A 10 year-old young man was having breakfast with his family on Sunday morning. He complained of sudden onset of headache, walked to his mother, and became unresponsive. His father (a physician) said that he was unresponsive except for muttering incomprehensible words when stimulated. No abnormal movements were noticed.

Father carried the child to the car and drove him to the emergency department. The child arrived in the emergency department with diminished level of consciousness but with eyes open. He would not follow commands or speak but would look briefly at the examiner when questioned. Pulse was 50; BP 100/62; temperature 36.3; oxygen saturation 99%. Pupils were equal, mid-position, and responsive to light. Gaze tended to be downward. The boy would withdraw extremities to painful stimulation; muscle tone seemed diminished.

What is your differential diagnosis and how would you proceed?
Intracranial hemorrhage in a young man secondary to arteriovenous malformation rupture.

Background, Risk Factors, and Epidemiology

Subarachnoid hemorrhages and intracranial hemorrhages do occur in children but have different causes and treatments than in adults.

Arteriovenous malformations are complex tangles of abnormal arteries and veins interlinked by fistulas. The fistulas allow high flow arterio-venous shunting causing arterial hypotension in the surrounding areas of the brain.[1] Many AVM’s have associated saccular or berry aneurysms.

Arteriovenous malformations occur in 1 per 100,000 children per year and are four times more common than aneurysm as a cause of subarachnoid hemorrhage in children younger than the age of 15 years.[2] They typically present before age 45 and only about 20% of symptomatic AVM’s manifest before age 15.[3] Their course cannot be easily predicted and they may remain stable in size, grow, or even regress; autopsy data suggests that as few as 12% are symptomatic during life.[1]

Clinical Presentation

Intracranial hemorrhage is the most common clinical presentation of an AVM.[1] Symptoms range from headache to coma depending on the anatomic location of the arteriovenous malformation and the size and location of the hemorrhage. The headache is typically that of subarachnoid hemorrhage with sudden onset of a severe headache. If intraparenchymal hemorrhage occurs, stroke-like deficit may be present. It is thought that AVM’s account for an estimated 2% of acute strokes.

Again, clinical manifestations of the intracranial hemorrhage reflect the anatomic location of the mass and rate of expansion. For cerebellar hemorrhages as in the case discussed above, the classic clinical manifestations are inability to walk, ataxia, and headache though there are many exceptions to this imperfect triad. Repeated vomiting, cranial nerve abnormalities, and progressive decline in consciousness are other findings described with cerebellar hemorrhage. Patients may abruptly become comatose and apneic if the hemorrhage expands in the small posterior fossa and compresses the brainstem. Some generalities hold for cerebellar hemorrhage; generally lateral hemorrhages are better tolerated than medial hemorrhages. Salvage with good clinical outcome may occur even in comatose patients with prompt neurosurgical evacuation. Neurosurgical decompression may be life-saving in some cases. Smaller hemorrhages may be managed non-operatively.[4]

Seizures are another common initial symptom of patients with arteriovenous malformations. The majority of seizures are of partial or partial complex type. Headache is the presenting symptom in another group.[1]
One series from a tertiary referral center reported congestive heart failure from rapid arterial-venous shunting as the presentation of the AVM in 18% of their pediatric-only population; this was a common presentation in neonates.[3]

Finally, progressive focal neurologic deficits (without hemorrhage) may be another presentation. The deficits will again vary depending on the location of the arteriovenous malformation.

Emergency department care primarily involves patient stabilization and then identification, proper consultation, and admission.

**Imaging Studies, Consultations, and Procedures**

Noncontrast cranial CT is the initial imaging modality of choice since it will identify all intraparenchymal hemorrhages and the majority of subarachnoid hemorrhages. If the history suggests subarachnoid hemorrhage and initial CT is unremarkable, lumbar puncture is recommended.

Further neuroimaging studies will be obtained in consultation with other physicians. MRI identifies vascular anomalies but angiography is still preferred by many for finer anatomic detail and vascular flow information.

Immediate neurosurgical consultation is recommended with identification of the intracranial hemorrhage or arteriovenous malformation.

