Long-term visual outcome after microsurgical removal of occipital lobe cavernomas

Clinical article

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Object. Cavernomas in the occipital lobe are relatively rare. Because of the proximity to the visual cortex and incoming subcortical tracts, microsurgical removal of occipital cavernomas may be associated with a risk of visual field defects. The goal of the study was to analyze long-term outcome after operative treatment of occipital cavernomas with special emphasis on visual outcome.

Methods. Of the 390 consecutive patients with cavernomas who were treated at Helsinki University Central Hospital between 1980 and 2011, 19 (5%) had occipital cavernomas. Sixteen patients (4%) were surgically treated and are included in this study. The median age was 39 years (range 3–59 years). Seven patients (56%) suffered from hemorrhage preoperatively, 5 (31%) presented with visual field deficits, 11 (69%) suffered from seizures, and 4 (25%) had multiple cavernomas. Surgery was indicated for progressive neurological deterioration. The median follow-up after surgery was 5.25 years (range 0.5–14 years).

Results. All patients underwent thorough neuroophthalmological assessment to determine visual outcome after surgery. Visual fields were classified as normal, mild homonymous visual field loss (not disturbing the patient, driving allowed), moderate homonymous visual field loss (disturbing the patient, driving prohibited), and severe visual field loss (total homonymous hemianopia or total homonymous quadrantanopia). At the last follow-up, 4 patients (25%) had normal visual fields, 6 (38%) had a mild visual field deficit, 1 (6%) complained of moderate visual field impairment, and 5 (31%) had severe homonymous visual field loss. Cavernomas seated deeper than 2 cm from the pial surface carried a 4.4-fold risk of postoperative visual field deficit relative to superficial ones (p = 0.034). Six (55%) of the 11 patients presenting with seizures were seizure-free postoperatively. Eleven (69%) of 16 patients had no disability during the long-term follow-up.

Conclusions. Surgical removal of occipital cavernomas may carry a significant risk of postoperative visual field deficit, and the risk is even higher for deeper lesions. Seizure outcome after removal of these cavernomas appeared to be worse than that after removal in other supratentorial locations. This should be taken into account during preoperative planning.

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Key Words • cavernoma • occipital lobe • microsurgery • visual deficit • vascular disorders

Cavernomas are usually detected between the 2nd and 5th decades of life.6,15,21,26 The most frequent manifestations of the disease are seizures, focal neurological deficits, and hemorrhage.1,2,18,23 The risk of symptomatic bleeding depends on the location of the cavernoma, generally increasing in deeper lesions of the brain; these risks amount to 0.1%–5% per patient-year (rebleeding risk 5%–60% per patient-year).7,10,11,16,19,21,24,26 Cavernomas located in the occipital region are relatively rare. Microsurgical removal of cavernomas in this region may be associated with a considerable risk of visual field deficits. However, when a patient presents with progressive visual field loss or seizures, surgical removal may be warranted. Literature on the natural history of occipital cavernomas and outcome after surgical treatment is scarce. The purpose of our study was 3-fold: to assess the frequency and pattern of visual field deficits in patients with occipital cavernomas pre- and postoperatively, to assess the role of visual disorders and seizures causing disability after surgery, and to assess the recovery potential of visual disorders in long-term follow-up.

Abbreviation used in this paper: DRE = drug-resistant epilepsy.
Methods

Patients and Symptoms

Of the 390 consecutive patients with cavernomas of the brain and spine treated at Helsinki University Central Hospital between January 1980 and January 2011, 19 (5%) had occipital lobe cavernomas. Sixteen patients (4%) were surgically treated and are included in this study. Patient files and images were analyzed retrospectively. The study protocol was approved by the local ethics committee of Helsinki University Central Hospital.

