Ruptured cerebral arteriovenous malformation

Rupture risk

Data suggest that the risk of cerebral arteriovenous malformation rupture is not increased during pregnancy and the puerperium. Therefore, Liu et al. would not advise against pregnancy in patients with intracranial AVM. However, they found that AVM rupture usually occurred in late gestational age; therefore, more attention should be provided to maintain relative hemodynamic stability in these patients during mid to late pregnancy. The issues facing patients with ruptured AVM in pregnancy remained challenging and required multidisciplinary management including neurosurgeons, obstetricians, pediatricians, and intensivists 1).

Outcome

Ruptured cerebral arteriovenous malformation often present as intracerebral hemorrhages (ICH), associated with 10 % to 30 % mortality.

Treatment

Patients who present with a hemorrhage from an AVM should be initially stabilized according to acute management guidelines for ICH. The characteristics of a lesion including its size, location in eloquent tissue, and high-risk features will influence risk of rupture, prognosis, as well as help guide management decisions. Given that rupture is associated with an increased risk of 6 % re-rupture in the year following the initial hemorrhage, versus 1 % to 3 % predicted annual risk in non-ruptured lesions only, definitive treatment is encouraged after ICH stabilization. A rest period of 2 to 6 weeks after hemorrhage is recommended before definitive treatment to avoid disrupting friable parenchyma and the hematoma. Treatment may consist of endovascular embolization, surgical resection, radiosurgery, or a combination of these three interventions based on the lesion 2).

Case series

2017

Data of patients with AVM were collected prospectively. Cases were identified in which an AVM was resected and an associated space-occupying ICH was evacuated at the same time, and divided into “group 1,” in which the surgery was performed acutely within 48 h of presentation (secondary to elevated intracranial pressure); and “group 2,” in which selected patients were operated upon in the presence of a liquefying ICH in the “subacute” stage. Clinical outcomes were assessed using the modified Rankin Scale, with a score of 0 to 2 considered a good outcome. Obliteration rates were assessed using postoperative angiography.

From 2001 to 2015, 131 patients underwent microsurgical resection of an AVM, of which 65 cases were included. In “group 1” (n = 21; Spetzler-Ponce class A = 13, class B = 5, and class C = 3), 11 of 21 (52%) had a good outcome and in 18 of 19 (95%) of those who had a postoperative angiogram the AVMs were completely obliterated. In “group 2” (n = 44; Spetzler-Ponce class A = 33, class B = 9, and class C = 2), 31 of 44 (93%) had a good outcome and 42 of 44 (95%) were obliterated with a single procedure. For supratentorial AVMs, the ICH cavity was utilized to provide an operative trajectory to a deep AVM in 11 cases, and in 26 cases the ICH cavity was deep to the AVM and hence facilitated the deep dissection of the nidus.

In selected patients the presence of a liquefying ICH cavity may facilitate the resection of AVMs when
performed in the subacute stage resulting in a good neurological outcome and high obliteration rate.

Two hundred eighty-eight patients with cerebral AVMs underwent microsurgical resection between 1983 and 2012 performed by the same surgeon (Johannes Schramm). This is a prospective case collection study that represents a consecutive series. The results are based on prospectively collected, early-outcome data that were supplemented by retrospectively collected, follow-up data for 94% of those cases. The analyzed data included the initial presentation, Spetzler-Martin grade, obliteration rates, surgical and neurological complications, and frequency of pretreatment with embolization or radiosurgery. The total cohort was compared using “small-AVM,” Spetzler-Martin Grade I and II, and ARUBA-eligible AVM subgroups. RESULTS The initial presentation was hemorrhage in 50.0% and seizures in 43.1% of patients. The series included 53 Spetzler-Martin Grade I (18.4%), 114 Spetzler-Martin Grade II (39.6%), 90 Spetzler-Martin Grade III (31.3%), 28 Spetzler-Martin Grade IV (9.7%), and 3 Spetzler-Martin Grade V (1.0%) AVMs. There were 144 unruptured and 104 ARUBA-eligible cases. Preembolization was used in 39 cases (13.5%). The occlusion rates for the total series and small AVM subgroup were 99% and 98.7%, respectively. The mean follow-up duration was 64 months. Early neurological deterioration was seen in 39.2% of patients, of which 12.2% had permanent and 5.6% had permanent significant deficits, and the mortality rate was 1.7% (n = 5). Outcome was better for patients with AVMs smaller than 3 cm (permanent deficit in 7.8% and permanent significant deficit in 3.2% of patients) and Ponce Class A status (permanent deficit in 7.8% and significant deficit in 3.2% of patients). Unruptured AVMs showed slightly higher new deficit rates (but 0 instances of mortality) among all cases, and in the small AVM and Ponce Class A subgroups. Unruptured Spetzler-Martin Grade I and II lesions had the best outcome (1.8% permanent significant deficit), and ARUBA-eligible Spetzler-Martin Grade I and II lesions had a slightly higher rate of permanent significant deficits (3.2%). CONCLUSIONS Microsurgery has a very high cure rate. Focusing microsurgical AVM resection on unruptured lesions smaller than 3 cm or on Spetzler-Martin Grade I and II lesions is a good strategy for minimizing long-term morbidity. Well-selected microsurgical cases lead to better outcomes than with multimodal interventions, as in the ARUBA treatment arm, or conservative treatment alone. Long-term prospective data collection is valuable.

