Glioneuronal tumor case series

A retrospective study of Liu et al., from University of Tennessee Health Science Center, of LGGs and LGGNTs diagnosed in patients younger than 12 months at St. Jude Children's Research Hospital (1986-2015) was conducted.

For the 51 patients (including 31 males), the mean age at diagnosis was 6.47 months (range, 0.17-11.76 months), and the mean follow-up period was 11.8 years (range, 0.21-29.19 years). Tumor locations were hypothalamic/optic pathway (61%), hemispheric (12%), brainstem (12%), cerebellar (8%), and spinal (8%). There were 41 patients with histological diagnoses: 28 had World Health Organization grade 1 tumors, 6 had grade 2 tumors, and 7 had an LGG/LGGNT not definitively graded. Forty-one patients required an active intervention at diagnosis. Throughout their treatment course, 41 patients eventually underwent tumor-directed surgeries (median, 2 surgeries; range, 1-6), 39 received chemotherapy (median, 2 regimens; range, 1-13), and 21 received radiotherapy. Forty patients experienced disease progression (median, 2 progressions; range, 1-18). Ten patients died of progression (n = 5), malignant transformation (n = 2), a second cancer (n = 2), or a shunt infection (n = 1). The 10-year overall survival, progression-free survival, and radiation-free survival rates were 85% ± 5.3%, 16.9% ± 5.3%, and 51.2% ± 7.5%, respectively. Forty-nine patients experienced health deficits (eg, endocrinopathies, obesity, seizures, visual/hearing impairments, neurocognitive impairments, and cerebrovascular disease). Predictors of progression and toxicities were defined.

Infantile LGG/LGGNT is a chronic, progressive disease universally associated with long-term morbidities and requires multidisciplinary intervention [1].

2016

Data from 35 patients diagnosed with GNTs, including 24 gangliogliomas and 11 dysembryoplastic neuroepithelial tumors, were retrospectively collected. DNA was extracted from GNTs tissues and BRAF V600E mutation was examined by DNA sequencing. The correlations between BRAF V600E mutation and clinical features were analyzed.

Totally, BRAF V600E mutations were detected in 11 patients with GNTs, the rate of mutation were 33.3% and 27.3% in GGs (8/24) and DNTs (3/11), respectively. The probability of BRAF V600E mutation in females (7/12, 58.3%) was significantly higher than that in males (4/23, 17.4%) (P=0.022). Moreover, patients with BRAF-mutated GNTs had a significantly wider variety of seizure types compared to GNTs with BRAF wild-type status (P=0.027). However, no significant correlation between the BRAF status and certain clinical features, such as age of seizure onset, duration of epilepsy, age at surgery, location of the tumor and postoperative seizure free, were observed.

Zhang et al., demonstrated the presence of BRAF V600E mutation in Chinese epileptic patients with GNTs, which was significantly correlated with gender and multiple seizure types. Large sample studies and long-term follow-up are required for further confirmation [2].

2012

Chandrashekhar et al, reviewed 244 cases of neuronal/glioneuronal tumors of the CNS diagnosed over the last decade and they constituted 0.86% of all CNS tumors (244/28061) received in that period.

Mean age at presentation was 25.06 years (range: 1-75 years) with male preponderance (M:F = 1.54 : 1). The majority occurred in third decade (76 cases, 31.4%), with only few cases occurring beyond fifth decade (13 cases, 5.3%). Ganglioglioma/gangliocytoma (94 cases, 38.52%) was the most frequent followed by central neurocytoma (86 cases, 35.24%), paraganglioma (32 cases, 13.52%), dysembryoplastic neuroepithelial tumors (DNET) (21 cases, 8.6%), desmoplastic infantile astrocytoma/desmoplastic infantile ganglioglioma (DIA/DIG) (6 cases, 2.45%), papillary glioneuronal tumor (PGNT) (3 cases, 1.22%) and rosette-forming glioneuronal tumor (RGNT) (1 case, 0.4%). Association with seizures was noted in 40.95% of cases. Glioneuronal tumors are an expanding group of tumors with varying spectra of morphologic patterns and biological behavior. An improved understanding has direct clinical implications for optimizing current treatments and developing novel therapeutic approaches. Although most glioneuronal tumors carry a favorable prognosis, other factors such as inaccessibility to surgical resection and rarely, malignant transformation, make it difficult to accurately predict the biological behavior based on histopathology alone. Reliable prognostic markers remain to be defined [3].

1)
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2)
Zhang YX, Shen CH, Guo Y, Zheng Y, Zhu JM, Ding Y, Tang YL, Wang S, Ding MP. BRAF V600E mutation in epilepsy-associated glioneuronal tumors: Prevalence and correlation with clinical features in a Chinese population. Seizure. 2016 Dec 9;45:102-106. doi: 10.1016/j.seizure.2016.12.004. [Epub ahead of print] PubMed PMID: 27984807.
3)
Chandrashekhar TN, Mahadevan A, Vani S, Yasha TC, Sampath S, Chandramouli BA, Devi BI, Arvinda HR, Shankar SK. Pathological spectrum of neuronal/glioneuronal tumors from a tertiary referral neurological Institute. Neuropathology. 2012 Feb;32(1):1-12. doi: 10.1111/j.1440-1789.2011.01206.x. PubMed PMID: 21410777.

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