Dysembryoplastic neuroepithelial tumor case series

2022

A total of 477 differentially expressed genes (DEGs) (180 upregulated; 297 downregulated) were observed in the epileptogenic cortical tissues (ECTs) compared to non-epileptic controls. Gene ontology analysis revealed enrichment of genes belonging to the following Kyoto Encyclopedia of Genes and Genomes (KEGG) pathways: (i) glutamatergic synapse; (ii) nitrogen metabolism; (iii) transcriptional misregulation in cancer; and (iv) protein digestion and absorption. The glutamatergic synapse pathway was enriched by DEGs such as GRM4, SLC1A6, GRIN2C, GRM2, GRM5, GRIN3A, and GRIN2B. Enhanced glutamatergic activity was observed in the pyramidal neurons of ECTs, which could be attributed to altered synaptic transmission in these tissues compared to non-epileptic controls. Besides glutamatergic synapse, altered expression of other genes such as GABRB1 (synapse formation), SLIT2 (axonal growth), and PROKR2 (neuron migration) could be linked to epileptogenesis in ECTs. Also, upregulation of GABRA6 gene in ECTs could underlie benzodiazepine resistance in these patients. Neural cell-type-specific gene set enrichment analysis (GSEA) revealed transcriptome of ECTs to be predominantly contributed by microglia and neurons. This study provides first comprehensive gene expression profiling of nonneoplastic ECTs of dysembryoplastic neuroepithelial tumors (DNTs) patients and identifies genes/pathways potentially linked to epileptogenesis [1)]

2021

DNTs patients who were admitted to the Department of Neurosurgery of Xiangya Hospital between 1 January 2010 and 31 December 2018 and underwent surgical resection were retrospectively analyzed. Clinical, neuroimaging, and pathological features of DNTs were compared among patients with different outcomes and analyzed using the Kaplan-Meier curves and univariable Cox regression analysis.

Results: Thirty-three DNTs patients were included finally, of which the average age at seizure onset was 11.59 ± 7.46 years old and the average duration of seizures prior to surgical resection was 3.00 ± 4.68 years. After surgical resection, the patients were followed up for 2.39 ± 1.97 years, and 28 patients (84.85%) were seizure-free (class I of the Engel Outcome Scale) while five patients (15.15%) were seizure-continuous (class II or III of the Engel Outcome Scale). When compared with seizure-free patients, seizure-continuous patients had greater age at seizure onset and longer duration of seizures before surgical resection ($p < .05$). No variables were found to be statistically significantly associated with prognosis in univariable Cox regression analysis, but patients with extra-temporal DNTs were found to have better prognosis than those with temporal DNTs (log-rank test $p = .048$).

Elder seizure onset age, longer duration of seizures prior to surgical resection, and a temporal location may be risk factors of poor prognosis for DNTs patients after surgical resection [2)].

2016

Data from 35 patients diagnosed with glioneuronal tumors (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 [3].

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27 patients with drug resistant epilepsy and brain tumor, aged up to 19 years at the time of surgery, were studied between 1996 and 2013 and followed up for at least one year. The mean interval between the onset of seizures and the diagnosis of the tumor was 3.6 years, and from diagnosis to the surgery, 18 months. The location of the tumor was in the temporal lobe in 16, with ganglioglioma and dysembryoplastic neuroepithelial tumors being the most frequent. Among the patients, 92.5% and 90.4% were seizure-free in the first and fifth year after surgery, respectively. Twelve of 16 children were successful in becoming drug-free, with complete withdrawal by 3.2 years. Surgery proved to be potentially curative and safe in these cases, suggesting that the tumor diagnosis and surgery cannot be postponed [4].

2008

A retrospective cohort of 23 patients seen at two major epilepsy centers, with localization-related epilepsy associated with histopathologically demonstrated DNETs. We assessed clinical, electrographic and surgical outcome features in patients with adult- and childhood-onset epilepsy. We were particularly interested in the level of congruence of EEG and MRI data and the need for intracranial recordings. We evaluated seizure outcomes at last follow-up.

The mean age was 33.3 years (range: 5-56 years). Ten patients had adult-onset epilepsy. Thirteen patients (57%) had simple partial, 21 (91%) had complex partial, 16 (70%) had secondarily generalized seizures and 5 patients had only simple partial seizures. Status epilepticus did not occur. Non-enhancing lesions on MRI were located in the temporal lobe in 17 patients, the frontal lobe in 3 patients and the parietal/occipital region in 2 patients. One patient had a DNET that involved both frontal and temporal areas. Ictal scalp EEG and MRI were congruent in 17 patients (74%). Eleven patients (48%) underwent lesionectomies, while the rest required some resection of extralesional cortex as well. Five patients required intracranial EEG. There was no association with cortical dysplasia. Seventeen patients (74%) had an Engel class 1 outcome, in a follow-up period that ranging from 5 to 98 months.

Burneo et al found no difference in outcomes between adult- and childhood-onset cases. Although

epileptogenicity was complex, congruence between electro-clinical and neuroimaging studies was high and allowed good surgical outcomes at 1 year of follow-up [5].

1)
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