2015

A retrospective study of 139 consecutive patients operated for a ruptured cerebral arteriovenous malformation (rAVM) between 2002 and 2012.

The age at diagnosis and the WFNS score were recorded for each patient before treatment. All patients were re-evaluated 3 months after treatment using the modified Rankin Scale (mRS). Conventional angiography was performed in the first 2 postoperative weeks and then a year later to detect any remnant or recurrence.

The mean age at diagnosis was 30.8 years (range 4-69 SD: ±5) and 44 patients had an age at diagnosis <18 yo. The mRS score 3 months after treatment was ≤2 in 104 patients (83%). Predictive factors of good functional outcome were age at diagnosis <25 yo, initial WFNS score ≤ 2, SPM grade ≤ 2 and absence of acute hydrocephalus (p<0.05). Complete obliteration was obtained in 123 patients (89.5%) after the first microsurgical treatment. Early postoperative conventional angiography revealed a rAVM remnant in 16 patients (10.5%). Late conventional angiography showed a recurrence in 6 patients (4.5%). All of them were <18 yo. Predictive factors of postoperative rAVM remnant were an initial WFNS score >2, SPM grade >2 and preoperative evaluation limited only to CT angiography in
emergency situation (p<0.05).

**Functional outcome after microsurgery** was good in 83% of patients with rAVM. Good results were also recorded in 28% of patients with poor initial neurological status and severe intracerebral hemorrhage, which required immediate surgery. In case of remnant, a further treatment should be decided in a true multidisciplinary discussion to protect the patient from any rebleeding ⁵).

**2010**

Wong et al retrospectively reviewed patients with bAVMs, aged 18 or below, managed at their hospital between January 1992 and December 2008. Clinical outcome was assessed using the modified Rankin Scale (mRS). Patients with ruptured bAVMs were analyzed; their clinical parameters and computerized tomography findings on admission were recorded. Clinical outcome was then evaluated against admission scores using the Glasgow Coma Scale (GCS), the World Federation of Neurosurgical Societies grading of Subarachnoid Hemorrhage (WFNS-SAH), the Spetzler-Martin AVM grading system, the ICH Score and 2 other independent parameters, namely pupillary response and significant focal neurological injuries. Spearman's rank correlation coefficient, linear regression analysis and multivariate logistic regression analysis were used for data analysis.

40 pediatric patients with bAVMs were found and 32 of them presented with hemorrhage (80%). In the 32 children with ruptured bAVMS, follow-up ranged between 7 and 204 months (median 100.5 months). The complete excision/obliteration rate as confirmed by digital subtraction angiography was 73.3%. The mRS scores at the last follow-up were: grades 0-II in 87.5%; grades IV-V in 6.25%, and grade VI in 6.25%. The Spetzler-Martin grade, the intracerebral hemorrhage score, the WFNS-SAH grade, the GCS scores, and a combined scoring scale consisting of the GCS, pupillary response and significant focal neurological injuries correlated significantly with clinical outcome 6 months after hemorrhage. However, on the scatter diagrams, it appeared that only the combined scoring scale might be valid for clinical practice. Multivariate logistic regression analysis showed that the combined scoring scale was a statistically significant independent predictor of clinical outcome 6 months after hemorrhage.

In this series of pediatric patients with ruptured bAVMs, although various grading scales correlated significantly with clinical outcome 6 months after hemorrhage, only the combined scoring scale might have the potential to be applied to predict clinical outcome in these children ⁶).