The patient population comprised 7 male (44%) and 9 female (56%) patients. Their median age was 39 years (range 3–59 years); 2 patients (12%) were children (< 18 years). In most cases, patients underwent initial examination in the neurological department of the referring hospitals and were then referred to our neurosurgical center for further evaluation and treatment. All but 1 patient had neurological symptoms on admission. Five patients (31%) presented with visual field deficits, and all 5 had a history of seizures. Eleven (69%) of the 16 patients suffered from seizures. Six (55%) of these 11 patients had only 1 seizure preoperatively, whereas 5 (45%) suffered from chronic epilepsy before surgery. Six patients with seizures (55%) had a history of hemorrhage. Altogether, 7 patients (56%) experienced hemorrhage from the cavernoma before surgery: 5 (71%) had a single hemorrhage, 1 had 2 hemorrhages, and 1 had 3 hemorrhages. Of these patients, 2 underwent surgery within 1 week of the hemorrhage because of progressive neurological deterioration, and 1 underwent surgery several months after the hemorrhage. None of these patients had a life-threatening hemorrhage with significant mass effect. Four of the 16 patients had headaches as the main symptom. The headache was associated with a CT-confirmed cavernoma hemorrhage in only 1 patient. Headaches were typical for cavernomas affecting the ventricular wall and in 2 of the 3 patients with a cavernoma of the trigone.

Imaging Procedures

Prior to admission to our department, each patient underwent MRI, except for the 1 patient admitted at a time when only CT was available. In patients with acute sudden headache or first seizure, a CT scanning study was performed soon after symptom onset to exclude hemorrhage (Fig. 1). If a cavernoma was suspected, an MRI study was performed within 1 month after ictus. The radiological features of the lesions were classified according to Zabramski’s classification system by the first author (J.K.) (Table 1). Type II cavernomas were the most common, occurring in 47% of patients. In 4 of 16 patients, the cavernoma had a pial surface extension. In the remaining 12 patients, the lesion did not reach the cortex; in 6 patients (38%), the shortest distance from the cortical surface to the lesion was less than 2 cm, and in the remaining 6 patients (38%), the lesion was located deeper than 2 cm from the pial surface, partially occupying the white matter. In 3 patients, the lesion was located in the trigone and was partially embedded in the ventricle wall. The median size of the cavernoma core in the series was 10 mm (range 5–30 mm). Four patients (25%) had multiple cavernomas. In all of these patients, an occipital cavernoma was suggested to cause symptoms and was radiologically the most active (Zabramski Type I or II), indicating active treatment. One patient had an unruptured cerebral aneurysm (middle cerebral artery) that was treated microsurgically before cavernoma removal.

All patients underwent CT imaging on the 1st postoperative day. Follow-up MRI was performed if the patient exhibited progressive worsening of symptoms.

Surgical Treatment

Surgery was indicated due to progressive neurological deterioration in all but 1 patient who was asymptomatic preoperatively. Microneurosurgical removal was performed via standard craniotomy placed according to the location of the cavernoma in the occipital region. Frameless neuronavigation was used in 10 patients, allowing optimization of the craniotomy to provide the shortest and safest route to the lesion. When the cavernoma did
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| TABLE 1: Classification of cerebral cavernous malformations* |
|-----------------|-----------------|-----------------|
| Lesion Type | MRI Signal Characteristics | Pathological Characteristics |
| I | T1, hyperintense core; T2, hyper-intense core | subacute hemorrhage |
| II | T1, reticulated mixed signal core; T2, reticulated mixed signal core w/ surrounding hypointense rim | lesions w/ thrombosis of varying ages |
| III | T1, iso- or hypointense; T2, hypointense lesion w/ hypointense rim magnifying the lesion size | chronic hemorrhage w/ hemosiderin staining w/in & around the lesion |
| IV | T1 & T2, not seen; GRE, punctate hypointense lesion | tiny cavernoma or telangiectasia |

* Adapted from Zabramski et al. Abbreviation: GRE = T2*-weighted gradient echo sequence.

not reach the cortical surface, a transsulcal approach was used (Fig. 3). During sulcal preparation a discoloration of the brain parenchyma was seen as a reliable sign of an underlying lesion. In patients with recent extracerebral bleeding, a hematoma was initially aspirated and further dissection of the lesion was carried out. Because of the vicinity of the visual cortex and tracts, no marked resection of the perifocal hemosiderosis was performed. When identified, a neighboring developmental venous malformation remained untouched during surgery to avoid venous infarction. After surgery, patients were discharged when stable (median 5 days [range 3–6 days]), or they were transferred to the referring hospital for further rehabilitation.

