LINC-21. PROGNOSTIC FACTORS AND SURVIVAL OF LOW-GRADE GLIOMAS IN CHILDREN AND ADOLESCENTS – A MULTICENTER STUDY IN PERU

Rosdali Diaz-Coronado[1], Eddy Hernández-Broncano[1], Sandro Casavilca-Zambrano[1], Danny Campos-Sanchez[2], Ivan Maza[3], Mariela Tello[2], Esmeralda Leon[4], Hernan Cañari[5], Javier Custodio[3], Luis Ojeda-Medina[1], Tatiana Negreiros[1], Olga Cordova[3], Pamela Mora-Alferez[1], Juan Garcia-Leon[1], Liliana Vasquez[6], Daniel Moreira[7], Ibrahim Qaddoumi[7]; [1]Instituto Nacional de Enfermedades Neoplasicas, Lima, Lima, Peru. [2]Instituto de Salud del Niño- San Borja, Lima, Lima, Peru. [3]Hospital Nacional Edgardo Rebagliati Martins, Lima, Lima, Peru. [4]Hospital Nacional Guillermo Almenara Irigoyen, Lima, Lima, Peru. [5]Instituto Nacional del Niño-Breña, Lima, Lima, Peru. [6]Organizacion Mundial de la Salud, Columbia, Washington, USA. [7]St. Jude Children's Research Hospital, Memphis, Tennessee, USA

BACKGROUND: Low-grade gliomas (LGG) are the most common central nervous system (CNS) tumors in children. Peru is an index country for the Global Initiative for Childhood Cancer (GICC). As part of the Initiative, a multidisciplinary brain tumor team was formed in 2020 that includes five national reference centers: National Institute of Neoplastic Diseases, National Children's Health Institute-San Borja, National Children's Health Institute-Breña, Edgardo Rebagliati Martins National Hospital and Guillermo Almenara Irigoyen National Hospital. This multicenter study sought to evaluate the survival and prognostic factors of patients younger than 18 years diagnosed with LGG, an index cancer for the GICC. METHODS: A retrospective study included all patients diagnosed with LGG in the five centers between 2014 and 2018. We analyzed clinical characteristics, histology, and treatment modalities. We used the Kaplan-Meier method for survival. RESULTS: 194 patients were registered; 136 patients were included. M/F ratio was 1.2, mean age 7 years old. The most frequent location was infratentorial (42.2%), supratentorial (34.9%), optic/chiasm/sellar (11%) and brainstem (11.9%). The most frequent histological types were pilocytic astrocytoma (61%), diffuse astrocytoma (10.3%), oligodendroglioma (5.2%), and other low-grade neoplasms (23.5%). Surgery was performed in 109 patients (83.2%). Chemotherapy alone was used in 17 (12.5%), while radiotherapy in 20 (14.7%). Overall survival at 5 years was 82.9% (95% CI 73.3 - 89.4). Age younger than 3 years (p=0.002), diffuse histological type (p=0.04), and location in the brainstem (p=0.001) were factors associated with a worse prognosis. CONCLUSIONS: Within the framework of the GICC, this work is one of the first steps to understand the current context of pediatric CNS tumor care in Peru. Although the reported survival rate is about the GICC goal of 60%, further improvements in care are needed to increase survival to level closer to high-resource setting and decrease long term morbidity.

LINC-22. PRIMARY CENTRAL NERVOUS SYSTEM (CNS) GERM CELL TUMORS (GCT) IN CENTRAL AMERICA AND THE CARIBBEAN REGION: AN AHOPCA 20-YEAR EXPERIENCE

Ana Veronica Giron[1], Patricia Calderon[2], Estuardo Pineda[3], Margarita Montero[4], Yamel Lizardo[5], Ute Bartels[6], Diana S. Osorio[7]; [1]Unidad Nacional de Oncologia Pediatrica, Guatemala, Guatemala. [2]Hospital Escuela La Mascota, Managua, Nicaragua. [3]Hospital Nacional de Niños Benjamin Bloom, San Salvador, El Salvador. [4]Hospital Infantil Robert Reid Cabral, Santo Domingo, Dominican Republic. [5]Hospital Infantil Regional Dr. Arturo Grullon, Santiago de los Caballeros, Dominican Republic. [6]The Hospital for Sick Children, Toronto, Canada. [7]ICON, Clinical Research Organization, ,, USA

