Intracerebral hemorrhage from ruptured cerebral arteriovenous malformation

A brain arteriovenous malformation may not cause any signs or symptoms until the AVM ruptures, resulting in bleeding in the brain (hemorrhage). In about half of all brain AVMs, hemorrhage is the first sign. But some people with brain AVM may experience signs and symptoms other than bleeding related to the AVM.

The most common presentation of a pial arteriovenous malformation (AVM) is spontaneous intracerebral hemorrhage (ICH). The peak incidence of ICH is early in the third decade of life.

Outcome

Ruptured Intracranial Arteriovenous Malformation rupture is highly morbid. Outcomes after Intracranial Arteriovenous Malformation rupture differ from other types of brain hemorrhage. There are no specific widely used grading systems designed to predict clinical outcome after AVM rupture.

To develop an all-comers scoring system to grade patients with AVM rupture and predict clinical outcome more accurately than grading systems currently in use.

Silva et al. retrospectively reviewed patients who presented to the Department of Neurosurgery, Brigham and Women's Hospital, and Jackson Memorial Hospital, with a ruptured AVM. Using change in modified Rankin Scale (mRS) as the response variable, they generated an ordinal logistic regression model to test for significant predictor variables. The full model was sequentially condensed until the simplest model with the highest area under the receiver operating curve (AUROC) was achieved.

A total of 115 patients who presented with ruptured AVMs were included in the study, with a mean follow-up time of 4 yr. The Ruptured AVM Grading Scale (RAGS) consists of the Hunt and Hess (HH) score (1-5), patient age (<35 = 0, 35-70 = 1, and >70 = 2), deep venous drainage (1), and eloquence (1). The RAGS score outperformed other neurosurgical grading scales in predicting change in mRS, with an AUROC greater than 0.80 across all follow-up periods.

The RAGS score is a simple extension of the Hunt and Hess Stroke Scale that predicts clinical outcome after AVM rupture more accurately than other grading systems.

It carries significant morbidity and mortality. This complication is particularly important in the young and otherwise healthy population, in whom AVMs cause up to one-third of all hemorrhagic strokes. A previous rupture is the single most important independent predictor of future hemorrhage. Current treatment modalities for AVM are microsurgery, endovascular embolization, and radiosurgery. In acutely ruptured AVMs, early microsurgical excision is usually avoided. The standard is to wait at least 4 weeks to allow for patient recovery, hematoma liquefaction, and inflammatory reactions to subside. Exceptions to this rule are small, superficial, low-grade AVMs with elucidated angioarchitecture, for which early simultaneous hematoma evacuation and AVM excision is feasible. Emergent hematoma evacuation with delayed AVM excision (unless, as mentioned, the AVM is low grade) is recommended in patients with a decreased level of consciousness due to intracranial hemorrhage, posterior fossa or temporal lobe hematoma of >30 ml, or hemispheric hematoma of >60 ml. The applicability of endovascular techniques for acutely ruptured AVMs is not clear, but feasible options, until a definitive
treatment is determined, include occluding intranidal and distal flow-related aneurysms and 'sealing' any rupture site or focal angioarchitectural weakness when one can be clearly identified and safely accessed. Radiosurgery is not performed in acutely ruptured AVMs because its therapeutic effects occur in a delayed fashion.  

Case series

241 consecutive AVM patients (mean age 37+/-16 years, 52% women) from the prospective Columbia AVM Databank initially presenting with hemorrhage were evaluated using the Rankin Scale (RS) and the National Institute of Health Stroke Scale (NIHSS). From the 241 AVM patients, 29 (12%) had subsequent intracranial hemorrhage during follow-up. For further comparisons, 84 non-AVM patients with intracerebral hemorrhage from the Northern Manhattan Study (NOMAS) served as a control group.

Results: In 241 AVM patients presenting with hemorrhage the median RS was 2 and the median NIHSS was 1 (49% RS 0 to 1, 61% NIHSS <2). The median time between hemorrhage and clinical evaluation was 11 days (mean 219 days). Recurrent AVM hemorrhage during follow-up resulted in no significant increase in morbidity (median RS 2, P=0.004; median NIHSS 3, P=0.322; time between hemorrhage and study evaluation: median 55 days, mean 657 days). Among AVM-hemorrhage subtypes, parenchymatous AVM hemorrhage was associated with higher stroke morbidity (odds ratio, 2.9; 95% CI, 1.5 to 5.8 for NIHSS > or =2) than nonparenchymatous hemorrhages. Parenchymatous AVM hemorrhage had a significantly better outcome (median NIHSS 1) than non-AVM related hemorrhage (median NIHSS 12; P<0.0001).

Conclusions: Hemorrhage, either at initial presentation or during follow-up of untreated AVM patients appears to carry a lower morbidity than intracranial hemorrhage from other causes. These findings support a careful weighing of risks from interventional treatment and natural history.

Case reports

a 4-year-old boy with a spontaneous intracerebral hemorrhage (ICH) related to an arteriovenous malformation (AVM) that was not found with good-quality magnetic resonance (MR) imaging and MR angiography. Both modalities were used serially in the acute phase and at 2 and 7 months of follow-up. Digital subtraction angiography identified the peripheral AVM when the patient experienced rehemorrhaging 1 year after his initial presentation. This case illustrates the need for a complete diagnostic evaluation including conventional angiography in cases of idiopathic ICH in children.