Neuroophthalmological Follow-Up

The first clinical follow-up was performed at a median of 2 months (range 1.5–3 months) after the procedure. When needed, further follow-up was arranged, mostly by the neurologists at the referring hospitals. All but 4 patients underwent neuroophthalmological assessment, including complete ophthalmological examination in June 2011 to assess visual outcome after surgery. In these 4 patients, a long-term ophthalmological examination, including visual field test, was conducted at an earlier stage since patients moved to another city or were not available for reexamination. The visual fields of the patients were examined using static perimetry with the aid of the Octopus 101 perimeter (Haag-Streit, Inc.), and kinetic perimetry was performed with the aid of the Goldmann perimeter (Haag-Streit, Inc.) by a skilled perimetrist in a standardized fashion. Automated static perimetry was preferred, but kinetic perimetry was chosen if the patient could not cooperate with automated static procedure or if it proved unreliable. Visual field test results were compared with those obtained immediately after surgery or preoperatively, if available. For better systematization of the results, patients were assigned to 4 groups based on their visual field findings as follows: Group 0, normal visual field; Group 1, mild homonymous visual field loss (not disturbing the patient, driving allowed); Group 2, moderate homonymous visual field loss (disturbing the patient, driving prohibited); and Group 3, severe visual field loss (total homonymous hemianopia or total homonymous quadrantanopia). We examined whether fixation was lost and whether visual field loss was relative or absolute. The median follow-up after surgery was 5.25 years (range 0.5–14 years).

Statistical Analysis

Statistical analysis was carried out using SPSS 17.0 software (SPSS Inc.). Clinical characteristics were presented as frequencies and percentages for categorical variables and as mean ± SD for continuous variables. Nonparametric data were analyzed using the Pearson chi-square test, and continuous variables were compared using the Student t-test. The level of significance was set at p < 0.05. All tests were 2-sided.

Results

Operation

Seizures, visual field deficits, and progressive headaches with or without hemorrhage were common indications for surgery in our patients. One patient was asymptomatic preoperatively, and the cavernoma was removed to eliminate potential risks of epilepsy or hemorrhage since the patient was relatively young (24 years) and the 10-mm lesion was on the cortex, allowing for a potentially safe operation. Surgery was performed using microneurosurgical techniques. All patients were informed about the risks of visual field deficits after surgery. Twelve patients (75%) underwent a single operation. In 11 of them, the cavernoma was successfully removed, but in 1 patient the lesion could not be found despite using frameless neuronavigation; this patient refused further surgeries, and her neurological state remained the same with no progression of visual field deficits. Four patients (25%) underwent a reoperation; indications for surgery were a cavernoma residual in 2 patients and failure to initially locate the cavernoma in the other 2. The surgical approach was tailored according to the location of the cavernoma in the brain, attempting to minimize

Fig. 3. Tiny occipital cavernoma (arrow) causing repetitive seizures within the same day, indicating surgical removal. Perimetry tests after several weeks postoperatively showed new visual field deficits. Left: Axial T2-weighted MRI study. Right: Axial T2*-weighted gradient echo MRI study.
damage to the visual cortex or incoming optic tracts. When a lesion was situated near the midline and the interhemispheric approach was used, special attention was paid to avoiding damage to the bridging veins.

The most common complaint immediately after surgery was a visual field deficit, reported by 10 patients (62%). On discharge from the hospital, 7 of these patients (70%) complained of partial field deficits that did not disturb walking, and 3 (30%) complained of complete homonymous hemianopia limiting their reading and even walking significantly. One patient with a cavernoma in the parietooccipital border developed contralateral hemiparesis, but his visual field remained intact.