Treatment issues are complex. While it is agreed that the rate of recurrent hemorrhage is less than for aneurysmal hemorrhage, the rate of AVM rehemorrhage is still not clear. For patients that presented with hemorrhage, the annual rate of rehemorrhage was found to be 18% compared to a rate of 2% in patients with arteriovenous malformation who presented in a manner other than hemorrhage.[1]

Once hemorrhage has occurred, treatment is generally recommended with options including surgical resection and various interventional angiography procedures. Some report a rapid decrease in hemorrhage reoccurrence after the first year. Some studies suggest that for unruptured AVM’s, surgery does not improve overall outcomes.[5]

Arteriovenous malformations of the posterior fossa are uncommon accounting for less than one-quarter of intracranial AVM’s. The incidence of hemorrhage is reported to be higher with greater morbidity for AVM’s in this location. [6]

Surgery is generally recommended for AVM’s that have bled if they are accessible; other treatment options may include endovascular occlusive techniques or gamma-knife therapy. Again, the natural history of these malformations remains unclear.
Sudden Severe Headache and Unresponsiveness in a 10 Year-old Boy

Clinical course and outcome

After examination and establishment of intravenous access, the child was taken to neuroradiology with physicians in attendance for airway management if needed. CT confirmed a cerebellar hemorrhage and he was admitted to the pediatric intensive care unit with neurosurgical consultation. He was started on dexamethasone and famotidine. An MRI/MRA later that day showed a cerebellar arteriovenous malformation with the right superior cerebellar artery being the primary feeding vessel.

Level of consciousness was decreased on the second hospital day and he was not following commands or opening his eyes spontaneously; repeat CT showed hydrocephalus. A ventriculostomy was placed but this was not followed by improvement. A suboccipital craniotomy with resection of the AVM and clot removal was performed hours later. Postoperative course was unremarkable; repeated angiography showed no remnant of the arteriovenous malformation.

He was transferred to a rehabilitation hospital for therapy of ataxia and speech difficulties; these improved over two weeks. He currently engages in sports and performs well in school.
Sudden Severe Headache and Unresponsiveness in a 10 Year-old Boy

Annotated Bibliography

References tend to be either reviews or summaries of cases at one institution. There is nothing in the way of randomized controlled therapeutic trials for this condition.


   Masterful recent summary of literature on AVM’s from an international study group. Recommended for overview of the topic.


   Brief synopsis of several recent publications with listing of caveats for detecting and managing SAH in children.


   Summary of experience of a large tertiary referral center with emphasis on anesthetic techniques.


   A review of Dr. Fisher’s classic work on cerebellar hemorrhage contrasting diagnosis and treatment changes over the last forty years.


   A neurologist’s analysis of surgical and non-surgical treatments for arteriovenous malformations. Articulates the argument that for AVM’s that have not ruptured, surgical treatment offers no proven benefit.

Summary of experience from British referral center of their experience with posterior fossa AVM’s. These are unusual lesions and the natural history is not clear though the authors feel posterior fossa AVM’s are at greater risk of hemorrhage and morbidity that AVM’s in other locations.
Sudden Severe Headache and Unresponsiveness in a 10 Year-old Boy

Questions

1. The most common cause of spontaneous intracranial hemorrhage in children and young adults is:
   a. Aneurysm
   b. Arteriovenous malformation
   c. Tumor
   d. Moyamoya disease

2. The most common presentation of an arteriovenous malformation is:
   a. Hemorrhage
   b. Generalized seizure
   c. Focal neurologic deficit without hemorrhage
   d. Partial seizure

3. Which of the following is true regarding cerebellar hemorrhages?
   a. Coma is never a feature of cerebellar hemorrhage since the cortex is not involved
   b. Lateral cerebellar hemorrhages generally do better clinically than medial hemorrhages
   c. Most cerebellar hemorrhages are from rupture of an AVM
   d. Most cerebellar hemorrhages are from rupture of an aneurysm

4. If subarachnoid hemorrhage is strongly suspected, what is the diagnostic strategy of choice?
   a. Immediate CT
   b. Immediate CT followed by lumbar puncture if CT is non-diagnostic
   c. MRI- perfusion-weighted if possible
   d. Immediate angiography

5. The natural history of AVM’s is most consistent with...
   a. Steady growth until rupture
   b. Nonprogressive mass lesion
   c. Increasing steal phenomena leading to seizures
   d. Not clearly known
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Answers

1. **Answer b.**

The most common cause of spontaneous intracranial hemorrhage in children and young adults are ruptures of arteriovenous malformations. In adults, the most common cause of non-traumatic intracranial hemorrhage is aneurysmal rupture.

2. **Answer a.**

The most common presentation of an arteriovenous malformation is hemorrhage; seizures are the next most common presentation.

3. **Answer b.**

Lateral cerebellar hemorrhages generally do better clinically than medial hemorrhages; sometimes patients have only minimal residual neurologic deficits after evacuation of lateral hemispheric cerebellar hemorrhages.

4. **Answer b.**

Immediate CT followed by lumbar puncture if CT is non-diagnostic is the most common diagnostic strategy. CT is needed to exclude intraparenchymal hemorrhage and is thought to be 90-95% sensitive for subarachnoid hemorrhage; it is generally recommended that LP be employed to increase diagnostic sensitivity further.

5. **Answer d.**

The nature history of AVM’s is not known.