Surgical treatment of occipital lobe epilepsy

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Object. Occipital lobe epilepsy (OLE) accounts for a small percentage of extratemporal epilepsies and only few and mostly small patient series have been reported. Preoperative findings, surgical strategies, histopathological bases, and postoperative outcomes for OLE remain to be elucidated.

Methods. A group of 54 patients with occipital lobe involvement were identified from a prospective epilepsy surgery database established in 1989. Medical charts, surgical reports, MR imaging, and histopathology data were reviewed, and patients with additional temporal and/or parietal involvement were categorized separately. Seizure outcome was classified according to the Engel classification scheme (Classes I–IV). Two patients were excluded due to incomplete data sets. Fifty-two patients with intractable epilepsy involving predominantly the occipital lobe were included in the study, comprising 17.8% of 292 patients undergoing operations for extratemporal epilepsies.

Results. In nearly all cases (50 [96.2%] of 52), a structural lesion was visible on preoperative MR imaging. Of these cases, 29 (55.8%) had “pure” OLE with no temporal or parietal lobe involvement. Most patients (83%) had complex partial seizures, and 60% also had generalized seizures. All patients underwent occipital lesionectomies or topectomies; 9 patients (17.3%) underwent additional multiple subpial transections. Histopathology results revealed 9 cortical dysplasias (17.3%), 9 gangliogliomas (17.3%), 6 other tumors (11.5%), 13 vascular malformations (25%), and 15 gliotic scars (28.8%). Visual field deficits were present in 36.4% of patients preoperatively, and 42.4% had new or aggravat-ed visual field deficits after surgery. After a mean follow-up of 80 months, 36 patients were seizure free (69.2% Engel Class I), 4 rarely had seizures (7.7% Engel Class II), 8 improved more than 75% (15.4% Engel Class III), and 4 had no significant improvement (7.7% Engel Class IV). Multifactorial logistic regression analysis revealed that early age at epilepsy manifestation (p = 0.031) and shorter epilepsy duration (p = 0.004) were predictive of better seizure control. All other clinical and surgical factors were not significant in predicting outcome.

Conclusions. Occipital lobe epilepsy is an infrequent but significant cause of extratemporal epilepsy. Satisfactory results (Engel Class I or II) were obtained in 77% of patients in our series. Postoperative visual field deficits occurred in a significant proportion of patients. In the modern MR imaging era, lesions should be investigated in patients with OLE and lesionectomies should be performed early for a better outcome. (DOI: 10.3171/JNS/2008/109/7/0057)

KEY WORDS • dysplasia • epilepsy • ganglioglioma • gliosis • occipital tumor • visual field

Abbreviations used in this paper: AVM = arteriovenous malformation; CPS = complex partial seizure; ECoG = electrocorti-coграфy; EEG = electroencephalography; FDG-PET = fluorine-18–labeled fluorodeoxyglucose–PET; IED = interictal epileptiform discharge; MST = multiple subpial transection; OLE = occipital lobe epilepsy; SPS = simple partial seizure; TLE = temporal lobe epilepsy; TSE = turbo spin echo; WHO = World Health Organization.

OCCIPITAL lobe epilepsy accounts for a small percentage of extratemporal epilepsies. Depending on selection and inclusion criteria, the frequency of OLE varies between 2 and 13% of symptomatic partial epilepsies.5,6,23,40 Few large series of patients with OLE have been reported so far.2,10,25,56 Moreover, some patient series cover time periods before the introduction of MR imaging.3,5,8,30,31,36 In one of the largest patient series, 42 occipital resections were performed between 1930 and 1991 at the Montreal Neurological Institute.21 In the modern era, however, preoperative diagnostic findings, surgical strategies, histopatho-
drome of nontumoral occipitotemporal epilepsy has also been defined. In addition, visual auras can also occur in temporomestal and temporoccipital seizures. Scalp EEG recordings are often not helpful or even misleading, but clearly lateralizing semiology was found to be predictive for satisfactory seizure relief. The identification of a lesion with high-resolution MR imaging and additional imaging modalities such as SPECT and FDG-PET play a major role in the diagnostic workup of patients with presumed OLE. For the definitive discrimination between OLE and TLE, invasive video-EEG monitoring using intracranial subdural and/or depth electrodes is often mandatory.

We describe a large series of consecutive patients who underwent occipital resections for lesional OLE during the era of modern MR imaging and video-EEG monitoring. We analyzed preoperative findings, surgical strategies, histopathological bases, and postoperative outcomes in this infrequent type of extratemporal epilepsy.

Methods

Study Population

From a prospective epilepsy surgery database established at the University of Bonn in 1989, we identified patients who underwent surgery for OLE between 1990 and 2005. Minimal requirements for inclusion in the study were as follows: 1) clinical history of medically intractable epilepsy; 2) preoperative MR imaging or histopathological results positive for occipital lobe involvement; 3) complete clinical and electrophysiological data sets; and 4) follow-up seizure outcome data. Of 292 patients undergoing surgery for extratemporal epilepsy in our database, we identified 54 patients (18.4%) with occipital lobe involvement. Of these 54 patients, 52 had adequate data for analysis. To determine the localization and extent of the lesion, MR imaging, operative reports, and histopathology data were reviewed. This review yielded 29 patients with “pure” OLE (9.9% of the total extratemporal epilepsy population in our series), 9 patients with additional temporal lobe involvement (oc- cipitotemporal), 10 patients with additional parietal lobe involvement (occipitoparietal), and 4 patients with additional involvement of both temporal and parietal lobes (oc- cipitotemporoparietal). These groups were then analyzed both separately and together as described below.

