ABSTRACT

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Primary leptomeningeal medulloblastoma: a case-based review.

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BACKGROUND: Medulloblastoma (MB) is the most common malignant pediatric brain tumor, accounting for 40% of childhood tumors in posterior fossa. Metastatic disease, occurring in 20-30% of all medulloblastoma cases at diagnosis, is largely exclusive to the leptomeninges. On the contrary, primary leptomeningeal medulloblastoma or so-called chameleon medulloblastoma, defined by the absence of a detectable intraparenchymal lesion with a widespread diffusion along leptomeninges, is a rare entity of difficult diagnosis with only a few cases reported in literature.

METHODS AND RESULTS: A comprehensive literature search of three databases (PubMed, Ovid Medline, and Ovid Embase) have been conducted to identify pertinent papers focusing on the diagnostic process, management, and treatment of primary leptomeningeal medulloblastoma and its peculiar features. To our knowledge, only eight cases are described in literature, including five pediatric patients and three adults, two of which with the initial involvement of the spinal cord. In addition, we report another two pediatric cases, showing widespread primary diffusion along leptomeninges of brain and spinal cord. Finally, we analyze in-depth the peculiar morphological MRI features of this tumor.

CONCLUSION: The classification and treatment of medulloblastomas are likely to change in the coming years due to new insights into the molecular biology of medulloblastoma. Primary leptomeningeal medulloblastoma could represent another potential challenge for biologists to start exploring the underlying mechanisms of this different clinical and pathological entity, with different implications for diagnosis and its management.

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