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Embryonal tumor with multilayered rosettes (ETMR), a newly coined term, including embryonal tumor with abundant neuropil and true rosettes (ETANTR), ependymoblastoma and medulloepithelioma, encompasses nearly 25% of all CNS-PNETs. To date, no clinical trial or consensus treatment approach has been reached on how to treat this unique clinical group. To better describe the clinical spectrum we have undertaken a retrospective literature review of all published cases of ETMR, PNET-1, ependymoblastoma and medulloepithelioma as well as our centre’s own clinical series with C19MC amplified tumors. By analyzing these patients together, it is an important start to critically evaluate current treatment approaches and guide our future study design into clinically rational directed therapy. A total of 206 unique cases were extracted of which 179 had follow-up and outcome data available. 3 year overall survival was 21% (95% C.I. 15% - 29%) and no deaths were reported after 3 years. Among the 179 patients with outcome details available, age over 4 was significantly associated with superior outcome on univariate analysis (p < 0.01). Among the 137 patients with radiotherapy and outcome data available there was a significant association between delivery of radiotherapy and improved survival (p < 0.0001). Among the 137 patients with chemotherapy and outcome data available high dose chemotherapy was also significantly associated with improved survival (p < 0.02). Among the 140 patients with surgical and outcome details available, gross total resection was associated with superior outcome (p < 0.0001). Ultimately a clear clinical picture will only become apparent with prospective enrolment in clinical trials based on C19MC amplification.