mutations might increase sensitivity of cancer cells to some chemotherapy through modulating gene expression and /or interfering with DNA repair mechanisms, therefore, affecting treatment outcome.

RARE-44. CLINICAL CHARACTERIZATION AND OUTCOME; OUR EXPERIENCE OF CHORDOMAS IN PEDIATRIC AND YOUNG ADULTS

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Pediatric chordomas are exceedingly rare and there are limited data to guide treatment decisions. We report a retrospective analysis of 19 patients with chordomas who received treatment at our institution from 2001–2020. Of the 19 patients, 15 had clival (79%), 3 cervical and 1 sacral chordoma. There were 9 males (47%). Median age at diagnosis of 10.6 years. Eight patients had gross total (42%) and 11 (58%) had sub-total resection. As front line therapy 15 patients (79%) underwent surgery followed by radiation (1 photon and 14 proton), 3 patients (16%) received surgery and chemotherapy (anaplastic histology) and 1 patient received only surgery. For patients treated with radiation therapy the average prescribed dose was 70 Gy (range: 52–74). Post-surgery and radiation, 14 of 15 patients remained in remission. Five (26%) patients had progressive disease (PD) with median time to progression of 13 months of whom 3 died of disease at median of 18 months. Treatment of PD consisted of chemotherapy and radiation for 3, re-resection with radiation for 1 and chemotherapy alone for 1 patient. The patients with metastatic and anaplastic disease mean survival is 21 month versus 45 for the rest of the cohort. In summary, post-operative adjuvant radiation provided an overall good outcome in majority of patients. Patients with anaplastic pathology and metastasis at diagnosis had worse outcome. Those who relapsed, subsequent treatment was palliative at best with short survival. Molecular analysis is warranted in future for better disease stratification.

RARE-45. SARCOMAS INVOLVING THE CENTRAL NERVOUS SYSTEM AT INITIAL PRESENTATION IN CHILDREN AND YOUNG ADULTS: A CASE SERIES

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Sarcomas of bone, soft tissue, or neural origin may occasionally invade the central nervous system (CNS), causing diagnostic and therapeutic challenges. We aim to investigate the clinical features of sarcomas involving the CNS at initial presentation. During 2015/01–2019/12, nine consecutive patients (4 Males and 5 Females) younger than 30 years of age treated at a University Healthcare System in Northern Taiwan were included. The median age was 8.7 years (range, 2–24 years); diagnoses were Ewing Sarcoma with *EWSR1* rearrangements (*n*=4), *CIC-NUTM1* Sarcoma (*n*=1), Osteosarcoma (*n*=2), Malignant Peripheral Nerve Sheath Tumor (MPNST; *n*=1), and extramedullary myeloid sarcoma (*n*=1). The tumors originated from the skull (*n*=1), dura (*n*=1), vertebra (*n*=4), spinal canal (*n*=1), or extra-CNS sites (*n*=2). Four patients had metastases (1 Ewing sarcoma, 2 osteosarcoma, and 1 extramedullary myeloid sarcoma). The main symptom at diagnosis was facial/eye pain (*n*=2), back pain (*n*=3), arm weakness (*n*=1), or gait disturbance (*n*=3). Upfront neurosurgical decompression (*n*=7) or urgent radiotherapy (*n*=1) was performed in most patients. At a median follow-up duration of 20.1 months, the overall survival rate was 70%. All patients with Ewing sarcoma (*n*=4) and *CIC-NUTM1* sarcoma (*n*=1) achieved Complete Response after surgery, interval-compressed chemotherapy, radiotherapy, and adjuvant chemotherapy. Patients with stage IV osteosarcoma (*n*=2) had Partial Response; the patients with MPNST and extraskeletal myeloid sarcoma died of Progressive Disease at 18 and 3 months after diagnosis, respectively. We conclude that timely decompression, early diagnosis, and histology-driven multimodality treatment are effective strategies in managing sarcomas involving the CNS.

