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Case series

Temporal lobe gangliogliomas associated with chronic epilepsy: Long-term surgical outcomes

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ABSTRACT

Objective: To review the clinical features and surgical outcome in patients with temporal lobe gangliogliomas associated with intractable chronic epilepsy.
Methods: The Rush University Surgical Epilepsy Database was queried to identify patients with chronic intractable epilepsy who underwent resection of temporal lobe gangliogliomas at Rush University Medical Center. Medical records were reviewed for age of seizure onset, delay to referral for surgery, seizure frequency and characteristics, pre-operative MRI results, extent of resection, pathological diagnosis, complications, length of follow-up, and seizure improvement.
Results: Fifteen patients were identified. Average duration between seizure onset and surgery was 14.3 years. Complex partial seizures were the most common presenting symptom. Detailed operative data was available for 11 patients – of these, 90.9% underwent complete resection of the amygdala and either partial or complete resection of the hippocampus, in addition to lesionectomy. Average follow-up was 10.4 years (range 1.6–27.5 years), with 14 patients improving to Engel's class I and one patient to Engel's class III. There were no recurrences, and permanent complications were noted in one patient.
Conclusions: Long-term follow-up of patients with temporal lobe gangliogliomas associated with chronic intractable epilepsy demonstrates excellent results in seizure improvement with surgery and increasingly low incidence of complications with improvements in microsurgical techniques.

1. Introduction

Ganglioglioma was first described by Courville in 1930 as a tumor composed of both glial cells and differentiated nerve cells [1]. Generally it is described as a relatively low-grade malignancy with an indolent clinical course. It is also one of the most common tumors causing intractable epilepsy – it has been found to be responsible for 37–51% of intractable epilepsy cases due to neoplasm in the literature [2–5]. Conversely, seizures were the presenting symptom for gangliogliomas in 63–85% of patients [6–9]. Surgical treatment of gangliogliomas has found success rates in patients achieving freedom from seizures ranging from 59% to 84% [2,8,10,11]. Of all CNS gangliogliomas, the incidence occurring in the temporal lobe was found to be 39% [7], and other tumor case series have shown temporal lobe gangliogliomas to cause epilepsy in 64–72% of cases [7,8]. We present the long-term surgical outcomes of patients with intractable epilepsy, shown pathologically to have gangliogliomas.

2. Methods

The Rush University Surgical Epilepsy Database was queried to identify patients undergoing temporal lobe tumor resections to treat intractable epilepsy in whom pathology revealed ganglioglioma from 1973 to 2009. All operations were performed by one of two surgeons (RWB and WWW). Patients were required to have follow-up sufficient to calculate an Engel's score (>1 year) to be included. Medical records were reviewed for age of seizure onset, delay to surgery, seizure frequency and characteristics, extent of resection, complications, length of follow-up, seizure improvement based on Engel's criteria, and post-operative MRI findings.

Preoperatively, non-invasive video electroencephalography, neuropsychological examination, and Wada testing were performed on all patients per our center's surgical epilepsy protocol.

In tumors involving the dominant temporal lobe, intraoperative or subdural cortical stimulation mapping localized speech function. After initial electrocorticography, a transcortical approach,

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Table 1
Characteristics of patients with a temporal lobe ganglioglioma undergoing surgery.

Case	Age (years), sex	Side	Years from seizure onset to surgery	Pre-op seizure frequency (per month)	Seizure type	Last follow-up (years)	Engel's score	Complications	Recurrence
1	8, F	L	7	300	Generalized	27.4	Ic	None	No
2	17, F	R	9	300	Complex partial	1.6	I	None	No
3	26, M	R	6	45	Complex partial	4.3	I	None	No
4	7, M	R	6	N/A	Complex partial	3.4	I	None	No
5	15, F	R	6	N/A	Simple partial	16.2	III	None	No
6	19, M	L	10	30	Complex partial	6.1	Ia	None	No
7	17, M	L	3	N/A	Complex partial	16.2	Ia	Hemiparesis	No
8	34, F	L	30	10	Simple partial	9.0	I	None	No
9	33, M	R	18	10	Complex partial	15.1	Ib	None	No
10	41, M	L	16	2	Complex partial	12.7	Ib	None	No
11	49, M	L	23	90	Complex partial	12.8	Ic	None	No
12	38, M	L	18	2	Complex partial	11.6	Ia	None	No
13	19, F	R	16	2	Complex partial	4.7	Ia	None	No
14	22, M	R	21	20	Simple partial	7.9	Ia	None	No
15	27, M	L	26	3	Complex partial and secondary generalized	6.3	Ia	None	No

