LINC-28. EPIDEMIOLOGICAL CHARACTERISTICS AND SURVIVAL OUTCOMES OF CHILDREN WITH MEDULLOBLASTOMA TREATED AT THE NATIONAL CANCER INSTITUTE (INCA) IN RIO DE JANEIRO, BRAZIL

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BACKGROUND: Medulloblastoma (MB), the most malignant brain tumor of childhood has survival outcomes exceeding 80% for standard risk and 60% for high risk patients in high-income countries (HIC). These results have not been replicated in low-to-middle income countries (LMIC), where 80% of children with cancer live. Brazil is an upper-middle income country according to WHO, with heterogeneous features of LMIC and HIC. METHODOLOGY: We conducted a retrospective review of 126 children (0–18 years) diagnosed with MB from 1997 to 2016 at INCA. Data on patients, disease characteristics and treatment information were retrieved from the charts and summarized descriptively; overall survival (OS) and event-free survival (EFS) were calculated using the Kaplan-Meier Method. RESULTS: The male/female ratio was 1.42 and the median age at diagnosis was 7.9 years. Headache (79%) and nausea/vomiting (75%) were the most common presenting symptoms. The median time from onset of symptoms to surgery was 50 days. The OS for standard-risk patients was 69% and 53% for high-risk patients. Patients initiating radiation therapy within 42 days after surgery (70.6% versus 59.6%, p=0.016) experienced better OS. Forty-five patients (35%) had metastatic disease at admission. Lower metastatic incidence was associated with lower OS (5/35% versus 14/20% p=0.49). A total of 20% of patients lived >40km from INCA fare better (OS 68.2% versus 51.1% p=0.032). Almost 20% of families lived below the Brazilian minimum wage. CONCLUSIONS: These findings suggest that socioeconomic factors, education, early diagnosis and continuous data collection, besides oncological treatment must be addressed to improve the survival of children with MB.

LINC-29. IMPACT OF RELA FUSION ON OUTCOMES OF CHILDHOOD SUPRATENTORIAL EPENDYMOMAS (ST-EPEN)

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BACKGROUND: Ependymomas are heterogeneous group of tumours with variable clinical course and diverse molecular features. RELA fusion status has been reported to have prognostic impact in ST-EPEN. Our retrospective study analysed the prevalence and clinical impact of RELA fusion in ST-EPEN.

METHODS: A prospective study was conducted at the Tata Memorial Centre. ST-EPEN patients from 2007 to 2019 were included (median age-10.2 years; boy:girl ratio-1.4:1) for analysis. Histopathology, histological features and immunohistochemical parameters (L1CAM expression vs. negative-expression) were comparable (61.1%/55% versus 62.6%/53.6%; p=0.391). The 3-year/5-year EFS of tumors expressing RELA fusion 1/2 by RT-PCR. Children were treated as per guidelines by the Neuro-Oncology and Radiation Oncology teams. Patients were followed up till May 2019. The OS and EFS of RELA-positive and RELA-negative tumors ranged from 0 to 33% in 4 cases: 1WNT; 3SHH; 1Group 4. TILs ranged from 0–60/mm² with a median of 1. TILs were significantly higher in SHH subgroup. CONCLUSION: PD-L1 positivity and number of TILs and TAMs were significantly more in SHH subgroup tumors followed by WNT tumors. CTLA-4 expression did not correlate with subgroups. All parameters showed a positive trend with increasing age.

LINC-31. TREATMENT OUTCOME IN CHILDREN WITH MEDULLOBLASTOMA IN MEDIUM-INCOME COUNTRY: AN EXPERIENCE FROM A SINGLE TERTIARY CENTRE IN KUALA LUMPUR, MALAYSIA

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INTRODUCTION: Medulloblastoma is the most common malignant brain tumour in children. The overall outcome has improved however, this was not translated to developing nations. METHOD: This was a retrospective review of patients from January 2000 to December 2017. Treatment was given using modified SIOP PNET 4 protocol; cranio-spinal irradiation (CSI), a total of 54G with vincristine followed by 8 cycles of adjuvant chemotherapy. Prior to year 2007, patients had CSI with or without adjuvant chemotherapy. Those <3 years old received modified UKCCSG/ SIOP CNS-1 protocol with 2 weekly chemotherapy for a duration of 392 days followed by CSI when required. All patients had MRI brain and spine, and tissue histopathological examination but without molecular subtype. RESULTS: Medulloblastoma comprised of 30% (n=31) out of total 103 brain tumour cases. Mean age at presentation was 7.6 years old (SD 4.4) with male to female ratio of 2:1. Average time of symptoms was 4.8 weeks. Majority, 77.4% was high risk and 19.4% was standard risk. There was high treatment abandonment rate (35.5%, n=11). Three patients returned and completed treatment after multiple surgeries in an average of 9 months. Three years OS were 69.4% and 48.4%, respectively.

