

institutions in Argentina between 2007–2019. Translocation was evidenced in all cases through molecular testing. Clinical characteristics, imaging, histopathology, and treatment response were evaluated. Extracranial and osseous lesions were excluded. RESULTS: 15 patients. Median age at beginning of symptoms: 8 yo (2–20). Most patients had intracranial hypertension syndrome (14/15). In brain MRI, 5/15 supratentorial lesions, 4/15 posterior fossa, 1/15 medullary, 2/15 supra and infratentorial, and 3/15 lesions diffuse leptomeningeal infiltration. Histopathologic findings showed diffuse pattern with small round blue cells in most cases, other patterns were also described. CD99 marked positive in all cases. Misdiagnosis with glial tumors (4/15), medulloblastoma (6/15) and infectious diseases (3/15); led to median delay to accurate diagnosis of 3 months (range 0–67). After correct diagnosis patients were treated with standard ES treatment (6 VIDE cycles plus radiotherapy) in 14/15 patients. Vincristine, irinotecan and temozolamide was used as second line treatment in all relapse cases whenever possible. EFS was 22 months (2–65). OS at 5 years of follow-up was 46.67% (mean OS 31 mo). CONCLUSION: Even though molecular assessment led to accurate diagnosis in all cases, treatment response and outcome showed two different groups of patients with long and very short survival. Adaptive therapy should be considered.

LINC-06. OBSERVATION ONLY IN A PATIENT WITH SUSPECTED LOW GRADE GLIOMA. SHOULD NEUROSURGERY ALWAYS BE THE FIRST STEP IN LOW AND MIDDLE INCOME COUNTRIES?

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BACKGROUND: Low grade gliomas (LGGs) are the most frequent pediatric brain tumor and they comprise a variety of histologies. Complete surgery is curative but sometimes its location makes it difficult. Recent publications highlight the excellent long-term outcomes of patients with LGGs with complete and incomplete resected tumors. Current strategies are focused on reducing risks of treatment related sequelae. METHOD: We describe a patient with a suspected LGG managed by close observation. We describe the case of a 6 year old female with 5 months history of focal onset seizures. During this time a brain MRI was requested and tumor was evidenced. After "tumor diagnosis" was made family visited a handful of private neurosurgeons with a uniformly dismal prognosis and high risk morbidity from procedures offered. When first seen at our Hospital, the clinical history seemed compatible with a LGG and seizures well controlled with antiepileptic drugs. Neurological examination was completely normal. MRI showed a large tumor (7x5x5 cm) hypointense on T1, hyperintense on T2, without contrast enhancement, involving the right temporal lobe white matter, insula, internal capsule, hippocampus, thalamus and mesencephalus with middle cerebral artery encasement. Interval imaging was proposed and after 4.5 years since diagnosis the tumor has been stable and patient clinically excellent. CONCLUSION: Overall survival in pediatric LGGs is excellent and risk of sequelae should always be part of multidisciplinary team considerations. In centers with significant neurosurgical morbidity, biopsy of large tumors that are compatible with LGG may not be required in selected cases.

LINC-07. PREVALENCE AND SPECTRUM OF EARLY ENDOCRINE DISORDERS IN SURVIVORS OF PEDIATRIC EMBRYONAL BRAIN TUMORS (PEBT): EXPERIENCE FROM INDIA

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BACKGROUND: Survivors of pediatric brain tumors are at high risk of developing endocrine disorders, potentially impacting growth, development and quality of life. METHODS: retrospective audit of 2-year survivors of PEBT (3-18 years at diagnosis) viz. medulloblastoma (MB), Central nervous system Primitive neuro-ectodermal tumors (CNS-PNET) and atypical teratoid/rhabdoid tumor (ATRT) treated January 2006-December 2017 at Tata Memorial Centre, Mumbai, with surgery, cranio-spinal irradiation (CSI); 35Gy in high-risk MB, CNS-PNET, ATRT and 23.4Gy in average-risk MB with tumor boost 19.8Gy) and six cycles of adjuvant chemotherapy (cyclophosphamide, cisplatin and vincristine). Patients were followed up by a paediatric endocrinology team specialized in management of PEBT. RESULTS: Of 249 PEBT treated during this period, 88 are alive in remission >2 years (69-MB, 15-CNS PNET, 4-ATRT), median age at diagnosis 6 years. At a median follow-up of 5.6 years (range 3-12.5 years), 63 patients (72%) had at least one endocrine disorder, 26 (29%) ≥2 hormonal deficiencies. The most common endocrine disorders were central hypothyroidism (57%), growth hormone deficiency (40%), central hypogonadism (5%) and central hypoadrenalism (3.5%). The median time to develop hypothyroidism was 2.8 years (range 5 months to 8.5 years) from CSI. Growth hormone replacement therapy began after a median period of 4.2 years (range 1.5 to 11.5 years) from CSI. Higher dose of CSI was associated with development of endocrine

disorder (odds ratio [OR] 2.71; 95% CI, 1.03 to 7.04, p=0.04). CONCLUSIONS: The high incidence of endocrine deficits in survivors of PEBT necessitates early and lifelong monitoring. Early and appropriate management is crucial to achieve full growth potential.

