Glioneuronal tumor

The septum pellucidum is the anatomic site of origin of a spectrum of uncommon neuroepithelial tumors that include central neurocytoma, subependymoma, and low-grade glioneuronal tumors morphologically resembling either dysembryoplastic neuroepithelial tumor (DNT) or Rosette forming glioneuronal tumor (RGNT). Multiple case series or individual case reports have described DNT-like or RGNT-like low-grade glioneuronal tumors of the septum pellucidum and lateral ventricle, which are distinct from the typical cortical and fourth ventricular locations of DNT and RGNT, respectively [1) 2) 3)].

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see Papillary glioneuronal tumor.

see Rosette forming glioneuronal tumor of the fourth ventricle.

see also BRAF V600E mutation in glioneuronal tumor.

Advances in the immunohistochemical detection of neuron-specific and neuronal-associated antigens have resulted in the discovery of neuronal elements in certain primary human brain tumors. The results have been not only to expand what neuropathologists commonly recognize as gangliogliomas, including the tumors now known as glioneurocytic tumor with neuropil rosettes and papillary ganglioneuroma, but also to expand the spectrum of tumor types to now include tumors such as central neurocytoma, dysembryoplastic neuroepithelial tumor, and desmoplastic infantile ganglioglioma. These discoveries have helped us to better understand the biology of these tumors and to refine our classification of them. Distinctions among these tumors include sites of predilection, such as the temporal lobe with the dysembryoplastic neuroepithelial tumors, and a spectrum of clinical aggressiveness that spans indolent “quasi-hamartomatous” lesions, such as the dysembryoplastic neuroepithelial tumor, to high-grade, highly aggressive tumors, such as the supratentorial primitive neuroectodermal tumor (World Health Organization Grade IV). Many of these tumors also commonly exhibit a glial component, as determined by both their histologic appearance and their immunoreactivity for glial fibrillary acidic protein.

For pathologists confronted by this growing array of tumors and subtypes, it is appropriate to focus on them and understand the differential diagnosis to be considered when confronted by them [4)].

Epidemiology

Zulch et al. from Germany in a large series reported that neuronal/glioneuronal tumors accounted for 0.4% (38/9000 cases) of all brain tumors, with similar incidence reported from Japan (0.4%), with higher incidence from Korea (2.1%).

Molecular features

Molecular analysis of these tumors has revealed the absence of IDH1/IDH2 mutation and co-deletion of chromosomes 1p19q that genetically define oligodendroglioma [5) 6)].

Additionally, they have been found to lack alterations in FGFR1, BRAF, MYB, and MYBL1 that

characterize the majority of DNT, RGNT, and other low-grade neuroepithelial tumors [7)]

Case series

see Glioneuronal tumor case series.

Case reports

Composite ganglioma and pleomorphic xanthoastrocytoma with anaplastic features in both components is an extremely rare glioneuronal tumor. 5 cases of anaplastic progression in the glioma component have been reported. These tumors generally affect young patients who suffer brain tumor-related epilepsy, are usually located in the temporal lobe or in the cerebellum, and may associate leptomeningeal spreading. Its current optimal treatment consists of maximal safe surgical resection and adjuvant chemoradiotherapy. Overall survival rate at 5 years is 33% in anaplastic pleomorphic xanthoastrocytoma, and 53% in anaplastic ganglioglioma. CASE DESCRIPTION: The authors describe a progression from ganglioglioma to this composite anaplastic entity after 32 months of follow-up, with apparently non-tumoral parenchyma separating the two components. Polymerase chain reaction revealed a wild-type BRAF gene. 7 months after concomitant chemoradiotherapy, radiological progression lead to a second line of chemotherapy, and a third line of chemotherapy was initiated after a subsequent progression at 11 months. CONCLUSIONS: This case may add some evidence in favor of the glioneuronal maldevelopment hypothesis to explain the oncogenesis of these neuroepithelial tumors [8)].

1)
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3)
Xiong J, Liu Y, Chu SG et al (2012) Dysembryoplastic neuroepithelial tumor-like neoplasm of the septum pellucidum: review of 2 cases with chromosome 1p19q and IDH1 analysis. Clin Neuropathol 31:31–38

4)
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6)
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8)
Rosselló A, Plans G, Vidal-Sarró N, Fernández-Coello A, Gabarrós A. Ganglioglioma progression to combined anaplastic ganglioglioma and anaplastic pleomorphic xanthoastrocytoma. Case report and literature review. World Neurosurg. 2017 Sep 19. pii: S1878-8750(17)31579-6. doi: 10.1016/j.wneu.2017.09.072. [Epub ahead of print] PubMed PMID: 28939541.