months. CONCLUSION: Our findings showed that pseudoprogression can occur early in the treatment course in CMMRD patients. Identification of this entity is important for appropriate clinical management.

RARE-16. SEVEN CASES OF RETINOBLASTOMA WITH CNS INVOLVEMENTS

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Treatment strategy for trilateral retinoblastoma (TRb: very rare RB with brain tumor) or retinoblastoma with central nervous system (CNS) involvement is not established yet. We retrospectively reviewed our seven cases of these rare almost fatal tumors. Their ages at diagnosis are 0y3m-1y10m (median 1y3m) (Male 4, Female 3). Only one had RB family history. Their affected eyes were bilateral 3, unilateral 3 and no 1. Their CNS involvements were suprasellar tumor 4, pineal tumor 1 and cerebrospinal fluid (CSF) cytology positive 2. Three of the suprasellar tumor patients had spinal metastasis. Four of the seven patients were TRb and one were genetically classified suprasellar retinoblastoma. All of them were treated with chemotherapy and four received high-dose chemotherapy. Three brain tumors of four TRb almost disappeared with chemotherapy. Two of them also received radiotherapy but relapsed. Although one radiation-free long-term TRb survivor developed secondary osteosarcoma, he got remission again and live 5 more years. One CSF positive Rb patient with chiasm invasion died of disease 11 months later. The other patient had no chiasm invasion nor CSF involvement at diagnosis, but his CSF cytology turned to positive after his second cycle of chemotherapy. He got remission with radiotherapy and high-dose chemotherapy, and alive without disease for 4 years. 2-year RFS and 2-year OS of all patients were 40% and 60%. Although our TRb patients responded to chemotherapy, it was difficult to avoid radiotherapy except one. Data accumulation is necessary for better treatment of these cancer-predisposed patients.

RARE-17. SURVIVAL BENEFIT FOR INDIVIDUALS WITH CONSTITUTIONAL MISMATCH REPAIR DEFICIENCY SYNDROME AND BRAIN TUMORS WHO UNDERGO SURVEILLANCE PROTOCOL. A REPORT FROM THE INTERNATIONAL REPLICATION REPAIR CONSORTIUM

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