LINC-08. INCREASED TREATMENT TOXICITIES AND INFERIOR OUTCOMES IN UNDERNOURISHED CHILDREN WITH BRAIN TUMOURS

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BACKGROUND: Children on treatment for brain tumours are known to be at high risk of undernutrition, the impact on outcome and toxicity is not well understood. METHODS: Retrospective audit of children (<18 years) diagnosed January 2017-December 2018 with embryonal brain tumours (medulloblastoma, primitive neuro-ectodermal tumors, pinealoblastoma, atypical teratoid/rhabdoid tumour) and treated at our centre. Data was retrieved from case records and electronic medical records. Nutritional status (NS) was defined as per World Health Organization (WHO) into severe malnutrition (SAM), moderate malnutrition (MAM), well nourished (WN) and overweight. Undernutrition (UN) was defined as SAM/MAM. Toxicity was documented till end of treatment, defined as treatment delay >1 week, significant infection or toxic death. RESULTS: Of 124 eligible patients who received entire chemotherapy at our centre, NS data was available in 73 at diagnosis and 58 at follow-up. At diagnosis -29, 16, 26 and 2 and at follow-up -20, 16, 22 and 0 were SAM, MAM, WN and overweight. During treatment, weight gain was documented in 26%, stable weight in 55% and weight loss in 19%. Those UN at diagnosis had worse outcomes at follow-up with 70% alive in remission compared to 88% of WN (p=0.14). There was increased toxicity in UN group (50%) compared to WN (24%), p=0.04. All 3 toxic deaths were in UN. Those who lost weight during treatment had higher toxicities (70%) compared to those with stable weight (30%) or weight gain (20%), p=0.02. CONCLUSIONS: In spite of nutritional intervention, children on treatment for brain tumours tend to lose weight. Increased treatment toxicities and inferior outcomes in undernourished children with brain tumours necessitates proactive and aggressive nutritional monitoring and intervention.

LINC-09. TREATMENT AND OUTCOME IN CHILDREN WITH LOW-GRADE GLIOMAS IN WESTERN MEXICO: EXPERIENCE AT HOSPITAL CIVIL DE GUADALAJARA

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BACKGROUND: Brain tumors are the most common solid tumors in childhood, 35% of them being low-grade gliomas (LGGs). Few data is available regard LGGs in low-and-middle-income countries. This study evaluates LGGs in a tertiary center in Mexico. DESIGN: A retrospective review of clinical files of 105 children diagnosed with LGG other than optic nerve glioma from 2007 to 2019 was done. RESULTS: Median age at diagnosis was 7.2 years (from 5 months to 18 years). Male to female ratio was 0.75:1. WHO Grade I represented 68% of the cases. Anatomic sites were: posterior fossa (41%), supratentorial (43.5%), spinal (8.5%), subependymal (6%) and pineal (1%). Ten percent of patients had a diagnosed phacomatosis. Treatment was observation without surgery in 3.8%, surgery followed by observation in 49.5%, only chemotherapy in 2.8%, only radiotherapy in 6.7%, and surgery combined with chemotherapy or radiotherapy in 37.2% of cases. Among patients who had surgical intervention, 40% achieved gross total resection, 44% subtotal resection and 16% only biopsy. One or more recurrences were found in 20% of patients. The 5 and 10-year overall survival (OS) was 83% and 73% respectively. The 5 and 10-year progression-free survival (PFS) was 66% and 44% respectively. CONCLUSIONS: In this series the OS were lower compared with countries with high income, reflecting the need to improve surgery, since only 40% achieved complete resection that is a determining factor for the prognosis. We observed a decrease in OS until 10-year follow and the PFS was even lower due to recurrence/progression.

LINC-10. SIOP PODC ADAPTED TREATMENT GUIDELINES FOR CRANIOPHARYNGIOMA IN LOW- AND MIDDLE-INCOME SETTINGS

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