Preoperative Evaluation

All patients had suffered chronic medically intractable epilepsy for > 1 year and had undergone adequate trials of ≥ 2 antiepileptic drugs before they were referred for presurgical evaluation. All patients underwent continuous noninvasive scalp video-EEG monitoring to determine ictal and interictal focal epileptiform abnormalities. The details of the preoperative workup for epilepsy surgery candidates at our institution have been previously described in detail.

Demographic, Clinical, and Radiological Data

Demographic and clinical data used for this analysis included the following variables for each patient: 1) sex; 2) age at epilepsy manifestation; 3) age at operation; 4) epilepsy duration; 5) seizure type; 6) seizure frequency; 7) presence or absence of other medical history; 8) preoperative neurological status; and 9) preoperative visual fields. For the other medical history variable, we examined whether the patient had a history of trauma, hypoxia, perinatal insult, or intracranial hemorrhage. For preoperative neurological status, preexisting localizing neurological findings (such as hemiparesis) were documented. Preoperative visual fields were categorized on an ordinal scale (0 = no defect, 1 = minimal/scotoma, 2 = quadrantanopsia, 3 = incomplete hemianopsia, 4 = hemianopsia).

Magnetic resonance imaging was performed using 1.5- or 3-T systems (Gyrosan ACS-NT, Gyrosan NT-Intera, Gyrosan Intera, Gyrosan 3T Intera, and Gyrosan 3T Achieva; Philips Medical Systems) according to a standardized protocol that has been described previously. Briefly, the following sequences were acquired: 1) a sagittal 3D T1-weighted gradient echo sequence with a voxel size of 1 mm³; 2) axial FLAIR TSE and T2-weighted TSE sequences at a slice thickness of 5 mm; and 3) coronal inversion recovery (at a section thickness of 5 mm), FLAIR (at a section thickness of 3 mm), and T2-weighted TSE sequences (at a section thickness of 2 mm). If a lesion was detected, T1-weighted spin echo sequences before and after gadolinium/diethylenetriamine pentaacetic acid administration were acquired. Based on the neuroradiologist’s original MR imaging examination, radiological data were classified into the following categories: dysplasia, tumor, scar/cyst formation, vascular malformation, or other lesion. When FDG-PET (12 [23.1%] of 52 cases) and SPECT (9 [17.3%] of 52 cases) studies were performed, results were noted.

Electrophysiological Data

Preoperative, intraoperative, and postoperative electrophysiological data were reviewed in each case. Intercal EEG studies were separated into the following categories: 1) IEDs over the posterior cortex (EEG leads P3/P4, T5/T6, and O1/O2) ipsilateral to the resection side; 2) ipsilateral IEDs ipsilaterally over the posterior cortex but also over other brain regions; 3) IEDs ipsilateral to the resection side but anterior to the above-named contacts; 4) bilateral IEDs or IEDs contralateral to the resection side; and 5) no IEDs. Ictal EEG studies were categorized as follows: 1) ictal onset over the posterior cortex EEG leads ipsilateral to the resection side; 2) ictal onset ipsilateral to the resection side over the posterior cortex but also over other brain regions; 3) ictal onset ipsilateral to the resection side but anterior to the above-named contacts; 4) bilateral or contralateral ictal onset; or 5) no localizing, regionalizing, or lateralizing ictal onset pattern (for example, because of artifacts).

Preoperative invasive diagnostic monitoring was performed in 22 (42.3%) of 52 patients, and included various combinations of depth, strip, and grid electrodes. The type and location of grid and strip electrodes were noted. Whether intraoperative ECoG was performed was also noted.

Operative and Histopathological Data

Operative details recorded and analyzed in the database included the date, side, location, and type of operation. The
location of the operation (pure occipital, occipitotemporal, occipitoparietal, or occipitotemporoparietal) was scored based on the concordance between operation report descriptions and MR images. The patients with MR imaging–visible lesions received lesionectomies, and 12 patients (23%) received additional MSTs in adjacent epileptogenic eloquent areas. In the MR imaging–negative cases, topectomies were performed guided by intracranial EEG recordings. Two patients (3.8%) with recurrent seizures underwent repeat resections, and the location and type of the second operations were noted.

Resected specimens were examined histopathologically using methods previously described. Tumors were classified according to the revised WHO classification scheme. Different subtypes of cortical developmental malformations were grouped together as “dysplasia.” For purposes of overall evaluation and correlations, histopathological diagnoses were categorized into 1 of 5 categories: 1) dysplasia; 2) ganglioglioma; 3) other tumor; 4) vascular malformation; or 5) scar/gliosis formation.

Outcome Data

Postoperative visual field outcome was categorized on the same ordinal scale as preoperative field outcome (see above) to detect new visual field deficits in individual patients. Cases with unclear or incomplete pre- and postoperative visual field testing (19 patients [36.5%]) were excluded from this evaluation. For seizure and neurological outcome, follow-up information was obtained either from the last regular annual outpatient visit or from telephone interviews. Patients were assigned to 1 of 4 outcome classes according to the Engel classification scheme as follows: Class I, seizure free or auras only since surgery; Class II, rare seizures (< 2/year or only nondisabling nocturnal seizures); Class III, reduction of seizure frequency > 75%; and Class IV, unchanged (< 75% reduction of seizure frequency). For further analysis, Engel Classes I and II outcomes were grouped as satisfactory seizure control, whereas Classes III and IV were grouped as unsatisfactory seizure control.