with or without lateral cortical resection, was used to gain access to the tumor. The primary goal of the operation was a gross-total resection of the lesion, and was aggressively pursued under the microscope. Further resection of epileptogenic temporal lobe tissue, the amygdala and hippocampus was then performed based on pre-operative testing, intraoperative electrocorticography and the location/infiltration of the tumor. In distinguishing incidental epileptic activity as compared to epileptogenic foci intraoperatively, simple spiking as noted on electrocorticography after lesionectomy was not addressed with surgical resection. However, persistent mesial temporal spiking that produced a broad spreading field was addressed with further resection in some cases.

The amygdala was resected to the superior level of the endorhinal sulcus. If focal tumor extended above this level, it was resected if it did not surround the anterior choroidal artery or the optic tract. A partial hippocampal resection extended to the level of the choroidal point and a resection of the head and body extended to the ambient cistern, where the tail of the hippocampus recedes medially around the thalamus.

3. Results

Fifteen consecutive patients were identified with a temporal lobe ganglioglioma and intractable epilepsy undergoing surgery between 1983 and 2005. This represents 38.5% of temporal lobe tumors causing intractable epilepsy in the Rush University Surgical Epilepsy Database. Table 1 shows details of the 15 identified cases. The group had a 2:1 male:female ratio. Mean age of seizure onset was 10.5 years and delay to surgery was 172 months. The mean age at surgery was 24.8 (range 7–49). Complex partial seizures were the most common presenting symptom, shown in 11 patients, and only one of these patients showed secondary generalized seizures. In patients with known preoperative seizure frequency, the median was 68 seizures per month (range 2–300 per month).

Preoperative MR imaging was preformed on 14 patients, in all of whom the tumor was hypointense on T1 sequences and hyperintense on T2 sequences. Cystic changes were observed in 2 patients, and enhancement was found in 3 patients. Typical MR imaging studies are shown in Fig. 1, demonstrating a right temporal lesion that is hypointense on T1-weighted imaging and hyperintense on T2-weighted FLAIR imaging. All tumors occurred in the medial temporal lobe. No patients required preoperative subdural and/or depth electrodes to achieve satisfactory localization for surgical planning.

Resection was carried out on the left side in 8 patients, and on the right in 7 patients. Complete lesionectomy was performed in all patients. The extent of amygdalohippocampectomy is known in 11 patients. Eight of these patients underwent complete resection of the amygdala with partial resection of the hippocampus. The remaining three patients had either sparing of the amygdala, sparing of the hippocampus, or complete resection of the hippocampus. 13 of 15 patients had postoperative MRI. Fig. 2 shows typical

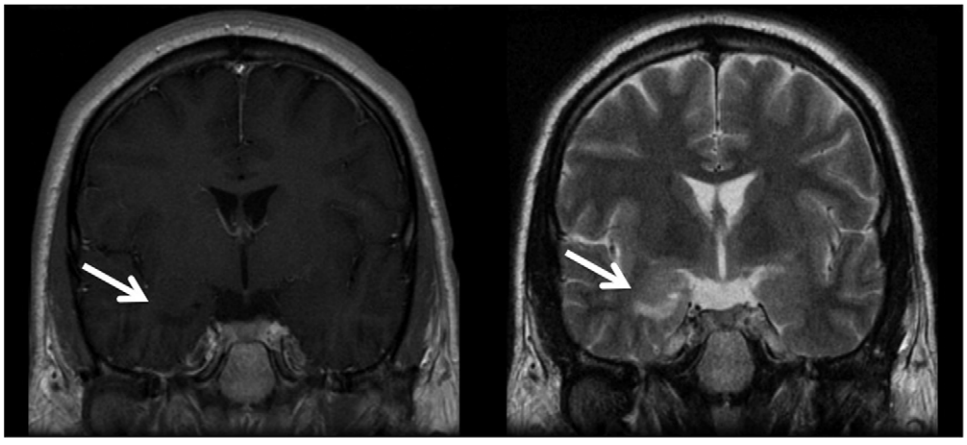

Fig. 1. Representative preoperative MR images showing a patient with intractable epilepsy and a right temporal lobe ganglioglioma. The lesion is hypointense on T1 (arrowed, left) and hyperintense on T2 (arrowed, right).