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Cerebral resective surgery in eloquent area

Surgery for dysembryoplastic neuroepithelial tumors and gangliogliomas in eloquent areas. Functional results and seizure control

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ABSTRACT

Introduction. – Dysembryoplastic neuroepithelial tumors and gangliogliomas are developmental glioneuronal tumors usually revealed by partial epilepsy. High epileptogenicity, childhood epilepsy onset, drug-resistance, temporal location, and seizure freedom after complete resection are common characteristics of both tumors. We report the specificity of surgical management, functional results and seizure outcome in cases of a tumor location in eloquent areas.

Methods. – Among 150 patients (88 males, 3–55 years) operated on for refractory epilepsy due to a glioneuronal tumor (1990–2015), 30 (20%, dysembryoplastic neuroepithelial tumors = 21; gangliogliomas = 9) had a tumor located in an eloquent cortex (sensory-motor, insular or language areas). Surgery was performed after a preoperative work-up, including stereo-electroencephalography in 48 patients (26%) and functional MRI in 100 (67%). MRI-guided lesionectomy was mainly performed in extra-temporal location, whereas an additional corticectomy was performed in a temporal location. Tumor microsurgical resections were guided using neuronavigation and cortical/subcortical electrical stimulations. Multiple stereotactic thermocoagulations were performed in two insular tumors.

Results. – New motor/language deficits related to eloquent areas occurred postoperatively in 6/30 patients (20%) without any major permanent disability. Minor sensorimotor ($n = 2$) and moderate language disturbance ($n = 1$) persisted in three of them. Postoperative seizure-free outcome (mean follow-up > 5 years) was obtained in 81% of the entire series, but significantly decreased to 60% in eloquent areas. Incomplete tumor resection was the main cause of surgical failure. However, unfavorable seizure outcome was also observed despite complete tumor resection. Malignant transformation occurred in one ganglioglioma.

Conclusion. – Epilepsy surgery for benign glioneuronal tumors in eloquent areas provides acceptable results regarding the functional risks. Complete tumor resection is crucial for long-term favorable outcome.

1. Introduction

Dysembryoplastic neuroepithelial tumors (DNT) and gangliogliomas (GG) are benign glioneuronal tumors (grade I in the WHO classification) [1,2] of neurodevelopmental origin. Due to their high epileptogenicity, DNT and GG are the most common tumors responsible for early onset epilepsy curable by surgery [3–5]. They represent 18% of current epilepsy surgery series in adults and 28% in children [6] and are predominantly located in the temporal lobe.

The proportion of each tumor type varies among the series, depending on criteria for establishing the histopathological diagnosis [5]. Differentiating DNT and GG is crucial as malignant transformation can occur in GG in about 3% of patients operated on for drug-resistant epilepsy [7,8], although it is exceptionally reported in DNT [9]. Despite some controversies, ranges of histopathological DNT subtypes have been recognized. They include the classical form based on the identification of a specific glioneuronal element (SGNE) associated with glial nodules and focal cortical dysplasia (FCD), corresponding to the so-called

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Table 1
Comparison of dysembryoplastic neuroepithelial tumors and gangliogliomas: clinical characteristics, postoperative status and seizure outcome in 150 patients operated on for intractable epilepsy at Sainte-Anne hospital during the period 1990–2015.

Population	DNT ($n=95$)	GG ($n=55$)	Total ($n=150$)	P-value
Gender	36 F, 59 M	26 F, 29 M	62 F, 88 M	0.26
Age at surgery, years (range, mean ± SD)	3–54 (26.2 ± 11.7)	10–55 (27.7 ± 11.2)	3–55 (26.8 ± 11.5)	0.46
Age at epilepsy onset, years (range, mean ± SD)	0.5–44 (12.9 ± 9.1)	1–40 (13.5 ± 8.7)	0.5–44 (13.2 ± 8.9)	0.75
Epilepsy duration, years (range, mean ± SD)	1–43 (13.5 ± 9.8)	1–44 (14.2 ± 10.6)	1–44 (13.7 ± 10.1)	0.64
Tumor lateralization	51 R, 44 L	24 R, 31 L	75 R, 75 L	0.29
Temporal location	67 (71%)	45 (82%)	112 (75%)	0.12
Mesial temporal Location	49 (52%) (21 L, 2 R-RHDL)	38 (69%) (19 L, 1R-RHDL)	87 (58%)	0.04
Extra-temporal location	19 (20%) (frontal = 4, central = 9, parietal = 4, insula = 2[a])	9 (16%) (central = 1, parietal = 4, insula = 3[a], posterior cingulate gyrus = 1)	28 (19%)	0.14
Multilobar location, including the temporal lobe	9 (9%)	1 (2%)	10 (7%)	
Location in eloquent areas	21 (22%)	9 (16%)	30 (20%)	0.4
SEEG	33 (35%)	15 (27%)	48 (32%)	
Follow-up, years (range, mean ± SD)	1–21 (7.2 ± 5.5)	1–20 (5.7 ± 4.8)	1–21 (6.6 ± 5.1)	
Engel I	75 (79%)	46 (84%)	121 (81%)	0.48
Engel IA	46 (48%)	30 (55%)	76 (51%)	0.25
Eloquent area/other locations				
Engel I	12/63 (57–85%)	6/40 (67–87%)	18/103 (60–86%)	0.001
Engel IA	10/36 (48–49%)	5/25 (56–54%)	15–61 (50–51%)	
Antiepileptic drug stop	27 (28%)	13 (24%)	40 (27%)	
Postoperative neurological deficit	10	4	14	
Due to surgical complications	5	3	8	
Due to location in eloquent area	5	1	6 (20%)	
Death during follow-up	3 (cancer = 1, unknown = 2)	1 (malignant transformation)	4 (2.7%)	