Statistical Analysis

We analyzed multiple potential prognostic factors with respect to their prediction of good seizure outcome: 1) histopathology versus outcome; 2) surgical procedure versus outcome; 3) age of onset and epilepsy duration versus outcome; 4) location versus outcome; 5) seizure type versus outcome; and 6) EEG characteristics versus outcome. In addition, we studied potentially interesting interactions among nonoutcome variables in this patient group, such as histopathology versus seizure characteristics and seizure type versus age of onset. Each factor was analyzed by chi-square or Fisher exact tests both for Engel Class (I–IV) and satisfactory (Engel Class I or II) versus unsatisfactory (Engel Class III or IV) seizure outcome. Continuous variables were tested with the Student t-test. For nonparametric testing, the Mann–Whitney U-test was applied. For multifactorial analysis, a stepwise logistic regression model was applied. Backward stepwise logistic regression was performed with critical probability levels of 0.05 for inclusion and 0.1 for exclusion of factors from the model.

Results

Demographic and Clinical Findings

Demographic and clinical data are summarized in Table 1 for the entire patient population (52 patients) and in Table 2 for the subgroup with pure OLE (29 patients) and the extended OLE subgroup (23 patients). The overall population consisted of 25 males and 27 females. Mean age at operation was 23.6 years (range 4–58 years), and mean preoperative epilepsy duration was 13.6 years (range 1–42 years). The preoperative seizure history for the pure OLE group was 17 years, however, compared with only 9.5 years in the extended OLE group (p = 0.008, Student t-test). Eight (89%) of 9 patients with cortical malformations had their epilepsy manifestation before the age of 10 years, compared with only 40–50% in all other diagnoses. Thirteen patients (25%) had a medical history notable for trauma, hypoxia, intracranial hemorrhage, or previous intracranial surgery, whereas 75% had no obvious contributing medical history. Forty-two patients (80.8%) had a normal preoperative neurological status (visual fields were categorized separately), and 10 patients had preoperative neurological deficits (cognitive deficits in 6 patients and hemiparesis or hemisensory syndrome in 4 patients). These latter demographic factors were not significantly different between the two OLE subgroups.

Seizure types were quite variable (Tables 1 and 2), but the majority of patients (43 patients [82.8%]) had CPSs with or without other seizure types. Complex partial seizures occurred with similar frequencies in the pure and extended OLE subgroups. Thirty-one patients (59.6%) had generalized seizures. The most common seizure-type combination was CPS and occasional secondary generalized seizures (32.7%). Seizure frequencies varied over a wide range. In the 43 patients with CPSs, these seizures occurred at a median of 15 per month (range 1–600/month). In the 23 patients with SPs, these seizures occurred at a median of 25 per month (range 1–300/month). In the 31 patients with secondary generalized seizures, these seizures occurred at a median of 3.5/year.

Adequate preoperative visual field information was available for 41 patients (78.8%). Of this subset, 26 had intact preoperative visual fields, 3 had minimal deficit or scotoma, 5 had quadrantanopsia, 3 had incomplete hemianopsia, and 4 had complete hemianopsia. Between the 2 subgroups, the presence of a visual field defect was significantly more frequent in pure OLE (48.1%) compared with extended OLE (14.3%; p = 0.003, Fisher exact test).

Electrophysiological Findings

Adequate interictal EEG studies were available for review in 49 (94.2%) of 52 patients. Twenty (40.8%) of these 49 patients had epileptiform activity exclusively over the hemisphere on which the resection was performed; of these, only 3 had IEDs over posterior EEG leads only, and another 9 had IEDs at ipsilateral posterior plus other electrode contacts, whereas 8 patients had only nonposterior epileptiform activity. Sixteen patients had IEDs bilaterally or contralaterally to the resection side (32.7%), and in 13 patients (26.5%) no IEDs were recorded.

Ictal surface EEG studies were available in 41 patients (78.8% of the total group). Thirteen (31.7%) of these pa-
In nearly all cases (50 [96.2%] of 52), a structural lesion was visible on preoperative MR imaging. Preoperative MR imaging interpretations were as follows: 10 developmental tumors/gangliogliomas, 4 other tumors, 5 dysplasias, 7 vascular malformations, 16 cases of scar/cyst formation, 8 MR imaging—visible lesions not otherwise specified, and 2 cases in which no lesion was visible.

In 12 cases (23.1%), FDG-PET was performed. In 10 of these cases, FDG-PET revealed decreased glucose utilization ipsilateral to the lesion; in the other 2 cases it was normal or nonspecific. Single-photon emission CT was performed in 9 cases (17.3%). In 6 of these cases, SPECT revealed decreased perfusion ipsilateral to the lesion in each case; in 1 case, ictal SPECT was performed and demonstrated ictal hyperfusion ipsilateral to the lesion in each case; in 1 case, SPECT results were discordant with FDG-PET results. A clear histopathological diagnosis was obtained in all 52 cases (Table 3). Nine dysplasias were seen, with 5 focal corticinal dysplasias, 2 glioneuronal hamartomas, and 2 cases of tuberous sclerosis (Fig. 1). Nine gangliogliomas were observed (Fig. 2). Six other tumors were observed (5 astrocytomas and 1 ependymoma). Of the astrocytomas, 2 were WHO Grade I, 2 were Grade II, and 1 was Grade III. Thirteen vascular malformations were found: 6 AVMs, 5 cases of Sturge–Weber disease, and 2 cavernous malformations (Fig. 3). Fifteen cases of scar/gliosis formation were found,
with variable evidence of other histopathological features such as cortical and subcortical astrocytosis, hemosiderin deposition, microglial activation, and cyst formation (Fig. 4).

The 8 lesions revealed on MR imaging as not otherwise specified were found to comprise 3 vascular malformations, 2 dysplasias, 2 developmental tumors/gangliogliomas, and 1 other tumor (Grade I astrocytoma). The 2 cases of no MR imaging–visible lesion were found to comprise 1 case of scar/gliosis and 1 case of cortical dysplasia.

