

INTRODUCTION

Pediatric AVM is a rare neurological insult that occurs in less than 1% of children (Berman et al., 2000). AVMs are arteriovenous shunts made up of feeding arteries, a mass of coiled vessels, and venous sinusoid spaces and veins. These vessels are connected directly without a capillary bed. The origin appears to be due to a failed differentiation of embryonic vessels into distinct arterial and venous networks, occurring between the third and twelfth week of fetal development. AVMs can range from several millimeters to several centimeters (DeVeber, 1999). Prior to rupture or after a hemorrhage occurs (through a warning leak or acute subarachnoid hemorrhage), AVMs initially present with seizures, headaches, and/or progressive neurologic deficits. They appear in two forms: hemispheric (the areas surrounding the main branches of the internal carotid artery) and deep midline (the area drained by the great vein of Galen). Surgery is warranted regardless of hemorrhage unless there are mitigating factors such as size, blood supply, location, and clinical condition of the child (Berg, 1994). In addition to operative procedures, nonsurgical and preoperative treatments which make high-grade lesions lower in grade and more suitable to resection, are all important secondary factors to consider. These treatments include angiogram, superselective embolization, irradiation, radiosurgery, and resection.

A dural arteriovenous fistulous malformation is an abnormal direct connection between a meningeal artery and a meningeal vein or dural venous sinus that result from injury/trauma, infection, or blood clotting in the vein. Borden Type III AV fistulas drain directly into subarachnoid veins. These veins can form aneurysms and bleed. Therefore, they need to be treated to prevent hemorrhage.

There is little in the neuropsychological literature regarding the course and outcome of pediatric AVM. A literature search revealed three case studies which demonstrated variable findings possibly due to factors such as the location and severity of AVM and length of time between surgery and follow-up. One case study found that psychological and physical abnormalities remitted at a 1 year follow-up (Walton, 1994), another found that most, but not all, abnormalities remitted by an 8-year follow-up (Eslinger, & Biddle, 2001; Eslinger et al., 1999), and a third case study found that several abnormalities persisted at a 7-year follow-up evaluation (Dikel et al., 2001). A recent case series describing the long-term psychosocial and adaptive outcomes in five participants with AVM (O'Toole, Borden, & Miller, 2006), found that the participants and their families were adapting variably, with more positive than negative findings. While overall emotional functioning appeared adequate, there were suggestions of defensiveness and worry. Global adaptive functioning was below average for most children and several parents responded with concern about their child's social functioning.

The purpose of this poster is to present a longitudinal case study to illustrate the long-term neuropsychological outcome in an adolescent with multiple repaired AVMs and to stress the importance for continued long-term neuropsychological follow-up, beyond medical clearance.

CASE REPORT

This is a young male who underwent neuropsychological assessment as an outpatient at three data points following initial diagnosis of a right mesial frontal AVM and a subsequent right tentorial dural AV fistula. The following timeline outlines the history of medical events and neuropsychological assessments.

March 1999	First AVM was diagnosed (right frontal superior gyrus extending to the genu of the corpus callosum pressing into the right lateral ventricle); see Figures 1, 2, 3, and 4)
April 1999	Embolization with subsequent complete surgical resection of AVM
August 1999	Neuropsychological Assessment (11 years, 5 months)
February 2003	Neuropsychological Assessment (14 years, 11 months)
Summer 2006	Patient sustained back injury in a dive from a bridge; this prompted a repeat angiogram
July 2006	Right tentorial dural arteriovenous fistula was diagnosed; see Figure 5.
August 2006	Surgical repair of dural arteriovenous fistulous malformation
December 2006	Neuropsychological Assessment (18 years, 8 months)

Developmental history was unremarkable until age 11 years, when he had a febrile seizure. An EEG study was completed and the results were abnormal. An MRI of the brain revealed a 4 cm arteriovenous malformation in the right frontal area, which was embolized and surgically resected by craniotomy in 1999. Following surgery, he participated in comprehensive therapies at a day rehabilitation program. A routine angiogram for follow-up of the previously resected AVM after he had hurt his back in a dive from a bridge was completed in 2006 prior to his attending college. Results revealed a dural right frontotemporal arteriovenous fistula. He underwent a craniotomy in August of 2006 when the lesion proved not amenable to embolization. One generalized tonic-clonic seizure occurred after surgery. He subsequently was prescribed Lamictal for seizure prevention and Concerta to decrease impulsivity. He also began experiencing double vision in his right eye and muscle weakness above his right eye. He has received ongoing psychotherapy since his first AVM surgery in 1999 due to anger management issues and impulsivity, both of which were present before detection of his AVM. He is currently enrolled at the University of Alabama. Although he has not been able to attend classes due to his surgery, he has been living on campus and is taking two on-line classes. His combined SAT scores were 940.