DNT: dysembryoplastic neuroepithelial tumors; GG: gangliogliomas; M: males; F: females; R: right; L: left; RHDL: right hemispheric dominance for language; SEEG: stereo-electroencephalography.

[a] Including 1 patient in each group treated by stereotaxic multiple radiofrequency coagulations after biopsy.

"complex form" [10], the "simple form" which presents similar features but is composed of SGNE alone [11] and the "non-specific forms" identified on the basis of the same glial and dysplastic components as in the complex forms but without SGNE [12]. These "non-specific forms" mainly correspond to the "diffuse" forms described in further studies [13,14]. GG are histopathologically characterized by the presence of large ganglion-like neurons and typically exhibit granular bodies and perivascular lymphocytic cuffing [5]. DNT and GG are frequently associated with FCD (type 3b according to the International League Against Epilepsy [ILAE] classification) [15,16] that may play a role in the high epileptogenicity of both tumors. Immunostaining markers (notably CD34 positivity) and chromosomal abnormalities (BRAF mutations) [17] may help to differentiate DNT and GG. However, this may remain difficult despite the use of more specific criteria.

Surgical resection of these tumors is usually followed by a high rate of seizure-free outcome (70–90%). The main prognostic factor for seizure freedom is the completeness of the tumor resection, as reported in most studies [18–28], which may be challenging in functional (eloquent) areas. The term of eloquent areas, commonly used in brain tumor surgery series [29], usually refers to primary motor, sensory, visual and language areas, supplementary motor area (SMA), and insular lobe. The latter is usually included in the eloquent areas because of its proximity and connections with eloquent cortical areas and white matter pathways, and whose surgical injury may cause permanent morbidity. Despite the frequency of eloquent cortex involvement, functional outcome is rarely reported in these specific tumor locations. Here, we discuss the specificity of surgical resection for DNT and GG located in eloquent areas and the results regarding functional and seizure outcomes based on a 25-year-long experience in our epilepsy surgical center.

2. Methods

2.1. Population and tumor location

Patient characteristics and tumor location are summarized in Table 1.

Based on the histopathological criteria mentioned above, 150 patients (88 males, mean age: 26.8 years; range, 3–55) underwent a surgical resection for drug-resistant partial epilepsy associated with DNT (95 cases) or GG (55 cases) in our center between 1990 and 2015. Mean age at surgery, age at seizure onset and preoperative duration of epilepsy were similar in the two groups. Male predominance (ratio: 1.6) was found in DNT patients. In most cases, epilepsy onset occurred during childhood or in young adults (mean: 13.2 years). Temporal location of the tumor was predominant in both DNT and GG groups (71% and 82% respectively) and equally found in both hemispheres. A mesial temporal lobe involvement was significantly more frequent in GG than in DNT (69% versus 52%, $P=0.04$). Conversely, extra-temporal and multilobar locations were more frequent in DNT than in GG without reaching significance (29% versus 18%, respectively, $P=0.14$). Eloquent areas were involved in 30 patients (neocortical temporal areas in 6 patients; extra-temporal areas in 24 patients, including 10 central, 8 parietal, 5 insular, and 1 multilobar cases).

2.2. Preoperative investigations

All patients underwent presurgical investigation including detailed analysis of ictal semiology, video-EEG monitoring, multisequence magnetic resonance imaging (MRI) at 1.5 Tesla since 2000 and 3 Tesla since 2010, and neuropsychological assessment. Motor, sensory, language or visual functional MRI (fMRI) and ^{18}F-fluorodeoxyglucose positron emission tomography (^{18}FDG-PET)