Arteriovenous malformation (AVM)

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Case Presentation

• 17 y/o female presented with dizziness, vertigo, and horizontal nystagmus for approximately 5 years

• Continued to have progressive symptoms when a pediatric neurologist ordered a MRI of the brain

• Found to have an AVM measuring 1.9 cm x 0.7 cm x 1.6 cm in the left cerebellar pontine cistern
Case Presentation (Arteriogram)
Background

• Arteriovenous malformation (AVM) is a congenital vessel abnormality with abnormal communication between artery and vein
• Nidus is a tangle of arteries and veins connected by at least one fistula
• Bleeding risk is 2 to 4% per year
Presentation

- Intracerebral hemorrhage (most common)
- Seizure
- Headache
- Focal neurologic deficit
- Incidental finding
Work-up

• CT of the head to rule out cerebral bleeding
• Angiography is gold standard

Optional

• CT angiography for vasculature
• MR angiography (MRA) and venography (MRV) for relationship of vasculature to normal brain structures
• Functional MRI (fMRI) to determine eloquent areas
# Spetzler-Martin Scale

<table>
<thead>
<tr>
<th></th>
<th>0 points</th>
<th>1 point</th>
<th>2 points</th>
<th>3 points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diameter</td>
<td>&lt;3 cm</td>
<td>3-6 cm</td>
<td>&gt;6 cm</td>
<td></td>
</tr>
<tr>
<td>Location</td>
<td>Noneloquent area</td>
<td>Eloquent area</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pattern of venous drainage</td>
<td>Superficial</td>
<td>Deep</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Adapted from Spetzler RF and Martin NA. A proposed grading system for arteriovenous malformations. *J Neurosurg* 1986; 65: 476-483.

The Spetzler-Martin scale is between 1-5. The higher the total point score, the worse the prognosis.
Treatment options

• Surgery
  – For Spetzler-Martin I-III score AVMs

• Radiosurgery
  – For lesions in deep or eloquent areas that are unresectable
  – For patients who cannot have surgery or do not desire to undergo surgery

• Endovascular embolization
  – For lesions <1 cm with one single arterial feeder
Treatment

• Goal of treatment is to obliterate the nidus
• Surgery has immediate benefits of decreased bleeding risk
• SRS takes 1-4 years to decrease bleeding risk
  – During the latent period, 54% decrease in bleeds
  – After obliteration, 88% decrease in bleeds
Treatment continued

• Aneurysms greater than 7 mm in diameter should be treated

• Do not embolize before radiation treatment since it can decrease the apparent size of the AVM and can lead to inadequate treatment because of geographic miss
Frame-based SRS Prescription

• Lesion average diameter < 3 cm: 21–22 Gy to the margin
• Lesion average diameter > 3 cm: 16–18 Gy to the margin
• For lesions in the brainstem, need to lower the dose
Dose-Response relationship for SRS in treatment of AVMs

<table>
<thead>
<tr>
<th>Minimum dose (Gy)</th>
<th>Percent in-field AVM Obliteration</th>
</tr>
</thead>
<tbody>
<tr>
<td>26.9</td>
<td>99</td>
</tr>
<tr>
<td>24.8</td>
<td>98</td>
</tr>
<tr>
<td>22.0</td>
<td>95</td>
</tr>
<tr>
<td>19.8</td>
<td>90</td>
</tr>
<tr>
<td>17.4</td>
<td>80</td>
</tr>
<tr>
<td>15.8</td>
<td>70</td>
</tr>
<tr>
<td>13.3</td>
<td>50</td>
</tr>
</tbody>
</table>

The lesion (orange arrow) is contoured (orange outline) on the day of Gamma Knife treatment on the arteriogram. The nidus is contoured alone while feeders are not included.
The contour (pink outline) is reproduced from the anteriogram on the MRI of the brain.
The patient received 18 Gy to the 50% isodose line.
A Randomised trial of Unruptured Brain Arteriovenous malformations (ARUBA) trial

• A trial reviewing preventive treatment of unruptured AVMs

• Patients were randomized to interventional therapy (surgery, embolization, SRS, or combination) or medical management alone

• Trial was stopped early when medical management alone was found to be superior
  – 10.1% (medical management group) versus 30.7% (interventional therapy group) for risk of death or symptomatic stroke
A Randomised trial of Unruptured Brain Arteriovenous malformations (ARUBA) trial

• Controversies
  – Short follow-up would detect procedure-related complications but not potential long-term benefits of prophylactic interventions
  – The intervention group had multiple interventions, which were grouped together
    • If one is inferior, could increase complication rate in entire group
Toxicities from SRS

• Early side effects include seizures, nausea, vomiting, headache

• Delayed side effects include seizures, radionecrosis, edema, venous congestion, cyst formation

• Risk of symptomatic adverse effects are 7% from RT treatment with 2.8% permanent side effects at 5 years
  
  – Increased risk of permanent side effects in thalamic and brain stem AVMs
Toxicities from SRS Continued

• Related to location
  – Higher PIE is associated with higher likelihood of injury

• Related to size of AVM and RT dose
  – 12-Gy (V12) is strong predictor

<table>
<thead>
<tr>
<th>Location</th>
<th>Post-radiosurgery injury expression (PIE) classification</th>
</tr>
</thead>
<tbody>
<tr>
<td>Frontal lobe</td>
<td>1</td>
</tr>
<tr>
<td>Cerebellum</td>
<td>1</td>
</tr>
<tr>
<td>Temporal lobe</td>
<td>2</td>
</tr>
<tr>
<td>Parietal lobe</td>
<td>2</td>
</tr>
<tr>
<td>Occipital lobe</td>
<td>3</td>
</tr>
<tr>
<td>Basal ganglia</td>
<td>3</td>
</tr>
<tr>
<td>Medulla</td>
<td>4</td>
</tr>
<tr>
<td>Thalmus</td>
<td>4</td>
</tr>
<tr>
<td>Intraventricular</td>
<td>4</td>
</tr>
<tr>
<td>Pons</td>
<td>4</td>
</tr>
<tr>
<td>Corpus callosum</td>
<td>4</td>
</tr>
</tbody>
</table>

Follow-up

• H&P and MRI every 6 months for 1-3 years
• Angiography to confirm obliteration of the AVM usually at year 2 or 3
• If patient does not have complete obliteration after 3 years, consider re-treatment
References