**Visual Outcome**

At the last follow-up, 4 patients (25%) had a normal visual field (Group 0); 6 (38%) had mild, nondisturbing visual field deficits (Group 1); 1 (6%) had moderate visual field impairment (Group 2); and 5 (31%) had significant disturbing and limiting visual field deficits (Group 3) (Table 2). Two of the 5 patients with visual field deficits preoperatively experienced complete improvement in their visual problems. A detailed neuroophthalmological examination in 12 patients confirmed absolute total homonymous hemianopia in 2 patients (17%), absolute homonymous visual field loss at 3–4 o’clock in 3 patients (25%), and relative quadrantanopia at 1.5–3 o’clock in 7 patients (58%) (Fig. 4).

In 4 (33%) of the 12 patients who experienced visual field problems, some improvement occurred after surgery, while 8 patients (67%) remained the same. Among these were 1 patient (Case 9) who had no visual field deficits but had postoperative cerebral visual worsening, including impaired visual memory and recognition of depth and motion perception, and another (Case 8) who had visual field deficits and visual nonepileptic hallucinations consisting of brief flashes of light. All but 1 patient had corrected visual acuities of 20/25 or better in both eyes (the patient in Case 1 had anisometropic amblyopia in 1 eye). No abnormal findings on dilated slit lamp and fundus examination that would affect perimetric testing were observed.

The patient in Case 8 had an ischemic lesion in the occipital lobe after surgery, followed by difficult neuro-psychological symptoms manifesting with disturbances of visual spatial defects, visual memory, and neglect. The patient in Case 9 suffered from DRE and began to have troublesome visual hallucinations 6 years after the operation. Brain PET imaging showed hypometabolic areas in the left temporal lobe. However, on video electroen-
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cerebrospinal fluid, no epileptic discharge or asymmetry was observed in the left tempororooccipital region when visual hallucinations occurred.

The only significant predictive factor for better visual outcome was the distance of the lesion from the cortical surface; 3 (75%) of 4 patients with cortical cavernomas had normal visual fields at the last follow-up, whereas of the 6 patients with lesions situated at a depth of more than 2 cm, a normal visual field was registered in only 1 (17%) (p = 0.034) (Table 3). No significant difference in visual outcome was detected between patients who underwent surgery via a transcortical or transsulcal approach. Furthermore, a reoperation was not associated with an additional risk of visual field deficits. Sex, history of bleeding, side and size of the cavernoma, and duration of follow-up were not correlated with better visual outcome.

Seizure Outcome

After a follow-up of a median of 5.25 years (range 0.5–14 years), 6 (55%) of the 11 patients who presented with seizures improved completely after surgery, while 5 (45%) continued to have seizures. Furthermore, 1 patient (Case 15), who was asymptomatic before surgery, developed seizures postoperatively. Among the 6 seizure-free patients, 4 (36%) continued to use antiepileptic drugs due to a fear of recurrent seizures. Of the 6 patients presenting with a single seizure, only 3 (50%) were seizure-free, while the remaining 3 suffered from recurrent seizures after surgery, requiring additional antiepileptic drugs. The patient in Case 5 was admitted to a referral hospital 6 months later due to status epilepticus with consequent MRI-confirmed damage in the occipital region. This patient had a history of alcohol abuse and used antiepileptic drugs irregularly. At the last follow-up, he continued to suffer from chronic epilepsy and was severely disabled.

In general, sex, size or side of the cavernoma, radiological appearance, distance from the cortex to the lesion, history of bleeding, use of the transsulcal or transcortical approach, and duration of follow-up were not correlated with the long-term outcome of seizures.

General Outcome

Of the 16 patients, 11 (69%) had no disability at the last follow-up, while 4 (25%) experienced moderate disability and 1 (6%) was severely disabled due to DRE with consequent mental retardation. No patient died. The reasons for disability (moderate or severe) were chronic epilepsy in 3 patients (19%) and visual field deficits in 4 patients (25%). Notably, among the 5 patients with absolute homonymous visual field deficits, 2 (40%) did not consider themselves disabled and returned to their routine daily activities and work. Four patients underwent neuropsychological examination because of their visual problems. A patient who developed a contralateral hemiparesis immediately after removal of a parietooccipital region cavernoma improved completely and was not disabled.

