Atypical Teratoid/Rhabdoid Tumors in the Cerebellar Vermis:
A Case Report

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Abstract: An atypical teratoid/rhabdoid tumor (AT/RT) is a rare pediatric brain tumor that clinicopathologically resembles medulloblastoma, but it is a more frequently occurring pediatric brain tumor. Making a differential diagnosis to distinguish between these tumors is sometimes difficult. We herein present a case of AT/RT occurring in the cerebellar vermis, which is the most common location for medulloblastomas. A nine-month-old boy presented with symptoms of repeated vomiting. Head computed tomography and brain magnetic resonance imaging scans showed a mass which was located in the cerebellar vermis which was associated with obstructive hydrocephalus and multiple disseminations. The initial clinical diagnosis was medulloblastoma, and a subtotal resection was thus performed. Histopathologically, the tumor was an admixture of basophilic or darkly stained portions and pale or lightly eosinophilic portions. The former consisted of small round cells with hyperchromatic nuclei and scant cytoplasm, which thus resembled medulloblastoma. The latter comprised tumor cells with vesicular nuclei, distinct nucleoli and pale cytoplasm, which thus resembled a large cell medulloblastoma. Rhabdoid cells with eosinophilic cytoplasm were also locally found. Necrosis, calcification and numerous mitoses were also noted. Based on these histological findings, the differential diagnosis included medulloblastoma with large cell portions vs AT/RT. Immunohistochemically, the tumor cells were positive for cytokeratin AE1/AE3, glial fibrillary acid protein (GFAP) and epithelial membrane antigen (EMA), while they were negative for INI1, thus supporting a diagnosis of AT/RT. In the present case, the application of INI1 immunostaining was very helpful for distinguishing AT/RT from medulloblastoma.

Key words: AT/RT, Cerebellar vermis, Differential diagnosis, INI1