BACKGROUND: Primary CNS GCT are rare neoplasms in pediatrics. Treatment depends on histological subtype and extent of disease. Overall survival (OS) is above 85% in high-income countries (HIC). OBJECTIVE: To describe the experience in treating CNS GCT in 4 of 8 countries, members of AHOPCA (Asociación de Hemato-Oncología Pediátrica de Centro América) and 5-year OS. DESIGN/METHODS: We conducted a retrospective chart review of patients treated for CNS GCT. Epidemiological, clinical characteristics, histology, treatment modalities and outcome were analyzed. RESULTS: From 2001 to 2021, 45 patients were included. Guatemala: 22, Nicaragua: 15, Dominican Republic: 3 and El Salvador: 5. Thirty (66.6%) were male, mean age at diagnosis was 8.6 years (range 12 months to 17 years). Presenting symptoms were headaches/vomiting (n=26, 57.7 %), visual disturbances (n=15, 33.3%), diabetes insipidus (n=7, 15.5%). Two patients (NGGCT) presented with precocious puberty. Upfront surgery was done in 33 cases (73.33%); 21 (63.6%) germinomas, 9 (27.2%) NGGCT and 3 (9%) CNS GCT. Eight patients were diagnosed and treated based on CSF tumor marker elevation; four germinomas (BHCG 11.32-29.41mUI/mL) and four NGGCT (BHCG 84.43-201.97mUI/mL or positive AFP>10 ng/mL). Tumor locations included suprasellar (n=15, 33.3%), pineal (n=14, 31.1%), thalamus/basal ganglia (n=5, 11%), other (n=6, 13.3%); and one bifocal. Four (8.8%) had metastatic disease, 6 with positive CSF, staging data was incomplete in 25 patients (55.5%). Patients were treated with varied chemotherapy and radiotherapy modalities. Nine patients had incomplete data regarding treatment. Five-year OS was 65% (61.5% for germinoma and 56% for NGGCT). Two patients with histologically confirmed germinoma relapsed. CONCLUSIONS: Germinoma was the most common histology and there was a male predominance. More than half patients had incomplete staging data and treatment was variable. OS is lower compared to HIC. Standardized treatment protocols will aid in adequate staging, treatment planning, prevent complications and improve survival.

LINC-23. FACTORS INFLUENCING OUTCOMES OF OLDER CHILDREN WITH MEDULLOBLASTOMA OVER 15 YEARS IN PERU, A RESOURCE LIMITED SETTING

Rosdali Diaz-Coronado[1], James B. Reinecke[2], Joseph R. Stanek[2], Jonathan L. Finlay[2], Eddy Hernandez-Broncano[1], Sharon Chavez-Paredes[1], Yesenia Miranda-Tunque[1], Adela Heredia-Zelaya[1], Sandro Casavilca-Zambrano[1], Pamela Garcia-Corrochano[1], Luis Ojeda-Medina[1], Enrique Orrego-Puelles[1], Ebert Torres-Malca[1], Raymundo Sernaque-Quintana[1], William Quispe-Valverde[1], Juan Garcia-Leon[1], Diana S. Osorio[2]; [1]Instituto Nacional de Enfermedades Neoplasicas, Lima, Lima, Peru. [2]Nationwide Children's Hospital and The Ohio State University, Columbus, Ohio, USA