RARE-46. A THIRTEEN YEAR PATIENT JOURNEY OF INFANT GIANT CLIVAL CHORDOMA: CASE REPORT AND LITERATURE REVIEW

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In 2006 we reported the youngest case of a large clival chordoma, a 15-week old baby, the second case to present without skull base involvement and the fourth case of chordoma in a patient with tuberous sclerosis. This unusually rare case (surgically un-resect able) underwent endoscopic skull-base diagnostic biopsy and a novel chemotherapy regime that aimed to control his disease[i]. Initial tumour control was achieved with chemotherapy (Ifosfamide, doxorubicin with dexrazonane, intrathecal hydrocortisone, methotrexate, cytarabine). Carboplatin and etoposide were later given for a further year. Following this, Sirolimus and imatinib were used for another twelve months due to primary tumour regrowth and three new skull-vault lesions. Sirolimus alone was continued for an additional year, but stopped due to optic neuritis. Imatinib was given until further progression two years later, leading to a change to everolimus. Surgery for the ventral foramen magnum was performed a year later. The patient received further surgery and radiotherapy for tumour recurrence. Sadly the tumour metastasised and he succumbed at age 13. Chordomas are aggressive and recur frequently. Complete primary resection followed by radiotherapy/proton beam therapy offers the best chance of cure but is not an option in infants with giant lesions, as in our case. We inform on alternative targeted treatment strategies and review the literature on these rare lesions. [i] Kambogiorgas D, St George EJ, Chapman S, English M, Solanki G: Infantile Clivus chordoma without clivus involvement: Case report and review of the literature, Childs Nerv System (2006) 22:1369–1374

RARE-47. DIFFUSE LEPTOMENINGEAL DISSEMINATED GLIONEURONAL TUMOR: CASE-SERIES

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Diffuse leptomeningeal disseminated glioneuronal tumor (DL-GNT) is a rare brain tumor that presents as a plaque-like subarachnoid tumor, commonly involving the basal cisterns and interhemispheric fissure of children but lacking intraparenchymal tumor. Here we report two cases focusing on clinicopathologic features. In all patients, radiography revealed characteristic leptomeningeal thickening and enhancement with minor superficial parenchymal lesions. The broadcast of the knowledge about this type of disease is important to increase awareness on this subject.

RARE-48. CHARACTERISTICS AND OUTCOME OF DIFFUSE LEPTOMENINGEAL GLIONEURONAL TUMOR (DLGNT): A SINGLE INSTITUTION EXPERIENCE

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Diffuse leptomeningeal glioneuronal tumors (DLGNT) are rare with an unknown etiology and unestablished incidence. Most frequently reported genetic alteration is *KIAA1549-BRAF* fusion. We present four DLGNT cases diagnosed between 2005–2018. Patient 1 is a female who presented with a 2-year history of back pain subsequently diagnosed with pilocytic astrocytoma. Re-imaging 3 months post-resection revealed a low grade glioneuronal tumor with BRAF duplication. Patient 2 is a female who presented with recurrent vomiting, dizziness, and hydrocephalus. The patient underwent biopsy which was consistent with oligodendrogliomatosis; no genetic analysis was done. Patient 3 is a male who presented with worsening headaches and intermittent vomiting. Approximately 5 months after resection, imaging showed leptomeningeal disease and further testing revealed *KIAA1549-BRAF* fusion and 1p deletion. Patient 4 is a male who presented with hydrocephalus. Imaging showed disseminated leptomeningeal enhancement without a dominant mass lesion; biopsy and clinical history confirmed the diagnosis. All four patients received chemotherapy, Patients 1 and 3 underwent radiation therapy, and Patient 3 received a MEK-inhibitor to which he had a great response. However, the patient was non-compliant and had PD which continued despite re-starting therapy. Patients 1, 2, and 3 have died of progressive disease; survival was Patient 1, 276 days, Patient 2, approximately 7 years and 8 months, and Patient 3, 2 years and 11 months. Patient 4 remains alive with disease 4.5 years from diagnosis. There is much to be learned about this rare, poorly understood disease but hope for improvement through therapeutic targeting of the MAPK pathway.

RARE-49. CHOROID PLEXUS ADENOMA WITHOUT ASSOCIATED HYDROCEPHALUS PRESENTING AS PRECOCIOUS PUBERTY

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Precocious puberty (PP) is a rare presentation of intracranial pathology unrelated to the pituitary. PP in this setting is considered a paraneoplastic phenomenon, achieved through synthesis of sex-hormones by the tumor itself or via alterations in the release of gonadotrophins from the pituitary. The latter has been described with masses adjacent to the pituitary or with those which cause hydrocephalus. We describe a case of a choroid plexus papilloma (CPP) without hydrocephalus presenting as precocious puberty.