, the rate of local progression of BM after initial surgery was analyzed in the follow-up period and overall survival data were calculated. RESULTS: Altogether, 548 patients with 649 surgically treated BM were included. According to postoperative MRI, complete resection was achieved in 407 (66%) of 649 BM and incomplete resection in 242 (34%) of 649 BM. No tumor progression was found in 25% of cases and resulted in an unexpected residual tumor which was evident on postoperative MRI in 122 (22%) BM. Preoperative Xray CT was more common in BM with residual tumors and this was associated over, local progression was significantly more common in cases with incomplete resection. CONCLUSION: Our data of this study including multiple centers indicate that postoperative MRI is capable to detect a relatively high rate of unexpected residual tumors following resection of BM. Since occult progression was more common in BM with residual tumors and this was associated with shorter survival, special attention should be paid to achieve a complete tumor resection.

P11.56.A. SHARED AND PERSONALIZED DECISION-MAKING IN PRE-SURGICAL CONSULTATIONS OF BRAIN TUMOR PATIENTS AND NEUROSURGEONS
I. J. M. Brau1,2, M. M. Siimkoort1,2, J. I. M. van Vugt1, M. C. W. Jooost1,2, A. P. Y. Hoogendoorn1, G. J. Rutten2, K. Gehring1,2, Elisabeth-TweeSteden Hospital, Tilburg, Netherlands, 2Tilburg University, Tilburg, Netherlands, 3Leiden University Medical Center, Leiden, Netherlands.

BACKGROUND: In order to assess whether the oncological benefits outweigh the functional risks of treatment for an individual patient, it is important that physicians involve patients in decision-making and personalize the process. This is especially important for brain tumor patients considering the limited treatment options available. The aim of this study is to evaluate shared decision-making and the personalization of the decision-making process during pre-surgical consultations of brain tumor patients and neurosurgeons. MATERIAL AND METHODS: For this observational study we collected 66 audio-recordings of pre-surgical consultations of adult brain tumor patients (glioma, meningioma) and their neurosurgeons. A preliminary analysis based on a shared decision-making template was conducted on 14/66 transcribed audio-recordings to assess in how many consultations the following...
key elements of shared decision-making were present, namely 1) offering a choice to the patient and emphasizing that the patient's perspective is important in making the decision, 2) discussing treatment options with benefits and harms, and asking about treatment preferences and values. Next, thematic analysis will be performed by two independent researchers until data saturation is reached to gain insight into personalization of the decision-making process as an integral part of shared decision-making. RESULTS: Preliminary findings show that 3) offering a choice to the patient and emphasizing that the perspective of the patient is important in decisions was done in 12/14 consultations; 2) discussing treatment options with benefits and risks was present in every consultation (14/14); and 3) in 12/14 consultations the neurosurgeon asked the patient about his or her treatment preference. However, explicitly asking the patient about personal goals occurred only in 3/14 consultations. By the time of the conference, we expect to have explored the decision-making process more in-depth and will be able to share the themes we have identified related to personalization of the decision-making process. CONCLUSION: Key elements of shared decision-making were present in most pre-surgical consultations, except for asking patients about their personal goals. Discussing personal goals that patients have concerning daily life may facilitate personalization of the decision-making process.

P11.57.B. COMBINED SURGERY AND RADIOTHERAPY FOR BRAIN METASTASES. RETROSPECTIVE ANALYSIS OF A CONSECUTIVE COHORT OF 118 PATIENTS

T. Kazda1, P. Fadrus1, I. Selingerova1, R. Jancek1, P. Propisil1, L. Hynkova1, J. Garcic2, V. Vybihal3, I. Roskova4, B. Belanova4, M. Smedka5, J. Prochaska5, P. Slampa6, 1University Hospital Brno, Brno, Czech Republic, 2Institute Brno, Brno, Czech Republic, 3University Hospital Brno, Brno, Czech Republic, 4St. Anne’s University Hospital, Brno, Czech Republic, 5Masaryk University, Brno, Czech Republic.