Discussion

Surgery of Occipital Cavernomas

Progressive neurological impairment, including visual field deficits and seizures with or without hemorrhage, is the most common indication for surgical removal of occipital cavernomas. However, since data on the natural history risks of these lesions are scarce, the treatment strategy usually depends solely on institutional experience and traditions. In our study, we found that microsurgical removal of occipital cavernomas might be associated with a significant risk of visual field deficits since 75% of patients experienced visual problems after surgery. Furthermore, only 40% of those who presented with visual field deficits experienced improvements. The risk of surgery-related visual field deficits was correlated with distance of the cavernoma from the cortical surface; removal of a lesion located more than 2 cm from the cortical surface was associated with visual field problems in 75% of cases (p = 0.034). By contrast, a visual field deficit appeared postoperatively in only 17% of patients with a superficial cavernoma. In our opinion, there are several reasons for this. 1) A lesion with pial extension can be identified immediately after dural opening, therefore not necessitating any additional dissection of the parenchyma. Using a high magnification of the operating microscope and appropriate microneurosurgical techniques, one may avoid any significant damage to the surrounding cortex. Even if a lesion is hidden under the cortex but is no deeper than 2 cm, it will usually be recognized easily by typical brain discoloration, which spreads for several millimeters outside the cavernoma. 2) Since the cortex is thicker over the crest of a convolution and thinner at the depth of a sulcus, the transgyral approach is thought to sacrifice more neurons than transsulcal dissection.

However, disruption of the arcuate U-fibers during transsulcal exposure is not proven to be less detrimental than disruption of vertical projection fibers with the transgyral approach. Consistent with this, we did not find any correlations with better visual outcome using either of these approaches in our patients. The mean depth of cortical sulci is around 2 cm, which explains why during removal of cavernomas located deeper than 2 cm from the cortical surface, damage of some fibers is almost unavoidable even when using delicate transsulcal dissection. Consequently, the deeper the lesion, the more injury is likely to be caused to either arcuate or vertical visual fibers during surgery, and although the cortex remains undamaged, patients will still experience visual field deficits.

Primary Visual Cortex, Optic Radiation, and Higher Visual Processing

When dealing with deep occipital cavernomas, one...
should pay attention to the anatomical background of the optic radiation and striate cortex and inform the patient about potential visual disorders. If the lesion is located in the occipital cortex, homonymous visual field deficits are very similar in both eyes, that is, they extend to the same angular meridian (congruity). The more congruous the visual field deficits, the nearer the lesion is to the occipital cortex, while optic tract lesions produce less similar visual field deficits in the eyes (incongruous). Cerebral visual impairment is a deficit of visual function caused by damage to the retrogeniculate visual pathways. There are 2 visual pathways, the dorsal stream and the ventral stream, the former of which passes between the occipital and posterior parietal lobes and the latter between the occipital and temporal lobes. Damage to the dorsal stream can impair higher visual processing, producing, for example, impaired depth and motion perception, difficulties in interpreting a visually complex environment, and hemispatial neglect. Damage to the ventral stream can impair the ability to recognize people and understand facial expressions and may cause difficulties with orientation. In the preoperative workup, in addition to visual field deficits, the clinical picture can also include visual-cognitive disorders, leading to increased disability and a negative effect on social behavior and learning. In the literature, diffusion tensor imaging tractography of the optic radiation has shown promising results with regard to mapping of the visual tracts, which may be useful during intraoperative navigation within the occipital white matter, thus improving postoperative results.

In our study, 3 patients had persistent central disorders of vision that were mainly emphasized in dorsal stream dysfunction in neuropsychological testing. One patient had ischemic sequelae after surgery, followed by difficult neuropsychological symptoms manifesting with visual spatial defects, disturbed visual memory, and neglect.

