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RE: DIFFUSE LEPTOMENINGEAL GLIONEURONAL TUMORS: A NEW ENTITY?

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We read with great interest the paper by Gardiman and colleagues regarding a pediatric series of four unusual neuroectodermal tumors presenting with diffuse leptomeningeal involvement in the "absence" of intraparenchymal lesions (5). The authors believe that cases with similar clinical and pathological features have been previously reported as diffuse leptomeningeal oligodendrogliomas (1–4, 6, 7, 10). However, based on the morphological and immunophenotypical findings, the authors suggest that these tumors might represent a new distinct nosological entity, which they called "diffuse leptomeningeal glioneural tumors." This new entity, typical of the pediatric age, would present a distinct radiological pattern and, based on their experience, would be characterized by an indolent clinical course.

We were surprised to find described in this series case 3, a case which we have studied and reported separately as leptomeningeal oligodendroglioma (9). We are fully familiar with the case, as most of the magnetic resonance imaging and both surgeries were performed in the hospital of Treviso. Therefore, we had the opportunity to study both the original biopsy, as well as the second biopsy. At difference from what reported, we find that the second biopsy shows an anaplastic tumor with a high mitotic index (8 mitoses/10 HPF) and a remarkably high MIB-1 labeling index (up to 50% of neoplastic cells). Furthermore, the patient expired as a consequence of the tumor. In our hands, despite the unusual morphological appearance of "pseudorosettes," there was no definitive evidence of neuronal differentiation by either immunohistochemistry or electron microscopy. We wonder if the "Neu-N positive ganglioid" cells described may represent entrapped distorted preexisting neurons rather than a true neoplastic component. The FISH studies for 1p19q, performed only on the second biopsy (the material from the first one was very limited), demonstrated a typical 1p19q codeletion (study performed in the Treviso laboratory and confirmed at the Mayo Clinic), as well as a typical translocation t(1p;19q) (q10;p10) as we illustrated in our report. Both findings are well-known distinct molecular features of oligodendroglioma in adults, although they might not be considered absolutely specific. In fact, it is also our experience that one can find 1p19q codeletion in a subset of extraventricular neurocytomas, and this feature seems to correlate with a more aggressive behavior in this tumor type (8). Therefore, on the basis of the morphological and molecular features which this subset of neurocytomas seems to share with the oligodendrogliomas, it is reasonable to hypothesize that these two entities might be pathogenetically related (8). However, we believe that in the particular morphological context of our case, in which the monomorphism and the roundness of the nuclei, the perinuclear halos, the back-to-back cellular arrangement and the mucoid background were highly suggestive of oligodendroglioma, these molecular features do have a confirmatory value. For these reasons, we chose to report the case as a leptomeningeal "oligodendroglioma." While we agree that there is an open controversy regarding the nature of unusual diffuse leptomeningeal tumors, we do not believe that this particular case can be used to support the existence of the reported cases as a new entity as a "glioneural tumor." We may suggest to complete the study of the other three cases reported with the use of additional techniques including electron microscopy and FISH for 1p19q.

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