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Review article

Treatment decision for occipital arteriovenous malformations (AVMs) to achieve hemorrhagic control while maximizing visual preservation: Our experience and review of literature

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ABSTRACT

Despite concern of hemorrhagic risk, patients with occipital AVMs are at significant risk for visual disturbances after treatment. We aim to characterize the hemorrhage risk and visual disturbance in occipital AVMs patients from our experience and literature review. We performed retrospective review of occipital AVM patients seen at our institution from 1990 to 2015. Patient characteristics were compared using multivariable logistic regression with follow-up visual disturbance as the outcome. We also systematically reviewed the PubMed database for English literature describing occipital AVMs (with exclusion of case reports). Ninety-seven patients satisfied inclusion criteria for our study. Mean age was 34.9 ± 16.4 years, with 50.5% male. Thirty-one (32.0%) presented with hemorrhage, and 32 (33.0%) presented with visual disturbance. Average AVM size was 4.0 ± 2.5 cm. Twenty-five (25.8%) were conservatively managed, 13 (13.4%) underwent surgery, and the rest were managed by radiosurgery (52.6%) or embolization (8.2%), with an obliteration rate of 38.9% in treated patients. During average follow-up of 5.4 years, 6 patients (6.7%) hemorrhaged yielding an annual hemorrhage rate of 1.2% for all patients, and 0.0% for surgically-treated patients. Thirty-seven (38.3%) patients experienced visual disturbance in some capacity, nineteen (21.1%) had de novo visual disturbance, fourteen of which were surgically treated patients (19.4%). Multivariable analysis reveals visual disturbance at presentation ($p = .012$) and microsurgery ($p = .047$) are significantly predictors of follow-up visual disturbance. While hemorrhage control remains the primary goal of AVM treatment, visual preservation in occipital AVMs is also a major concern. Recommending patients for microsurgery should be weighed carefully and individualized as it bears the highest risk of visual field disturbance despite most optimal hemorrhage control.

1. Introduction

Intracranial arteriovenous malformations (AVMs) are relatively rare and bear an extensive hemorrhage risk. Existing literature reports risk of hemorrhagic presentation between 30% and 82% [1–6], and subsequent annual bleeding rates between 1.9% and 4.61% [2–4,6–9].

Approximately 5% of all AVMs are occipital [10], and are unique in that their involvement of the visual cortex predisposes to visual disturbance (most commonly visual field deficits), at presentation or after treatment. Approximately 37–51% of patients reportedly present with visual disturbance [10–12], and 5–10% of treated patients experience worsening or new visual deficits [10]. Occipital AVM patients are also at greater risk of non-hemorrhagic, migraine-like headache as evidenced by literature reporting that 55%–60% of all occipital AVM patients present with such headaches [10,12].

Present treatment strategies for occipital AVMs are directed toward reduction or elimination of bleeding risk provided the significant morbidity and mortality associated with intracerebral hemorrhage (ICH) [3,13,14]. However, the eloquence of occipital lesions necessitates that treatment selection maximize visual preservation while achieving the primary goal of lesion obliteration. By report, headaches respond reasonably to treatment, whereas vision improvement is minimal when compared to conservative management. Notably, worsening of existing or induction of de novo visual disturbances appears most pronounced when patients are treated with microsurgery [10].

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Literature describing occipital AVMs remains limited, and to our knowledge, only one report compares treatment outcomes between varying treatment modalities and presents the associated, calculated annual hemorrhagic risk [10]. Optimal treatment decisions for occipital AVMs requires additional studies to confirm previous findings. Here we provide a comparative description of our institutional experience with occipital AVM treatment, and a systematic review of the literature.

2. Methods

2.1. Study cohort selection

We performed a retrospective chart review of all occipital AVM patients seen at our institution between 1990 and 2015. All information was collected into an IRB-approved institutional AVM database that was both retrospectively and prospectively maintained. Patients with AVMs involving the occipital lobe as confirmed by angiography were included in the study. Exclusion criteria included missing clinical or angiographic data, patients lost to follow-up, or those with hereditary hemorrhagic telangiectasia.

2.2. Variable definitions

Demographic, clinical, imaging, and follow-up data were collected. Age was defined as age at the diagnosis of AVM, and race as White, Black, Asian, and Others. Pretreatment functional status was inferred from provider clinical notes and represented using modified Rankin Scale (mRS). Visual disturbance was defined as consistent visual blur, visual field cut, or complete vision loss. Size of AVM was measured in centimeters. Lesion eloquence was defined as visual cortex involvement. Management modality was distinguished into four distinct categories: Surgery ± Embolization (SE), Radiosurgery ± Embolization (RE), Embolization only (Emb), and Observation (Obs). Follow-up duration was defined as the interval between diagnosis to last follow-up for untreated patients, and first treatment to last follow-up for treated patients. Lesion obliteration was confirmed by digital subtraction angiography (DSA). Lesions in patients undergoing radiosurgery with obliteration confirmed by magnetic resonance angiography (MRA) were also considered to be obliterated.