Six patients aged <3 years; half had advance disease on palliative care post surgery. Other survivors had severe learning difficulty and two had second malignancy (meningioma and thyroid carcinoma) at average 15.5 years after diagnosis. CONCLUSION: SIOP CNS-1 is not sufficient for developing nations. Crucial. Moreover, multidisciplinary management and molecular stratification are important in improving the outcome.

LINC-32. REPORT OF AN INITIAL SITE VISIT TO DETERMINE FEASIBILITY AND IMPLEMENTATION OF A COMPREHENSIVE NEURO-ONCOLOGY PROGRAM IN KENYA

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BACKGROUND: Pediatric central nervous system (CNS) tumors are the leading solid tumors in the United States, but vastly under-reported in the African population. There’s limited data on childhood brain tumors as well as the histopathological distribution in Kenya. This report surveys as an initial site visit to determine the feasibility of a comprehensive neuro-oncology program at Kenyatta National Hospital (KNH) in Nairobi, Kenya. DESIGN: This collaboration began with a visit from the director of neuro-oncology at KNH to our neuro-oncology program at Riley Hospital for Children at Indiana University Health in May 2019. This report includes recommendations from the May 2019 trip, as well as a reciprocal site visit to Kenya in January 2020.

RESULTS: Building off the May 2019 trip, a bi-directional collaboration has been initiated and maintained. Additionally, the KNH program has many necessary components to forming a comprehensive neuro-oncology program, including capable neurosurgeons with a neurosurgical training program, radiation oncology, early diagnosis and continuous data collection, besides oncological treatment must be addressed to improve the survival of children with MB.
Hospital has the expertise to build a comprehensive neuro-oncology program. The program currently lacks a dedicated nurse coordinator and “specialist” in neuro-oncology. Ongoing discussions with local stakeholders are aimed to galvanize national support to improve care for children with brain tumors and to plan a multidisciplinary neuro-oncology symposium in 2021. In the meantime, telemedicine efforts can support nursing education and reiterate the multidisciplinary needs for children with brain tumors.

LINC-33. MULTIDISCIPLINARY MANAGEMENT OF PAEDIATRIC PRIMARY CENTRAL NERVOUS SYSTEM LYMPHOMA: UPDATED EXPERIENCE FROM A REGIONAL CANCER CENTRE IN NORTH INDIA

Paediatric primary central nervous system lymphoma (PCNSL) constitutes 1% of all PCNSLs. Data pertaining to paediatric PCNSLs (2016–19) was abstracted by retrospective chart review. We identified 7 paediatric patients with PCNSL. None had congenital or acquired immunodeficiency. The median age at presentation was 13 years. The male to female ratio was 4:3.

The median EOCG performance status was 2. On neuro-imaging, 3 patients had solitary and 4 patients had multiple lesions. CSF cytology showed atypical lymphocytes in 1 patient. None had ocular involvement. Systemic lymphoma work-up was negative in all. Biopsy and resection of tumour were done in 4 patients each. Histopathology revealed DLBCL in 6 and B-cell NHL in 1 patient. All patients underwent induction chemotherapy (median-5 cycles). CNS disease was controlled in 5 patients. The median time to complete CNS therapy was 9 months. Paediatric PCNSL is a rare tumour entity and multidisciplinary management with high dose Methotrexate and Rituximab based chemo-immunotherapy and cranial radiotherapy leads to excellent early clinical outcome.

LINC-34. OPTIC NERVE INFILTRATION: RARE MANIFESTATION OF CHILDREN WITH ACUTE LYMPHOBlastic LEUKEMIA IN REMISSION

BACKGROUND: Optic nerve infiltration in acute lymphoblastic leukemia is a rare manifestation. This infiltration may appear months in advance as an isolated sign of extramedullary relapse and considered as one of the significant clinical findings of central nervous system leukemia. AIM: To describe the case of rapidly progressive optic nerve infiltration in a girl with ALL in remission. CASE: A 13-year-old girl in full remission following treatment for B-cell acute lymphoblastic leukemia presented with decreased vision and proptosis on the left eye. She completed the chemotherapy course two years before. On physical examination, we found the optic disc swelling in her left eyes. There were no signs of relapse from the hematological, cerebrospinal fluid analysis, and bone marrow aspiration. The orbital CT found a mass in the left retrobulbar (size 29x48x32 mm), suspected of optic nerve glioma. The mass has grown rapidly in a month, and she lost her left sight. The involved eye was exenterated (60x55x40 mm). The histopathology and immunohistochemistry showed the B-cell acute lymphoblastic leukemia. Unfortunately, the patient could not come for further follow-up due to the COVID-19 large-scale social distancing. Two months later, she came with pallor and pain in all of her body. The bone marrow aspiration showed leukemic relapse and she is undergoing chemotherapy. CONCLUSION: Optic nerve infiltration by leukemia requires both diagnostic certainty and urgent management. A routine ophthalmic assessment is recommended in patients with a history of acute lymphoblastic leukemia to diagnose optic nerve involvement due to leukemic infiltration.

