PNR-33. MOLECULAR RE-EVALUATION OF INSTITUTIONALLY DIAGNOSED CNS-PNETs: CLINICAL CONSEQUENCES OF CONFINED DIAGNOSTIC GROUPS

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Primitive neuroectodermal tumors of the central nervous system (CNS-PNETs) are highly aggressive, poorly differentiated embryonal tumors occurring predominantly in young children but also affecting adolescents and adults. Recently, by utilizing DNA methylation, it has been demonstrated that a large proportion of institutionally-diagnosed CNS-PNETs display molecular profiles indistinguishable from those of various other well defined CNS tumor entities, including high grade gliomas, AT/RTs, and ependymomas. Among institutionally-diagnosed CNS-PNETs that do not align with other tumor types, four distinct molecular entities were defined, each associated with a recurrent genetic alteration and distinct histopathological and clinical features: CNS-NR-FOXO2, CNS-HGNET-BCOR, CNS-HGNET-MN1, and CNS-EFT-CIC (Sturm et al., 2016). Interestingly, after identifying these new entities, additional cases aligning with these profiles were found among patients with historic diagnoses other than CNS-PNETs. In order to develop appropriate treatment strategies for these new molecular entities, it is essential to know the clinical response and outcome from previously applied treatment strategies. Conversely, it is valuable to analyze the outcome of patients with tumors treated as per CNS-PNET strategies that were reclassified into other known entities and compare these data to the clinical and outcome data from the group they were re-classified to. Within a broad international collaborative approach, clinical, molecular and histopathological data are collected for patients with historic diagnoses of CNS-PNET or with tumors not diagnosed as CNS-PNET but that fit the molecular profile of one of the newly defined molecular entities. Clinical and outcome data will be presented for patients with matched molecular profiles.