Figure 1. Dorsal view



Figure 2. Posterior view

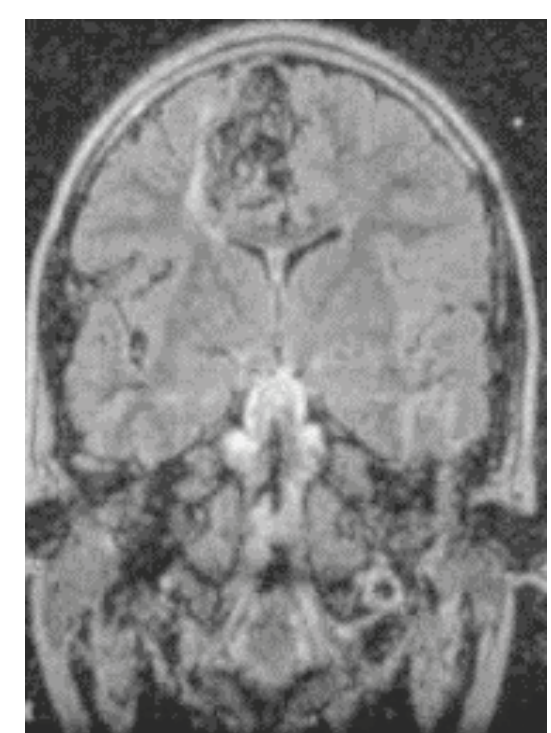


Figure 3. Lateral view

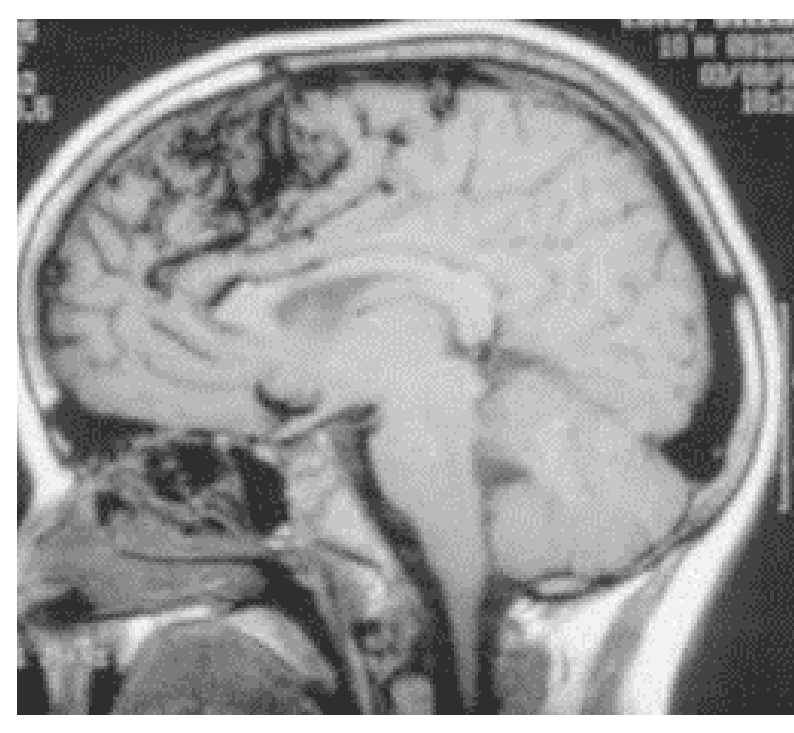


Figure 4. Angiogram (1st AVM)

