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of *SMARCB1*, which encodes a critical component of the SWI/SNF chromatin-remodeling complex. They can arise in the brain where they are called Atypical Teratoid/Rhabdoid Tumors (ATRTs) and in various extra-cranial locations where they are called Malignant Rhabdoid Tumors (MRTs). Both ATRTs and MRTs are frequently lethal diseases for which best therapeutic approaches remains to be determined. Herein, we describe the Sick Kids experience.

Design/Methods: Patients diagnosed with MRT at the hospital for Sick Children between 1996-2016 were identified and charts reviewed. Demographic data, treatment details and survival information was collected.

Results: We identified twenty-five patients diagnosed with MRTs between 1996 and 2016. Median age at diagnosis was 3.2 years (1 month -16.6 years). 64%(n=14) were female and 36%(n=8) were male. Thirty-two percent of the tumors were intra-abdominal (n=8), 24% (n=6) para-spinal, 16% in extremities (n=4), 8% nasopharyngeal (n=2), 8% multifocal (n=2), 4% Skull base (n=1), 4%Chest wall (n=1) and 4% neck (n=1). Most of the patients, 56% presented with advanced disease (stage III and IV). 8% (n=2) of the patients presented with multifocal disease and correspond to patients diagnosed with rhabdoid tumor predisposition syndrome. The median time to progression was 6 months (ranging from 1 month to 25 months). Most of the patients (n=20) were treated with multimodal therapy including chemotherapy in 18 patients, radiation therapy in 11 patients and surgery in 14 patients. Only 20% of patients are alive and free of disease. 64% of patients succumbed to the disease.

Conclusions: MRTs continues to be a disease with poor outcomes. Young patients and patients presenting with metastatic disease have a dismal prognosis. Insights in the biology of this entity are urgently needed to guide therapeutic approaches and the development of clinical trials.

P-272 | Paratestitular Rhabdomyosarcoma in Children: Surgical Analysis

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Background/Objectives: In children, about 7% of all cases of genitourinary rhabdomyosarcoma are paratesticular in origin. Prognosis for paratesticular rhabdomyosarcoma (RMS) is favorable; approximately 60% to 80% of patients have localized disease at diagnosis. The purpose of this study was to report a retrospective review of paratesticular RMS in children treated at the Pediatric Oncology Department at one institution in Brazil. The prognostic, surgical and histopathologi aspects were analyzed.

Design/Methods: From 1987 to 2017, a total of twenty fiv patients with ages ranging from 10 months to 16 years old wit pathologically confirmed diagnosis of paratesticular RM: treated at our institution.

Results: The median age at presentation was 12 years (rang 10 months – 16 years). Sixteen patients were ≥10 years ol (76%). Eighteen patients had initial surgery at non-oncologi hospital and seven needed a second surgery. One third o patients who underwent initial surgery outside our institu tion required second surgery (three hemiscrotectomy and fou lymphadenectomy). Partial cystectomy was performed in on patient due to vesical metastasis. The histopathological classi fication of RMS was: embryonal in 19, alveolar in four and no classified RMS in two. The patients were treated following th International Rhabdomyosarcoma Study Group from 1987 t 2008 (88%) and European Pediatric Soft Sarcoma Group pro tocol since 2009 (12%). All patients received chemotherapy and only seven received radiotherapy. The 5-year overall sur vival (OS) was 76%, with follow up from 6 months to 20 years (median 4 years). The mortality among patients ≥10 years of age was 37,5%, while all patients < 10 years are alive.

Conclusions: The 5-year OS was worse in patients ≥10 years old, confirming current data. The patients who underwent initial surgery at non-oncologic hospital needed second surgery more frequently, including hemiscrotectomy. Continued effort is required to educate providers on appropriate workup of scrotal masses to avoid scrotal violation.

P-273 | Our Experience with Primary Total Thyroidectomy and Lymph Node Biopsy in Papillary Thyroid Carcinoma

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Background/Objectives: There are concerns about the optimal primer surgery in case of papillary thyroid carcinoma. From 2006 we moved on the total thyroidectomy and regional lymph node biopsy as the first attempted procedures in all cases, to provide opportunity for postoperative iodine isotope therapy. There are few evidences about the results of this procedure.

Design/Methods: We retrospectively analyzed the data of the children underwent primary total thyroidectomy in our unit from 2006-2016.

Results: 29 patients were found, the mean age 15.3 year (7-20), M/F ratio 8/21. All had papillary thyroid carcinoma