BACKGROUND: Despite current advances in systemic therapy for brain metastases, neurosurgery remains the preferred method of choice in patients with limited brain metastases. Postoperative radiotherapy is indicated to reduce the risk of local recurrence in all patients after surgery. Therefore, the aim of this retrospective study is to describe clinical characteristics and survival outcomes in consecutive cohorts of patients treated by this combined local treatment. MATERIAL AND METHODS: Clinical data were retrieved from electronic medical records for consecutive patients who underwent surgery for brain metastases between 2007 and 2019. All patients underwent postoperative radiotherapy. Local progression free survival (localPFS) evaluated the local control at the operated site. DistalPFS at the other parts of the brain. Univariable and multivariable analysis of survival characteristics were performed. The median follow-up was 49 months. RESULTS: A total of 118 patients were included (54% women, median age 60 years, median Karnofsky index 80% at the time of radiotherapy). Single metastasis was treated in 66%, while 11% presented with more than 3 metastases. The most common primary diagnosis was lung cancer (33%) and breast (20%). Radical surgery was achieved in 92/117 (79% of patients). In total, only 48/118 (41%) of patients underwent targeted radiotherapy (mostly fractionated stereotactic radiotherapy of 25 Gy in 5 fractions). Significantly more patients (p=0.001) underwent postoperative radiotherapy, during 2017-2019 (43/48) compared to 2007-2015 period (3/48). A total of 20% of those who underwent postoperative whole brain radiotherapy (WBRT) had a specific technic of hippocampal sparing WBRT of WBRT with simultaneous integrated boost to remaining brain metastases. Median overall survival (OS) for all patients was 9 months (6.2 - 12), median localPFS 22 months (14 - not reached), median distalPFS 11 months (6.8 - 27) and median extracranialPFS 11 months (5.9 - 15). A significant (p=0.00017) difference in OS was when grouping patients according to median Karnofsky index (KPI) and median Karnofsky index (KPI) 80%. Greater median Karnofsky index (KPI) was associated with improved survival compared to postoperative WBRT in our cohort. Stereotactic radiotherapy should be considered in all patients with adequate radiotherapy technology. Supported by Ministry of Health of the Czech Republic AZV, NV18-03-00469 and NV18-03-00398.

P11.58.A. CASE OF A COMPLEX NEUROCUTANEOUS SYNDROME CHARACTERIZED BY EXTENSIVE PERIPHERAL NERVE SHEATH TUMORS AND SOMATIC ERBB2 MUTATION

M. Barden, J. Baehring; Yale School of Medicine, New Haven, CT, United States.

Schwannomatosis is a rare genetic tumor predisposition syndrome characterized by the presence of multiple non-intradermal schwannomas and the definitive absence of vestibular nerve involvement. Though considered benign, the burden of tumors can cause significant morbidity in the form of motor dysfunction and refractory neuropathic pain. Treatment is focused on mitigating these symptoms, which includes resection of offending tumors when disabling and anti-angogenic therapies and pain treatments. When feasible, targeted medical therapies are lacking. Schwannomatosis is molecularly distinct from the other neurofibromatoses and by definition lacks constitutional mutations in NF1 and NF2. Instead, constitutional mutations in SMARCB1 (also designated BAF1) or an array of tumor predisposition syndromes. WES on tumor tissue from both sites however did reveal a somatic ERBB2 variant (p.D769Y), suggesting mosaicism. ERBB2 D769Y has previously been classified as an activating mutation that confers sensitivity to agents like lapatinib, an irreversible tyrosine kinase inhibitors. Patients with ERBB2-mutated peripheral nerve sheath tumors may have broader therapeutic options in the variety of available tyrosine kinase inhibitors studied in other cancers.


A. Pellerito1, E. Bruno1, G. D’Alessandris2, V. Interno3, V. Polo4, E. Pronello5, T. Somma6, G. Spina7, T. Ius8, V. Esposito9,4, R. Ruda10,11, 1Division of Neuro-Oncology, Department of Neuroscience, University and City of Health and Science Hospital, Turin, Italy, IUOC Neurochirurgia, Fondazione Policlinico Universitario A. Gemelli IRCCS, Roma, Italy, 2Department of Interdisciplinary Medicine, Policlinico Hospital and University “Aldo Moro”, Bari, Italy, 3Department of Neuro-Oncology, Istituto Clinico San Giovanni di Dio, Fosso Fbaugh Hospital, Como, Italy, 4Department of Neurology Unit, Department of Translational Medicine, University of Eastern Piedmont, Novara, Italy, 5Division of Neurosurgery, Department of Neurosciences, Reproductive and Odontostomatological Sciences, University of the Second University of Naples, Naples, Italy, 6Division of Neuro-Oncology, IRCCS Fondazione Policlinico San Matteo, Pavia, Italy, 7Neurosurgery Unit, Department of Neurosciences, Santa Maria della Misericordia University Hospital, Udine, Italy, 8Department of Neurosurgery, IRCCS Veneto, Treviso, Italy, 9Department of Neurology, University, Roma, Italy.

BACKGROUND: Diffuse gliomas display heterogeneous biology, natural history, response to treatments, and outcome. According to the 2021 WHO Classification, an integration of histological and molecular factors is needed for the diagnosis of diffuse gliomas. The Italian Association of Neuro-Oncology (AION), with the participation of the Italian Society of Neuro-Oncology (SINcN), promoted a survey to explore how the 2021 WHO molecular diagnostic criteria are integrated into clinical practice in a national framework. MATERIAL AND METHODS: A web-based survey containing 38-item multiple-choice questions was sent to members of the AION and SINcN in February 2022 via the respective email listings of these organizations. RESULTS: We collected 152 answers. Most attendances were < 45-year-old (117, 77.0%). Participants from North, Centre and South of Italy were 85 (55.9%), 38 (25.0%), and 29 (19.1%). Academic and non-academic hospitals were 35 (46.1%) and 82 (53.9%). The presence of Italian neuro-oncology organizations. CONCLUSION: Overall, 152 responses were collected. The majority were < 45-year-old (<50%). The majority of respondents were from academic hospitals. The response rate from Southern Italy was low (<20%).