Thus the overall MR imaging sensitivity for detecting distinct histopathological lesions in this series was 96.2%. The MR imaging specificity was 78.6% for 15 tumors including gangliogliomas, 33.3% for any type of 9 dysplasias, 53.8% for 13 vascular malformations, and 86.7% for 15 cortical scars and gliosis.

Procedures and Complications

Forty-one patients underwent lesionectomies (extended, if possible), and 2 MR imaging–negative patients underwent topectomies. Five of these patients had undergone operations before, as described below. Twenty-five procedures were restricted to the occipital lobe, 8 were occipitotemporal resections, 7 were occipitoparietal procedures, and 3 resections were carried out in the occipitotemporoparietal region. Overall seizure outcome resulted in 30 pa-

TABLE 3
Histopathological diagnoses in 52 patients with OLE

<table>
<thead>
<tr>
<th>Histopathological Diagnosis</th>
<th>No. of Cases (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>dysplasia</td>
<td>9 (17.3)</td>
</tr>
<tr>
<td>focal cortical dysplasia</td>
<td>5</td>
</tr>
<tr>
<td>glioneuronal hamartoma</td>
<td>2</td>
</tr>
<tr>
<td>tuberous sclerosis</td>
<td>2</td>
</tr>
<tr>
<td>ganglioglioma</td>
<td>9 (17.3)</td>
</tr>
<tr>
<td>other tumor</td>
<td>6 (11.5)</td>
</tr>
<tr>
<td>Grade I astrocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Grade II astrocytoma</td>
<td>2</td>
</tr>
<tr>
<td>Grade III astrocytoma</td>
<td>1</td>
</tr>
<tr>
<td>ependymoma</td>
<td>1</td>
</tr>
<tr>
<td>vascular malformation</td>
<td>13 (25)</td>
</tr>
<tr>
<td>AVM</td>
<td>6</td>
</tr>
<tr>
<td>Sturge–Weber disease</td>
<td>5</td>
</tr>
<tr>
<td>cavernous malformation</td>
<td>2</td>
</tr>
<tr>
<td>scar/gliosis formation</td>
<td>15 (28.8)</td>
</tr>
</tbody>
</table>

Fig. 1. Example of a patient with an occipital focal cortical dysplasia. Preoperative sagittal (A, B, D, and E), axial (C), and coronal (F) FLAIR TSE images (at 3 T) of a 5-year-old girl with seizure onset at 2 years who experienced both SPSs and CPSs (~ 30/month). An extended occipital lesionectomy with preservation of the basal and mesial occipital cortex was performed using intraoperative ECoG. Histopathology revealed focal cortical dysplasia (Type IIb, Palmini and Lüders classification). Her preexisting contralateral superior quadrantanopsia was unchanged postoperatively, and she became seizure free (Engel Class I).
tients in Engel Class I (69.8%), and 4 patients in Engel Class II (9.3%), resulting in 79.1% satisfactory seizure control. Five 5 patients attained Engel Class III and 4 patients Engel Class IV, thus unsatisfactory seizure control was found in 20.9%.

Nine patients underwent lesionectomies together with additional MSTs of adjacent eloquent cortex. Four MSTs were restricted to the occipital area, 1 was occipitotemporal, 3 were occipitoparietal, and 1 was occipitotemporo-parietal. Seizures were fully abolished in 6 patients (66.7% Engel Class I), and seizures were reduced in frequency by >75% in 3 patients (33.3% Engel Class III). These results were not statistically significantly worse compared with the results of pure lesionectomies (\( p = 0.415 \), Fisher exact test).

Seizure control was less frequently achieved in the group of patients in whom invasive monitoring was required (22 patients, 42.3%): with noninvasive monitoring, 73.3% became seizure free, whereas the rate was 63.6% after invasive monitoring. In 7 patients with electrode implantations, however, results from intracranial monitoring resulted in the suggestion to perform lesionectomies and MSTs, and all but 1 patient became seizure free (85.7%, Engel Class I). Only 3 of these cases were occipital only, whereas the others involved adjacent temporal, parietal, or both lobes.

Five patients had undergone previous operations. One patient with a remote history of hypoxia and right frontal and occipital porencephaly with gliotic scar formation had undergone a right frontal lesionectomy 8 months prior without seizure relief; the occipital operation was the second operation and led to an Engel Class II outcome. Another patient with an occipital ganglioglioma had undergone an incomplete occipital lesionectomy without sufficient seizure control and remnants of the tumor were detectable on MR imaging; this patient underwent a second operation 6 months later that led to complete seizure control (Engel Class I). One patient had undergone an operation years before for an AVM, and another underwent an operation for a cavernoma. The former operations were only successful for extirpation of the malformations, but did not improve seizures, so that both patients underwent occipital reoperations to resect gliotic scars in the adjacent occipital or occipi-

![Example of a patient with an occipital ganglioglioma. Preoperative axial FLAIR (A), sagittal (B), axial (C), and coronal (D) inversion-recovery sequences (at 1.5 T) of a 14-year-old boy with seizure onset at 6 years of age who experienced 2.5 CPSs per month and 1 generalized seizure per year. An extended occipital lesionectomy and MSTs were performed using intraoperative ECoG. Histopathology revealed a WHO Grade I ganglioglioma. There was a new postoperative contralateral superior quadrantanopsia, and the patient was seizure free at 79 months (Engel Class I).](image-url)
totemporoparietal area. Both patients became seizure free (Engel Class I). Another patient had undergone an operation elsewhere for “gliosis,” which upon revision 3 years later was discovered to be a ganglioglioma, and an extended resection led to complete seizure relief. Thus, of 5 patients with a history of intracranial surgery, all 4 who underwent reoperations in the same area became seizure free. One patient with a distinct previous operation had an Engel Class II outcome.