**Visual Field Outcome**

In our patient series, 31% of patients had pretreatment visual field deficits. Neuroophthalmological examination performed a median of 5.25 years postoperatively showed that visual disorders remained unchanged if they were present immediately after surgery, confirming their very poor long-term recovery potential. Furthermore, in only 2 of 5 patients the preoperative visual field deficits disappeared. Many patients learned to tolerate visual field deficits quite well. In our series, 2 of 12 patients with postoperative visual problems did not notice them, although later perimetry tests confirmed these deficits. These could have been missed before surgery since not all patients underwent complete neuroophthalmological examination preoperatively. Undoubtedly, this is a limitation of the study. The extent of toleration of absolute homonymous hemianopia varied significantly, emphasizing the importance of prompt psychological aid.

Occipital region lesions seem to have a similar hemorrhage rate as other supratentorial cavernomas. None of the patients experienced massive life-threatening bleeding, and their visual outcome did not differ from that of nonhemorrhagic patients. Furthermore, rebleeding did not carry additional risks of permanent visual field deficits compared with the nonhemorrhagic group. However, because of the relatively small number of patients in our series, some correlations might have been underestimated.

**Seizure and General Outcome**

Favorable epilepsy outcome was registered in only 55% of patients after removal of an occipital cavernoma. This is worse than removal of cavernomas in other supratentorial locations, for example, the frontal or temporal lobe, where postoperative recovery from epilepsy occurs in 70%–84% of patients. This may be related to less extensive hemosiderin resection during cavernoma removal due to high eloquence of the occipital region. Nevertheless, no consistent data have emerged with regard to the usefulness of removing additional hemosiderin-stained parenchyma during cavernoma removal. While some studies have shown additional resection to be associated with better epilepsy outcome, a recently published retrospective meta-analysis of 31 studies on epilepsy outcome after supratentorial cavernoma removal in 1226 patients revealed no benefits of such strategy on long-term seizure outcome. In our department, we routinely perform additional removal of perilesional parenchyma in noneloquent regions of the brain. Undoubtedly, this issue warrants further studies.

The most common cause of disability after surgery was visual field deficits, occurring in 25% of patients. According to the grading system of operated cavernomas proposed by us earlier (Table 4), occipital region cavernomas presenting with visual field deficits may be assigned to Grade 2 lesions, with the estimated favorable outcome (Glasgow Outcome Scale Score 5) being 79%. Confirming this, 80% of our patients with Grade 2 cavernomas had no disability in the long-term follow-up. Such information might be useful in preoperative discussions with patients and their relatives on the risks of surgery. In the long-term follow-up, epilepsy caused disability (Glasgow Outcome Scale Score 4) in 4 of our patients, with 1 patient being severely disabled. This emphasizes the importance of timely treatment of patients with seizures since a long duration of epilepsy before surgery may be associated with worse seizure outcome.

**Conclusions**

In occipital cavernomas, operative risks should be weighed against natural history. Surgical removal of these lesions carries a significant risk of postoperative disability, and early recognition of visual field deficits can help in planning the surgical strategy.

**Table 4: Scoring in the grading system of brain and spinal cavernomas**

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<th>Variable</th>
<th>Score</th>
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<td>location</td>
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<tr>
<td>basal ganglia, infratentorial, spinal cord</td>
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</tr>
<tr>
<td>supratentorial</td>
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</tr>
<tr>
<td>focal neurological deficit</td>
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*From Kivelev et al.*
visual field deficits, occurring in 75% of patients in our series. Lesions located deeper than 2 cm from the cortical surface were associated with a 4.4-fold risk of visual field impairment relative to superficial cavernomas. Furthermore, only 55% of patients were seizure free postoperatively. Disability was mainly associated with visual field deficits and epilepsy. These findings should be taken into account when considering surgery. Further improvement of microneurosurgical techniques and preoperative functional radiological assessment are needed to diminish surgery-related risks in patients with symptomatic occipital cavernomas.

**Disclosure**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Kivelev. Analysis and interpretation of data: Kivelev, Koskela. Critically revising the article: Setälä, Niemelä, Hernesniemi.

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