

FINAL CATEGORY: EMBRYONAL TUMOR WITH MULTILAYERED ROSETTES (ETMR), OTHER EMBRYONAL TUMORS, RARE TUMORS

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ETMR-01. BRAIN STEM TUMORS IN CHILDREN LESS THAN THREE MONTHS: CLINICAL AND RADIOLOGIC FINDINGS OF A RARE DISEASE

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BACKGROUND: Brainstem gliomas account for 10-20% of all central nervous system pediatric neoplasms. Brain stem tumors in children less than three months old at diagnosis are extremely rare. Our aim is to study a retrospective cohort to improve the understanding of the disease course and guide patient management. **METHODS:** This is a multicenter retrospective analysis across the European Society for Pediatric Oncology SIOP-E HGG/DIPG Working Group linked centers and included patients with a brainstem tumor diagnosed between 2009 and 2020 and aged less than 3 months at diagnosis. Clinical data was collected, and imaging characteristics were analyzed blindly and independently by two experienced neuroradiologists. **RESULTS:** Five cases (female; n=4) were identified of which no patient received any therapy. The epicenter of 2 tumors was in the medulla oblongata alone and in the medulla oblongata and the pons in 3 cases. For patients with tumor in equal parts in the medulla oblongata and the pons (n=3) the extension at diagnosis involved the spinal cord; for the 2 patients with the tumor epicenter in the medulla oblongata alone (n=2) the extension at diagnosis included the pons (n=2) and the spinal cord (n=1). Biopsy was performed in one patient identifying a pilocytic astrocytoma. Two patients died rapidly. In one patient autopsy revealed a high-grade glioma (case 3). Three survivors showed either spontaneous tumor regression (n=2) or stable disease (n=1). The surviving patients were followed-up for 10, 7, and 0.6 years, respectively. One case had the typical imaging characteristics of a dorsal exophytic low-grade glioma. **CONCLUSIONS:** No patient fulfilled the radiologic criteria defining a high-grade glioma. Central neuroradiological review and biopsy (if feasible) may provide useful information with regards to the patient's management.