Complications were noted in 2 patients (3.8%). In 1 case, a postoperative epidural hematoma was found that required drainage, without resulting in neurological deficit. In another case, postoperative intracranial hemorrhage led to hemiparesis and dysphasia. A third patient with a left occipitoparietal lesion had postoperative dyscalculia, which was transient and not considered a complication.

Follow-Up and Seizure Outcome

In the overall group of 52 patients, the mean follow-up period was 6.7 years (range 4 months–14.4 years; Table 4). A total of 36 patients (69.2%) were classified as seizure free (Engel Class I), and 4 patients (7.7%) had rare nondisabling seizures (Engel Class II). Thus, overall satisfactory seizure outcome was achieved in 40 patients (76.9%). Eight patients (15.4%) were categorized in Engel Class III, and 4 patients (7.7%) experienced no significant improvement (Engel Class IV); thus, unsatisfactory seizure outcome was observed in 12 patients (23.1%).

In the subgroup of 29 patients with pure OLE, mean follow-up duration was 86.1 months (range 4–170 months). Twenty-three patients (79.3%) had > 24 months of follow-up. A total of 21 patients (72.4%) were seizure free (Engel Class I) and 2 patients (6.9%) had rare nondisabling seizures (Engel Class II); thus, satisfactory seizure outcome was achieved in 79.3% of the pure OLE population. Three patients (10.3%) were categorized in Engel Class III, and 3 patients (10.3%) experienced no significant improvement (Engel Class IV); these patients were grouped together as unsatisfactory seizure outcome (20.7%).

**Fig. 3.** Example of a patient with an occipital vascular malformation. Preoperative CT scans (A–C) and T1-weighted MR images (D–F) with gadolinium enhancement (at 1.5 T) of a 57-year-old man with seizure onset at 16 years of age who experienced 30 CPSs per month. Invasive monitoring with grid and subdural strip electrodes was performed, followed by an extended occipitobasal lesionectomy. Histopathology results revealed an AVM with calcification. There was a new postoperative contralateral hemianopsia, and the patient was not seizure free at 33 months (Engel Class IV) despite complete resection as revealed by repeated MR imaging.
Factors analyzed as predictors of seizure outcome included the location of lesion, sex, age at operation, age at epilepsy onset, duration of epilepsy, seizure characteristics, and histopathological diagnosis. Neither preoperative seizure frequency nor presence of specific seizure types (SPSs, CPSs, or generalized seizures) predicted postoperative seizure outcome. There was no statistically significant difference in seizure outcome by location of lesion (occipital, occipitoparietal, occipitotemporal, occipitotemporoparietal; Table 4) analyzed according to either Engel class (p = 0.368, Fisher exact test) or to satisfactory versus unsatisfactory seizure outcome (p = 0.478, Fisher exact test).

Age at operation did not have a statistically significant effect on seizure outcome (p = 0.181; Fisher exact test). Similarly, age at epilepsy manifestation was not a statistically significant predictor of seizure outcome according to chi-square analysis and nonparametric analysis (p = 0.168 for satisfactory seizure control; Mann–Whitney U-test), but was later significant in the logistic regression analysis (see below). Duration of epilepsy > 20 years was associated

<table>
<thead>
<tr>
<th>Group</th>
<th>No. of Patients</th>
<th>Mean Follow-Up Mos (range)</th>
<th>Engel Class* (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>I</td>
</tr>
<tr>
<td>all patients</td>
<td>52</td>
<td>80.3 (4–173)</td>
<td>36 (69.2)</td>
</tr>
<tr>
<td>occipital only</td>
<td>29</td>
<td>86.1 (4–170)</td>
<td>21 (72.4)</td>
</tr>
<tr>
<td>occipitoparietal</td>
<td>10</td>
<td>89.9 (12–173)</td>
<td>6 (60)</td>
</tr>
<tr>
<td>occipitotemporal</td>
<td>9</td>
<td>60.1 (10–102)</td>
<td>5 (55.6)</td>
</tr>
<tr>
<td>occipitotemporoparietal</td>
<td>4</td>
<td>60.1 (6–123)</td>
<td>4 (100)</td>
</tr>
</tbody>
</table>

* Number of patients in each class.

TABLE 4
Summary of follow-up and seizure outcome

Fig. 4. Example of a patient with an occipital glial scar. Preoperative axial T2-weighted gradient echo (A–C) and coronal FLAIR TSE images (D–G) of a 36-year-old man with seizure onset at 6 years of age who experienced 6 SPSs per month, 2.5 CPSs per month, and 2 generalized seizures per year. Invasive monitoring with depth electrodes and occipital interhemispheric strip electrodes was performed, followed by an extended occipital lesionectomy. Histopathology revealed glial scar/gliosis formation. The preoperative small scotoma enlarged into a postoperative contralateral upper quadrant-anopsia, and seizure outcome was Engel Class III (> 75% reduction in seizure frequency) at 80 months.
Surgery for occipital lobe epilepsy

TABLE 5
Seizure outcome according to epilepsy duration in 52 patients with OLE

<table>
<thead>
<tr>
<th>Epilepsy Duration (yrs)</th>
<th>No. of Patients</th>
<th>Satisfactory (Engel Class I or II)</th>
<th>Unsatisfactory (Engel Class III or IV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;5</td>
<td>16</td>
<td>13 (81.3)</td>
<td>3 (18.8)</td>
</tr>
<tr>
<td>5–10</td>
<td>10</td>
<td>9 (90.0)</td>
<td>1 (10.0)</td>
</tr>
<tr>
<td>&gt;10–20</td>
<td>13</td>
<td>12 (92.3)</td>
<td>1 (7.7)</td>
</tr>
<tr>
<td>&gt;20</td>
<td>13</td>
<td>6 (46.2)</td>
<td>7 (53.8)</td>
</tr>
</tbody>
</table>

* Number of patients in each category.