2.3. Treatment selection

Treatment selection for occipital AVMs at our institution is a shared decision making process between the attending physician and the patient and their family, and is individualized extensively to accommodate the daily functional needs of the patient. As a general protocol, for accessible lesions with acceptable surgical risk, microsurgery was recommended; whereas for those not amenable to surgery, radiosurgery was recommended. Non-curative embolization for high-flow pedicles may be considered, and at times is the only acceptable treatment provided the risk of surgery and radiosurgery, especially for occipital AVMs with involvement of critical structures such as thalamus or basal ganglia. Conservative management is generally recommended for AVMs with relatively low risk of hemorrhage with high treatment risk profile.

2.4. Statistical analysis

To explore potential factors associated with follow-up visual disturbance, patient baseline characteristics and outcomes were compared between those with follow-up visual disturbances and those without. Student's *t*-test or analysis of variance (ANOVA) was used for continuous variables and Chi-square or Fisher's exact test was used for categorical variables where appropriate. Significant variables were then considered for further examination using univariable and multivariable logistic regression. Factors not statistically significant but that portend critical clinical significance were also included in the analysis. Initial variable selection for multivariable logistic regression employed a $p < .10$ threshold; inclusion for clinically significant variables is more selective than univariable due to limited sample size. For reported statistical results, significance was defined as $p < .05$, and all p values were reported as two-sided. Statistical analyses were performed using R Statistical Software (Version 3.2.3, Vienna, Austria).

2.5. Literature review

A systematic approach was utilized for literature review of published occipital AVMs on PubMed. The search strategy was developed by the first author (W.Y.) who has previous formal training in library science informatics. Inclusion criteria consisted of: (1) studies reporting on occipital AVMs, (2) English literature, and (3) case series. Exclusion criteria included: (1) studies describing "vascular malformations" including malformations of other type, (2) AVM studies not focused on occipital AVMs, (3) non-English literature, (4) case reports, (5) occipital AVMs study with non-extractable basic clinical, angiographic, or outcome information, and (6) articles with extractable full text. Literature was obtained exclusively from PubMed. Our search strategy is depicted as follows: (AVM [Title/Abstract] *OR* AVMs [Title/Abstract] *OR* Arteriovenous Malformation [Title/Abstract] *OR* Arteriovenous Malformations [Title/Abstract]) *AND* (Occipital [Title/Abstract]).

We reviewed full text for all results returned from the search strategy. Literature screening and data extraction was performed independently by two reviewers, and we included study period, number of patients, demographics, clinical data (such as ruptured presentation, type and frequency of visual disturbances), and angiographic data (such as size, venous drainage pattern, Spetzler-Martin grading) on patients, type of treatment, follow-up length, treatment outcomes and adverse events such as hemorrhage at follow-up in our data collection form. Risk of bias was assessed for included studies, and results from each study were individually summarized. Data aggregation was performed in studies where data was available for each variable.

3. Results

3.1. Study population and baseline characteristics

From our database of 708 sporadic AVMs, a total of 123 patients (17.4%) were found to have a lesion involving the occipital lobe. Incomplete baseline data including missing angiographic data or critical clinical data was observed in 15 patients, and 11 patients were missing follow-up data. Our final study cohort was therefore comprised of 97 patients, with 25 (25.8%) in the Obs group, 13 (13.4%) in SE, 51 (52.6%) in RE, and 8 (8.2%) in the Emb group. Fig. 1 details the patient selection process. Average age of all patients was 34.9 ± 16.4 years (range: 3.4–81.1 years), with 50.5% being male. Race distribution was: White (n = 77, 79.4%), Black (n = 6, 6.2%), Asian (n = 5, 5.2%) and Other (n = 9, 9.3%).

Among all patients, 32.0% presented with hemorrhage attributable to AVM rupture. There was no difference in distribution of rupture presentation between patients with and without follow-up visual disturbances ($p = .137$). Headache at presentation was found in 71.1% of patients and visual disturbance in 33.0%. Patients presenting with visual disturbance were more likely to experience subsequent follow-up visual disturbances ($p = .010$). As shown in Table 1, most patients had a mRS of 1 (37.1%) or 2 (39.2%) before

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