LINC-35. THE ST. JUDE GLOBAL ACADEMY NEURO-ONCOLOGY TRAINING SEMINAR: A MULTIDISCIPLINARY, INTERNATIONAL EDUCATION PROGRAM

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The success of the treatment of children with central nervous system (CNS) tumors relies on a effective multidisciplinary team, with up-to-date and broad knowledge and skills. The St. Jude Global Academy Neuro-Oncology Training Seminar was launched as course in globally applicable content in pediatric neuro-oncology with a focus on multidisciplinary work-up across the world. A routine ophthalmic assessment that is most relevant for the learners, a needs assessment survey that included evaluation of team dynamics, treatment capacity, existing knowledge, and educational goals was designed. Survey questions in 11 domains were prepared by 24 sites in 11 countries across the world. An online course was used to create the course that consists of two components: a 9-week online course and a 10-day workshop at the St. Jude campus. 72 participants from 11 institutions enrolled in the online portion and 20 participants were selected based on grades to attend the workshop. The workshop included: a retrospective post-test evaluation established that learners improved their understanding of the barriers to care, possible solutions to improve care, understanding of diagnosis and treatment, and methodology to implement projects (<0.01). All participating teams developed projects that are locally implemented. Those presented at the workshop formed a multidisciplinary, international collaborative group (Global Alliance in Pediatric Neuro-Oncology). This experience establishes that educational programs with systematically created curricula can not only improved knowledge but be a mechanism to share experiences and create collaborative networks. Ultimately, patient outcomes will be tracked to monitor the true impact of the course.

LINC-36. TRILATERAL RETINOBLASTOMA: A REPORT OF FOUR CASES

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Retinoblastoma is the most common primary intraocular cancer that usually develops in early childhood. About 5% of those patients are at risk of developing trilateral retinoblastoma (TRB). In developing countries, the vast majority of them come from the late stage; therefore, ocular and patient survival rates are lower than in developed countries. From 2015–2019, we found four cases of trilateral retinoblastoma. Two of them had bilateral retinoblastoma, and two had unilateral retinoblastoma. They all presented with leukocoria and had no family history of retinoblastoma. The mean age was 13.8 months (range 9–24 months of age). The diagnosis of trilateral retinoblastoma was made from initial head CT/ MRI. They were treated conservatively with high dose VEC chemotherapy, and three of them have definitive retinoblastoma treatment. Trilateral retinoblastoma is usually fatal and needs multidisciplinary treatment care. In developing countries, it is important to evaluate distant metastasis. Head CT or MRI from the initial diagnosis to exclude the trilateral retinoblastoma.

LINC-37. 500 CONSECUTIVE SURGICAL CASES FROM THE PEDIATRIC ONCOLOGY NEUROSURGERY GROUP: UNDERSTANDING THE PERSPECTIVE OF A TERTIARY CENTER IN BRAZIL

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With this presentation we aim to present cases submitted to surgery by the same group of surgeons since 2010, presenting the physical structure, medical assistance, scientific production and the challenges that we need to overcome in the second decade of the twenty-first century, in a developing country.

LINC-39. PERFORMANCE STATUS OF PEDIATRIC PATIENTS WITH CENTRAL NERVOUS SYSTEM TUMORS TREATED IN MEXICO, A SINGLE-CENTER EXPERIENCE

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BACKGROUND: Central nervous system (CNS) tumors are the most common malignancies in children and adolescents. They account for about 15% of all cancers at this age. Little is known about the specific epidemiology of this group in Mexico and there are no reports of results focused on the Performance Status of patients who are treated in a multidisciplinary setting. OBJECTIVE: To describe the Performance Status of CNS pediatric patients after being treated with a multidisciplinary approach in a tertiary center. METHODS: We report a retrospective chart review of all pediatric patients who presented to the Neuro-Oncology Clinic at Teleton Pediatric Oncology Hospital in Queretaro, Mexico, from December 2014 to January 2020. We analyzed age, gender, the extent of surgical resection and histopathology. Performance Status was assessed using ECOG and Karnofsky/Lansky scores during every patient’s last follow-up visit. RESULTS: A total of 56 patients were treated, epidemiology and histopathology variants are similar to those described in the international literature. With a median follow-up of 33 months, 35 patients are alive (62.5%), 28 of them (74.2%)