TABLE 6
Seizure outcome according to histopathological group in 52 patients with OLE

<table>
<thead>
<tr>
<th>Histopathological Group</th>
<th>No. of Patients</th>
<th>Engel Class* (%)</th>
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</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>I</td>
</tr>
<tr>
<td>all patients</td>
<td>52</td>
<td>36 (69.2)</td>
</tr>
<tr>
<td>dysplasia</td>
<td>9</td>
<td>6 (66.7)</td>
</tr>
<tr>
<td>ganglioglioma</td>
<td>9</td>
<td>8 (88.9)</td>
</tr>
<tr>
<td>other tumor</td>
<td>6</td>
<td>3 (50)</td>
</tr>
<tr>
<td>vascular malformation</td>
<td>10</td>
<td>10 (76.9)</td>
</tr>
<tr>
<td>scar/gliosis formation</td>
<td>15</td>
<td>9 (60.0)</td>
</tr>
</tbody>
</table>

* Number of patients in each class.

with poorer outcome (46.2% satisfactory seizure outcome) compared with < 20 years (p = 0.034, Fisher exact test; Table 5). These findings were confirmed by a nonparametric analysis comparing age and seizure outcome, in which a younger age at surgery correlated with better seizure control (p = 0.003 for Engel Class I outcome and p = 0.002 for satisfactory seizure control [Engel Class I or II]; Mann–Whitney U-test).

Seizure characteristics (seizure frequency or seizure types) were not associated with outcome. A positive past medical history for conditions such as trauma, intracranial hemorrhage, or hypoxia was not associated with altered outcome (p = 0.811, Fisher exact test). In addition, the presence or absence and type of intracranial invasive monitoring failed to predict outcome.

We next considered whether histopathology results may have been associated with outcome. Table 6 demonstrates seizure outcome (Engel Class) according to histopathological group. The patients with the best seizure outcome were found in the ganglioglioma group (89% Engel Class I, 11% Engel Class II), and the poorest outcome was seen in patients in the “other tumor” group (50% Engel Class I, 0% Engel Class II). This difference was not statistically significant (p = 0.565, Fisher exact test).

Multifactorial Analysis of Prognostic Factors

To formally analyze multiple clinical and surgical factors simultaneously in the prediction of outcome, we performed a multifactorial univariate logistic regression analysis. The dependent variable was satisfactory versus unsatisfactory seizure outcome. Factors tested in the model were sex, seizure outcome (Engel Class I or II), age at epilepsy manifestation (p = 0.187). Instead, female sex was found to be correlated with being seizure free (p = 0.003).

Visual Field Outcome

In 33 cases, both adequate preoperative and postoperative visual field data were available for comparison. Of these 33 cases, 21 patients (63.6%) started with intact visual fields and 12 patients (36.4%) had preoperative visual field deficits. Of those 21 patients with intact preoperative fields, 10 patients (47.6%) still had intact visual fields postoperatively; therefore, 11 patients (52.4%) had new deficits. These deficits consisted of a new scotoma/subquadrant deficit in 2 cases, new quadrantanopsia in 3 cases, new incomplete hemianopsia in 3 cases, and new hemianopsia in 3 cases. Of the 12 patients with preoperative visual field deficits, 9 patients (75%) had no worsening of visual field deficit and 3 patients (25%) experienced worsened deficits postoperatively. In summary, new or aggravated visual field deficits were seen in 14 (42.4%) of the 33 cases in which all the information was available.

Discussion

Diagnosis of OLE

Due to the relatively small number of patients who have undergone operations so far for OLE, experience is still limited, for example, compared with TLE. Occipital lobe epilepsy appears to account for ~ 2–13% of extratemporal epilepsies.\(^3\),\(^6\),\(^23\),\(^40\) The literature indicates that there is a strong selection bias in extratemporal epilepsy patient series, depending on referral, inclusion criteria, and consideration of potential surgical candidates.

We found an epilepsy duration of 17 years in the subgroup with pure OLE, whereas the epilepsy duration was only 9.5 years in the group with extended OLE (occipitotemporal, occipitoparietal, or occipitotemporoparietal epilepsies). This difference in seizure history was statistically significant. In this context, it is interesting to note that a longer duration of epilepsy was 1 of 2 factors predictive of poorer outcome after surgery. Long epilepsy duration prior to surgical therapy may be related to several challenges in diagnosing and treating OLE. Many patients developed their epilepsies before the introduction of modern MR imaging and before the reinstitution of epilepsy surgery in Germany in the 1980s. In contrast, more extended forms of OLE resulted in shorter individual seizure history. One can
assume that although these latter epilepsies are more widespread in nature, they may be considered as evolving from less eloquent areas. The core of pure OLE is the mesial occipital area and the occipital pole, both highly eloquent areas, whereas extended forms of OLE mostly involve more lateral areas in which surgical resections can be performed with less risk of persistent neurological deficits. This explanation is supported by the observation that patients with pure OLE had significantly more preoperative visual field deficits compared with the patients with extended OLE (48 vs 14%, respectively; \( p = 0.003 \)). This finding is consistent with the study of Blume and colleagues who found visual field deficits in 42% of patients with mesial OLE and none in lateral OLE, whereas other clinical symptoms did not allow them to distinguish between mesial and lateral OLE. Interestingly, semiological features characteristic of TLE were found in 78% of the patients in the latter study. This is consistent with our finding of 83% CPSs in the pure OLE and the extended OLE subgroups, suggesting that ictal features do not allow differentiation between mesial or lateral OLE, nor between OLE and TLE.