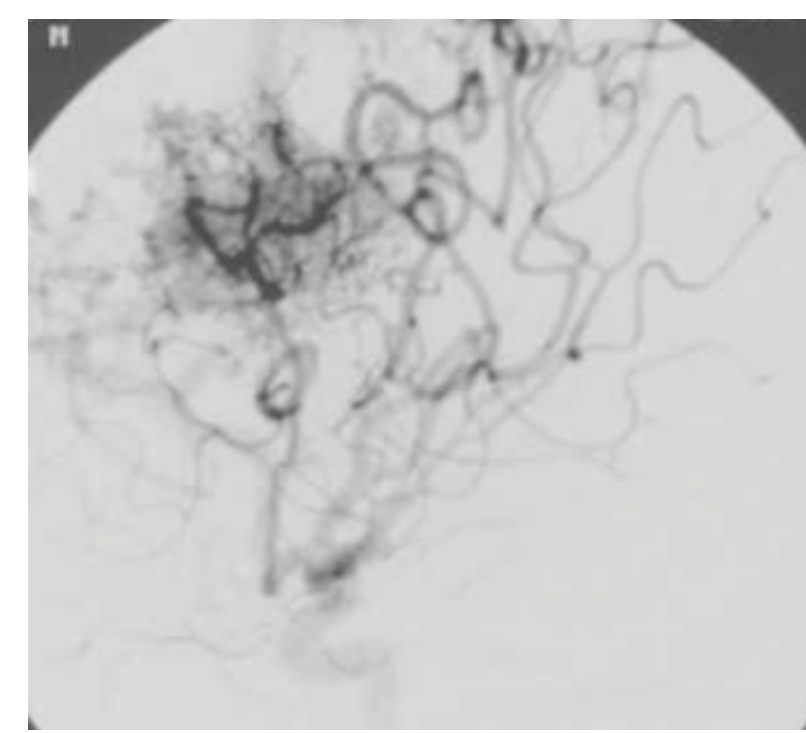
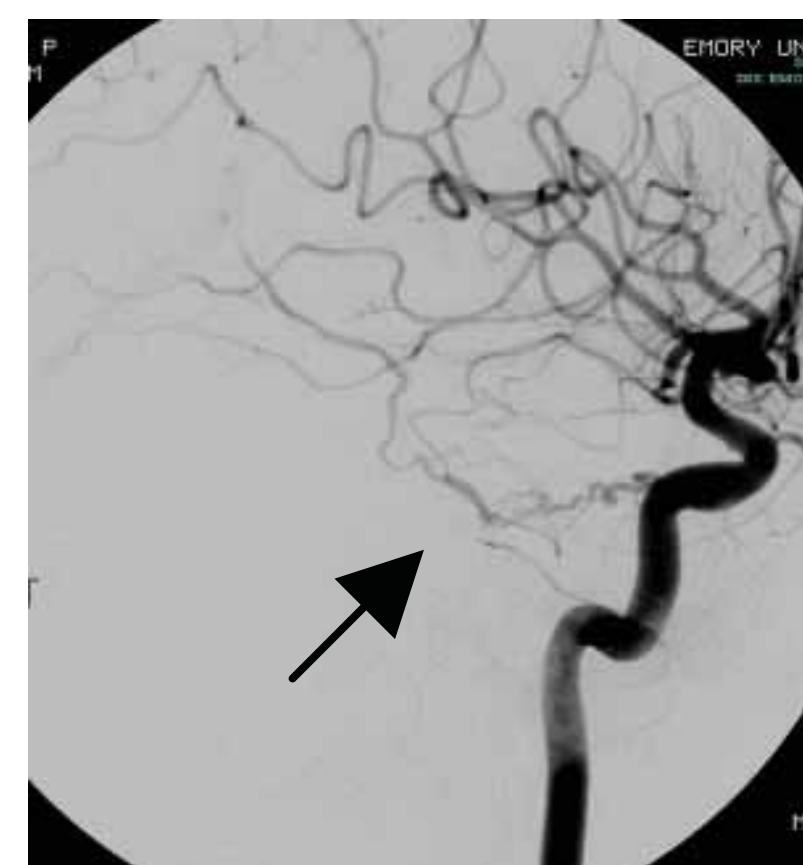


Figure 5. Angiogram (2nd AVM)



METHODS

For each neuropsychological assessment, relevant information was obtained through a review of the patient's medical record and interview with his mother, various questionnaires completed by his mother and teacher, observations of the patient's behavioral repertoire, and his performance on a variety of psychological tests. The domains assessed at each assessment included intellectual functioning, language, motor/visual perceptual/spatial functioning, memory, executive functioning, attention, achievement, and social/emotional/behavioral functioning.

RESULTS AND DISCUSSION

Neuropsychological Assessment I:

Overall cognitive ability was variable with significantly stronger verbal than nonverbal abilities. This may be associated with abnormal brain development due to the arteriovenous malformation. Within this context, the neuropsychological profile revealed deficits in listening comprehension, analysis/synthesis of meaningful information, organization and integration of nonverbal output in the left visual field, and nonverbal memory when he had to rely on visual processing to encode and retrieve visual stimuli. Behavioral observations revealed an impulsive response style and organizational difficulties. In addition, mild weaknesses were noted in word retrieval. In contrast, single-word receptive vocabulary, expressive language skills on unstructured tasks, certain nonverbal skills (e.g., visual scanning, spatial orientation), fine motor speed and dexterity, and nonverbal memory for verbally encodable sequences of information, verbal learning and memory, and certain executive skills (e.g., cognitive set-switching, flexible problem-solving) were secure. Academic skills were mostly average to high average with a relative weakness in written expression skills. On this latter task, the patient omitted some words and had difficulty with organization of ideas and output on paper. Emotionally, his mother endorsed somatic complaints. He did not endorse significant symptoms of anxiety and depression, although he expressed concern about controlling his anger outbursts and swearing. The examinee was benefiting from counseling to explore grief issues regarding his AVM, to learn to control his temper, and to move on past this crisis.

