Well-differentiated pediatric glial neoplasms with features of oligodendroglioma, angiocentric glioma and dysembryoplastic neuroepithelial tumors: a morphological diagnostic challenge.

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Abstract

OBJECTIVE: Oligodendrogliomas are rare in the pediatric population, and most oligodendroglioma-like tumors in this age group may belong to other entities. In addition, accurate diagnosis and grading of such lesions using criteria developed for adult oligodendrogliomas prove difficult, and often controversial.

MATERIAL AND METHOD: During a study of tumors previously diagnosed as pediatric oligodendroglioma, we identified four tumors displayed features of that resembled oligodendroglioma, angiocentric glioma and dysembryoplastic neuroepithelial tumor but could not be classified as either one of these entities. Their clinical, histological and immunohistochemical features of these cases were investigated in this study.

RESULTS: Two male (both 9 years old) and two female (ages 4 years and 20 months) patients presented with new onset of seizures. All patients were treated surgically, and two required reoperation. Histologically, the tumors were well-differentiated glial neoplasms with focal angiocentric pattern, delicate vascularity, diffuse growth, infiltrative margins, cortical nodules, focal myxoid areas, and leptomeningeal extension. Immunohistochemical studies showed diffuse nuclear positivity with Olig-2 and GFAP antibodies, whereas staining with neuronal markers, EMA, p53, and IDH1 were negative. Fluorescent in-situ hybridization analysis demonstrated intact 1p/19q in all tumors, and there was no ultrastructural evidence of ependymal differentiation. All patients were alive with disease with a mean follow-up of 112 months.

CONCLUSION: These four cases illustrate the morphological diversity of well-differentiated, oligodendroglioma-like glial neoplasms and the uncertainty in their classification among pediatric tumors.


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