Electrophysiology and OLE

Because neither semiological nor EEG findings provide reliably specific findings in OLE, diagnosis and selection of patients for surgery is difficult. In the present study, all patients had medically refractory epilepsy; however, MR imaging findings played a major role in nearly all patients in suggesting occipital seizure localization. This lesion-based hypothesis was then furthered by analysis of clinical and EEG findings. Thus, our approach was predominantly lesion-directed, and the vast majority of patients had lesions documented in the occipital lobes or additionally in adjacent areas of parietal and/or temporal lobes.

Elementary visual hallucinations have been consistently found as part of OLE semiology; however, TLEs can exhibit comparable features. Typical visual auras were found in 61–68% of patients in recent studies of OLE or posterior epilepsies, but they were not reliably lateralizing. Salanova and colleagues described visual auras in 73% of 42 patients who underwent operations for OLE over a period of > 60 years. Williamson et al. found only initial ictal symptoms to provide information on the presumed seizure focus in 88% of patients with OLE, whereas symptoms in the later course of seizures were nonspecific: 44% experienced seizures with typical temporal semiology, and the same amount had multiple types of seizures. Böesebeck and associates hypothesized that with precise characterization of auras it should be possible to lateralize the seizure focus. This characterization had a significant impact on outcome prediction: lateralizing auras resulted in complete seizure relief in 69%, whereas the rates were 28% only without lateralizing auras, and false lateralization resulted in a 0% rate of seizure control. These findings were mostly explained by the arbitrary borders of the occipital lobe and strong connections with temporal and parietal lobes, which enable fast propagation of seizure activity. Thus, if the initial symptoms are not determined, misdiagnosis of TLE may result.

Interictal and ictal surface EEG recordings may contribute to the generation of an adequate focus hypothesis in some cases insofar as they frequently indicate the hemispheric (40.8% of interictal studies and 31.7% of ictal studies) or even brain region (24.5% of interictal EEG studies, 24.4% of ictal EEG studies) that is eventually shown to contain the ictal onset area. To clarify potentially misleading clinical and electrophysiological results, evaluation using intracranial electrodes may be necessary. Invasive monitoring with implanted electrodes was performed in 42% of patients in our study to define the epileptogenic area and to reliably differentiate seizure spread from occipital origin versus nonoccipital seizure origin.

Imaging of OLE

Only 6 modern studies of OLE included only patients who had undergone MR imaging prior to surgery. As in these modern studies, the vast majority of patients in our study (50 [96%] of 52) showed MR-detectable lesions. Thus, we took a lesion-directed approach in this study, but identification of a lesion alone is not sufficient to offer epilepsy surgery to the patient. Magnetic resonance imaging proved to be very sensitive, but MR imaging specificity was lower: 87% for scars and cysts, 79% for tumors, and surprisingly only 54% for vascular malformations. The least specificity was attained in cortical malformations (33%). Clinically a high sensitivity is most important, however, and clearly in this group of patients MR imaging proved to be the major adjunct to the diagnostic armamentarium. The presence of a lesion on MR imaging is generally accepted to portend a better prognosis for becoming seizure free. The introduction of modern MR neuroimaging has facilitated the identification of occipital lesions. It should be noted that 2 lesions were undetected using MR imaging in our sample, but nevertheless MR sensitivity was 96%. The clinical impact of the lower specificities may be of less importance, because initial treatment planning is not primarily dependent on histopathological diagnosis.

In nonlesional cases or cases with ambiguous MR images, FDG-PET and ictal SPECT have been especially helpful in defining the epileptogenic zone. In 23% of cases, we made use of FDG-PET to support the clinical and/or imaging hypothesis. In 10 of 12 cases, the FDG-PET data supported the MR imaging and clinical findings, and in 2 cases the results were discordant. Ictal SPECT was performed in 3 patients in our series, and in each case hyperfusion ipsilateral to the lesion was observed. When ictal injection was not possible, however, ictal SPECT was performed in 9 patients, and only 67% resulted in lateralizing findings. Kim et al. found ictal SPECT to be sufficiently lateralizing in 76%, but the correct localization was possible in only 29%. In their study, FDG-PET was superior in localizing epileptogenic zones, showing 93% correct lateralization and 69% correct localization. Thus, FDG-PET and SPECT can be regarded as diagnostic adjuncts, but they must be interpreted carefully and further study is necessary to determine the specific role of each modality.

In 1 case in our series, FDG-PET and SPECT revealed contradictory results.

Surgery and Complications

The majority of patients (43 [83%] of 52) underwent lesionections tailored to include the lesion and the surrounding presumed epileptogenic zones as derived from noninvasive or invasive EEG monitoring. In a few cases,
Surgery for occipital lobe epilepsy

results from FDG-PET and SPECT were also considered for resection planning. In some cases, the resection was limited when preservation of visual fields was intended, but restricting the extent of resection may worsen outcome. Epileptogenic zones are known to be quite extensive in occipital epilepsies, often involving posterior temporal and parietal regions, so that it is often difficult to achieve complete resection with an acceptable risk for permanent neurological deficits. Furthermore, it is possible that the epileptogenic focus may be distant from the morphological lesion, due to either fast propagation and/or intense coupling of the structures involved. Proximity to eloquent areas is the main reason that most patients underwent extended lesionectomies, and only 9 patients (17%) underwent lobectomies. In 9 patients, the planned lesionectomies were not able to encompass the entire presumed epileptogenic zone, as derived from invasive EEG monitoring; these patients underwent additional MST. Six of these 9 patients became completely seizure free, and another 3 had significant improvement in seizure frequency. Other studies have previously shown that MST may be a helpful surgical adjunct in eloquent areas, because seizure outcome can be superior when combined with a resective approach.

