Medulloblastoma with extensive nodularity

Treatment for medulloblastoma carries significant risks, particularly in resource-constrained settings.

Aristizabal et al. report a case of a Mexican infant with desmoplastic/nodular medulloblastoma. Given the nature of her tumor, we developed a tailored regimen following subtotal resection to avoid both radiation therapy and the high-dose cisplatin therapy offered at most centers in the United States. The patient is in remission 4 years after the initial diagnosis. This case suggests an alternative treatment plan for this particular tumor variant that accommodates the limited resources of many centers around the world and avoids the risks associated with radiation therapy at a young age [1].

Four cases of medulloblastoma with extensive nodularity (MBEN) are described. The patients were 3 years of age or younger at diagnosis. Cranial CT scan disclosed multiple coalescing nodules with peculiar 'grape-like' architecture in three patients. A near total excision of the tumour was performed in all four patients. The patients are currently receiving radiation and chemotherapy. Histologically, the tumours were characterized by extreme nodularity with intranodular uniformity and low proliferative index. The internodular zones were extremely cellular, composed of undifferentiated, mitotically active cells. Bcl-2 protein expression was observed in the cellular zones but was distinctly absent within nodules.

MBEN represents a variant that occurs in very young children but has a good prognosis. The favourable outcome is probably related to its spontaneous neurocytic differentiation. The pattern of Bcl-2 immunoreactivity in MBENs indicates that this protein could be a key player in the regulation of neuronal differentiation in medulloblastomas [2].

References

1)
Aristizabal P, Burns L, Rivera-Gomez R, Levy ML, Roberts W, Crawford JR. Medulloblastoma With Extensive Nodularity: Tailored Therapy in a Low-resource Setting. J Pediatr Hematol Oncol. 2017 Mar 6. doi: 10.1097/MPH.0000000000000798. [Epub ahead of print] PubMed PMID: 28267079.
2)
Suresh TN, Santosh V, Yasha TC, Anandh B, Mohanty A, Indiradevi B, Sampath S, Shankar SK. Medulloblastoma with extensive nodularity: a variant occurring in the very young-clinicopathological and immunohistochemical study of four cases. Childs Nerv Syst. 2004 Jan;20(1):55-60. Epub 2003 Dec 5. PubMed PMID: 14657995.