Long-Term Recurrence and Risk of Second Surgeries after Resection of Symptomatic Cavernous Malformations in Pediatric Patients

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Disclosures

- None
Introduction

- **Cavernous malformations (CMs)** are dilated capillary vessels that lack a blood-brain barrier and are prone to focal microhemorrhage and hemosiderin deposition\(^1\)

- CMs are the *second most common vascular malformation* to developmental venous anomalies (DVAs) and account for 10-15% of all vascular malformations\(^2\)

- However, **data on the natural history of pediatric CMs after resection** are limited

Methods

- Children (age ≤ 18) receiving surgery at our institution for removal of a CM between January 1996 and December 2017 were identified
  - Approval was granted by our university’s Institutional Review Board

- Children not receiving surgery for CM removal were not included

- Primary outcomes were lesion recurrence, progression, and secondary CM resections

- Multivariable regression models (logistic regression and Cox proportional hazards) were used to evaluate factors associated with primary outcomes of interest
Risk of Recurrence, Progression, and *De Novo* Lesions

**Panel A.** Progression/recurrence risk after CM resection stratified by resection extent

**Panel B.** Risk of *de novo* lesions stratified by number of CMs at initial resection
Secondary Surgeries after Initial Resection

Panel A. Causes of secondary surgeries

Panel B: Risk of subsequent surgeries stratified by number of CMs at initial resection

Panel C: Risk of subsequent surgeries stratified by resection extent of initial lesions

Panel D: Identification of risk factors independently associated with risk of secondary surgeries. Significant covariates are indicated by a red symbol.

*Only including surgeries for residual or recurrent lesions.
Acute hemorrhage and multiple CMs at presentation were the only factors independently associated with need for secondary surgeries.

We established the following heuristic risk model:

- **Low Risk**: No hemorrhage and single CM at initial presentation
- **Medium Risk**: Acute hemorrhage OR multiple CMs at initial presentation
- **High Risk**: Acute hemorrhage AND multiple CMs at initial presentation
Potential Treatment Guidelines

Based on the risk model we proposed, a potential series of treatment and monitoring guidelines may be constructed:

Low risk patients may not require extended follow-up as risk of developing additional symptomatic lesions requiring secondary surgeries is extremely low.

Medium and high risk patients may benefit more from long-term follow-up by radiographic imaging and may also consider genetic testing for predisposing factors.
Discussion

- Gross total resection of CMs did **reduce the incidence of subsequent surgeries for residual or recurrent lesions** compared to subtotal resection.

- However, most secondary surgeries were for either **pre-existing asymptomatic lesions** or for **de novo lesions**.

- Multivariable analysis identified **acute hemorrhage** and the **presence of multiple CMs at initial presentation** as independently prognostic factors for increased **risk of subsequent surgeries for symptomatic CM**.

- Additional **external validation** of our model is necessary.
Summary Points

- Most newly symptomatic lesions after initial CM resection are either previously asymptomatic or de novo

- Acute hemorrhage at diagnosis and number of initial CMs present are risk factors for secondary CM resection surgeries

- Based on a simple risk model, high and medium risk patients should be monitored for longer and more frequently than low risk patients