Five patients had a history of previous intracranial surgery for different diseases, which resulted mostly in incomplete resection. All 4 patients who underwent reoperations in the same area as before became seizure free after occipital epilepsy surgery. Thus, reoperation should be considered in those cases with residual MR imaging–visible lesions, and resection of a glial scar alone may be quite beneficial in individual cases.

A concern for occipital lobe surgery is aggravation of existing or creation of new visual field defects. Preoperatively, visual field deficits were present in 36% of patients, and 42% had new or aggravated visual field defects after surgery. These results compare well with those in published findings. In addition, several authors have found that visual field deficits are rarer in lateral OLE compared with mesial OLE, which is understandable based on the location of the calcarine cortex and is also supported by our data on extended OLE. Clearly, the informed consent process must involve a frank discussion of the likelihood of visual field loss following OLE surgery.

Histopathological Findings

Histopathological diagnoses were obtained in all 52 cases. The most common finding was glial scars/gliosis (29%), followed by vascular malformations (25%), gangliogliomas (17%), dysplasias (17%), and other tumors (12%). The spectrum of these occipital lesions is different from other areas of the brain and has not previously been described in detail. Other investigators have found a predominance of malformations of cortical development or tumors. In our series, 2 of the astrocytomas were WHO Grade I, 2 were Grade II, and 1 was Grade III, which corresponds to larger patient cohorts who underwent epilepsy surgery. The most notable result was that 15 cases of glial scar/gliosis were found, with variable evidence of other pathological features such as cortical and subcortical astrogliosis, hemosiderin deposition, microglial activation, and cyst formation. Some of these cases were related to clear evidence of prior trauma. The predominance of this diagnosis emphasizes the importance of glial scars not only in the classic location of mesial temporal sclerosis but also in contributing to occipital epileptogenesis. Gliotic lesions can be associated with continuous epileptiform discharges.

Outcome Predictors

Of 52 patients with a mean follow-up of 6.7 years, 36 (69.2%) were classified as seizure free (Engel Class I), and 4 (7.7%) had rare nondisabling seizures (Engel Class II). Thus, overall satisfactory seizure outcome was achieved in 40 patients (76.9%). Eight patients (15.4%) were categorized in Engel Class III, and 4 patients (7.7%) experienced no significant improvement (Engel Class IV); thus, unsatisfactory seizure outcome was observed in 12 patients (23.1%). Our outcome results are better compared with those in older patient series, although direct comparison is difficult due to differences in patient selection and use of different classification schemes. As illustrated in Table 7, however, seizure-free outcomes were attained in 33–50% of patients in most of the larger studies, which were published in the 1990s. The results of the more recent studies on surgical treatment of OLE described seizure control rates similar to ours. It appears that the introduction of modern MR imaging, video-EEG monitoring, and individualized use of invasive monitoring improved the seizure-free outcome of epilepsy surgery in the occipital lobe by ~ 20%. Similar improvements over time have been noted for TLE surgery.

In the subgroup of 29 patients with pure OLE, a total of 21 patients (72.4%) were seizure free (Engel Class I) and 2 patients (6.9%) had rare nondisabling seizures (Class II); thus, satisfactory seizure outcome was achieved in 79.3% of the pure OLE population. With more extended occipital surgery in 23 patients, similar results were obtained (65.2% Engel Class I and 8.7% Engel Class II), resulting in 74% satisfactory seizure relief in this subgroup. There was no significant difference in seizure outcome according to the location of the lesion (occipital, occipitoparietal, occipitotemporal, or occipitotemporoparietal). Furthermore, neither clinical, semiological, or EEG factors correlated with outcome. With respect to histopathology, the best seizure outcome was noted in the ganglioglioma group (89% Engel Class I and 11% Engel Class II), and the poorest outcome was noted in the “other tumor” group (50% Engel Class I and 0% Engel Class II). Similar findings have been described for TLE, and these results support the idea of the special behavior of the developmental tumors and their role in long-term epilepsy.

We found that young age at epilepsy manifestation was a significant predictor of satisfactory seizure outcome in some analyses. Duration of epilepsy was associated with poorer outcome in our study, whereas age at operation was not predictive. Other studies have reported that duration of epilepsy may affect outcome, although this is not consistently reported to be significant. In a recent study of 44 patients with posterior cortex epilepsies, Dalmagro et al. found that a favorable outcome was associated with shorter epilepsy duration. This finding, together with our data, suggests that an earlier operation is preferable. The epilepsy duration of 17 years in our pure OLE subgroup should
be regarded as a challenge to accelerate patient selection and presurgical evaluation.

Conclusions

Occipital lobe epilepsy is a rare but significant cause of extratemporal epilepsy. Satisfactory results (Engel Class I or II) were obtained in 77% of patients in our series with a mean follow-up of 6.7 years. Our study supports the improvement of seizure outcomes in the modern era with high-resolution MR imaging and careful video-EEG monitoring and other appropriate tests. Postoperative visual field deficits occur in a significant proportion of patients. Long epilepsy duration is a negative prognostic factor, suggesting that diagnosis and treatment of patients with lesional OLE should be performed early for better outcome.

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