BACKGROUND: Medulloblastoma is the most common malignant brain tumor in children. While survival has improved in high-income countries (HIC), the outcomes for patients in low-middle-income countries (LMIC) are unclear. Therefore, we sought to determine the survival of children with Medulloblastoma at the Instituto Nacional de Enfermedades Neoplasicas (INEN) between 1997 and 2013 in Peru. METHODS: Between 1997-2013, data from 103 children older than three years with Medulloblastoma were analyzed. Fourteen patients were excluded. The patients were split into two distinct cohorts, 1997 – 2008 and 2009 – 2013, corresponding with chemotherapy regimen changes. Event-free (EFS) and overall survival (OS) were calculated using the Kaplan-Meier method, while prognostic factors were determined by univariate analysis (log-rank test). RESULTS: Eighty-nine patients were included; median age was 8.1 years (range: 3-13.9 years). The five-year OS was 62% (95% CI: 53 – 74%) while EFS was 57% (95% CI: 48 – 69%). The variables adversely affecting survival were anaplastic histology [compared to desmoplastic; OS: HR=3.4, p=0.03], metastasis [OS: HR=3.5, p=0.01; EFS: HR=4.3, p=0.004], delay in radiation therapy of 31-60 days [compared to ≤30 days; EFS: HR=2.1, p=0.04], and treatment 2009 – 2013 cohort[OS: HR=2.2, p=0.02; EFS: HR=2.0; p=0.03]. CONCLUSIONS: Outcomes for Medulloblastoma at INEN were low compared with HIC. Anaplastic sub-type, metastasis at diagnosis, delay in radiation therapy, and treatment in the period 2009 - 2013 negatively affected the outcomes in our study. Multidisciplinary teamwork, timely delivery of treatment and partnerships with loco-regional groups and colleagues in HIC is likely beneficial.

LINC-24. SURVIVAL AND GENETIC EVALUATION OF PATIENTS DIAGNOSED WITH CHOROID PLEXUS TUMOR TREATED AT A BRAZILIAN INSTITUTION

Milena Oliveira, Nasjla Silva, Andrea Cappellano, Jessica Rodrigues, Daniela Almeida, Sergio Cavalheiro, Patricia Dastoli, Frederico Silva, Maria Teresa Seixas, Fernanda Lima; IOP, São Paulo, SP, Brazil

INTRODUCTION: Choroid plexus tumors (CPT) are rare intraventricular neoplasms of epithelial origin. They usually occur in the 2nd year of life, corresponding to 0.4-0.6% of intracranial tumors in this age group. They are classified, according to WHO 2021: choroid plexus carcinoma (CPC), atypical choroid plexus papilloma (ACPP) and choroid plexus papilloma (CPP). Li-Fraumeni syndrome (LFS) is present in 50% of patients with CPC. In Brazil, the TP53p.R337H mutation affects 0.3% of the population in the South/Southeast. OBJECTIVE: Evaluate the genetic assessment and overall-survival (OS) of patients with CPT treated at IOP/GRAACC/UNIFESP. PATIENTS/METHODS: Between 1995-2021, 48 patients were diagnosed with CPT in our institution, 27 CPC, 18 CPP and 3 ACPP. From 2012-2021 patients were referred for genetic evaluation, 16 of which had CPC (2 had previously CPP). Family history for neoplasms was present in 87.5%, 37.5% compatible with Li-Fraumeni Syndrome (LFS) of which 50% with positive mutations. The molecular investigation of TP53 gene in patients with CPC was performed and positive in 62,5%:R337H(6 patients), R110C*, R158H*, H179R*, R196* (*1 patient each). For those with R337H, p53 protein immunohistochemistry resulted in 90-100% positivity. Treatment strategies consisted of surgery with gross total resection being achieve in all ACPP, 94,4% CPP and 55,5% CPC. Chemotherapy was performed in only 1 case of ACPP and all CPC with a variety of schemas, including high dose of chemotherapy and autologous bone marrow transplant for slow responders or recurrent disease. The three- and five-years OS for CPC was 72.5% and 61.7%, respectively. For CPP and ACPP 5y OS was 100%. CONCLUSION: The present study is in agreement with the literature showing an excellent survival for CPP and ACPP but an inferior outcome for patients with CPC and TP53 mutation despite being assisted by a well-prepared multidisciplinary team with an adequate treatment.