Neuropsychological Assessment II:

The results of this neuropsychological evaluation suggested that the examinee was slightly struggling cognitively. Overall cognitive ability was low average to average. In the area in which the AVM was found, there were slight and subtle adverse sequelae. For example, his ability to construct and organize complex spatial information in paper and pencil format was a relative weakness. In contrast, when he was asked to perform nonverbal spatial tasks in the absence of a motor component, his performance was better developed. This suggested that motor interference was a culprit in any spatial difficulties that he was having. Analysis of his manual motor dexterities, again frontal functions, showed speed that was decreased for his dominant hand, but not for his non-dominant hand. Manual motor dexterity was within normal limits for his dominant hand and low average for his non-dominant hand. It was likely that this variability within his manual motor functioning contributed to minor difficulties he experienced in his construction and production of spatial information.

Language skills spanned the average to high average range and suggested a relative strength. He was able to learn rote information, although there was minor difficulty with monitoring of performance. His ability to recall contextual verbal information was average, although he is significantly aided by recognition cues, suggesting slight verbal retrieval interference. Visual scanning and attentional skills improved significantly since the previous assessment. Academic skills were within the average to high average range in the area of reading and in mathematics. Spelling skills remain a solidly high average strength.

Emotionally, he was not reporting any significant symptoms of a mood or anxiety disorder at this point in time. His mother and teacher were not reporting any concerns regarding his emotional behavior, although his mother reported that there were attentional/executive issues which interfered with his home functioning that were not seen in school.

Neuropsychological Assessment III:

The results of this neuropsychological evaluation suggest that the examinee made great gains since the previous neuropsychological evaluation in 2003 when he showed localized deficits in keeping with his right frontal location of AVM and subsequent surgery. He now demonstrates verbal intellectual functioning that is within the high average range, which is a better representation of his intellectual potential than his nonverbal intellectual functioning, which still shows some residual difficulty in the ability to organize spatial information, either when a motor component is present or not present. Within the area of language functioning, language skills are generally secure and he shows great gains in his ability to understand complex linguistic concepts.

Within the motor/sensory/perceptual domain, his performance is variable, again in keeping with the location of his AVM and subsequent surgeries. His manual motor dexterity and fluency is within normal limits bilaterally for his dominant and non-dominant hands, although he does not demonstrate the dominant advantage. His perceptual organization of spatial stimuli, as evidenced by his ability to copy complex visual information is mildly impaired. When a motor component is deleted from spatial tasks, he struggles slightly if he is required to be the "executive" of a spatial task, such as re-visualizing single details. However, when information is spatial and linear, and he only is required to recognize a response, his performance is high average.

Within the area of learning and memory, he shows an excellent pathway through the auditory/verbal modality, especially when information is contextual. In this situation, his performance is high average to superior. His performance in the verbal domain when information is rote is more variable and suggestive of mild interference with attaining an attentional set that is rectified by trial learning. His ability to learn such rote verbal information is adequately developed over time. His visual memory for complex spatial stimuli shows a "consolidation effect," or that he needs more time than usual to encode information into long-term storage.

With regard to attention and executive functioning, he can perform rote tasks of attention with adequate ability, while he struggles slightly with working memory, especially in the juggling of auditory-rote information. His visual attention at this point in time is compromised and slowed by double vision, and is low average. On executive tasks, he benefits from feedback, rules, and demonstrates an impulsive tendency that can be rectified by feedback.

Academically, his performance is within the high average to superior range across all areas of academic functioning, more in keeping with his verbal intellectual presentation.

Emotionally, he appears to be resilient, is optimistic about his future, and is not reporting significant symptoms of a mood disorder. He is handling his current recuperation from surgery with realism and patience. He reports typical and developmentally appropriate wishes, as well as a nurturing background, which suggests that he appreciates his family. His parents are not reporting significant internalizing or externalizing behavioral issues. They are reporting that his attention is adequately developed on his current regimen of Concerta.

In summary, post-surgical neuropsychological data suggest localized deficits, including difficulties with planning, organization, emotional control, pragmatic language, and nonverbal encoding/retrieval following repair of the right frontal AVM and double vision and difficulties with organization of nonverbal material following repair of the right tentorial dural AVM. Long-term data indicate resolution of most localized deficits. Parent report collected through diagnostic interview revealed residual ongoing, but intermittent executive difficulties, particularly those associated with the orbitofrontal area (e.g., impulsivity, poor decision-making).

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