

Life without the dominant hemisphere - a holohemispheric arteriovenous malformation in an adult

Aki Laakso, MD, PhD¹, Jussi Numminen, MD, PhD², Johan Marjamaa, MD, PhD¹, Reza Dashti, MD, PhD³ and Juha Hernesniemi, MD, PhD¹

Departments of ¹Neurosurgery and ²Radiology, Helsinki University Central Hospital; ³Department of Neurosurgery, Cerrahpasa Medical faculty, University of Istanbul

Introduction

A 30-year old woman was recently admitted to consultation because of a large, unruptured AVM. The lesion was originally diagnosed at the age of 9 (in 1990) due to mild right-sided hemiparesis and hemianopy and focal epileptic seizures. Her parents were told at the time that the lesion was inoperable. She has no apparent cognitive symptoms, and her academic success has been normal, although she has experienced some mild dysphatic symptoms. She has a college degree in economy, and is currently studying in college for a degree in nursing. She was now referred to us by her neurologist, with a question if something could be done to the AVM with current treatment modalities. Her neurological symptoms have remained stable for years, and she receives medical treatment for her epilepsy.

Figure 1. A gadolinium-enhanced T1-weighted MRI in axial, coronal and sagittal planes demonstrate a giant AVM encompassing the whole left hemisphere, with some cortical sparing in frontobasal and temporobasal regions..

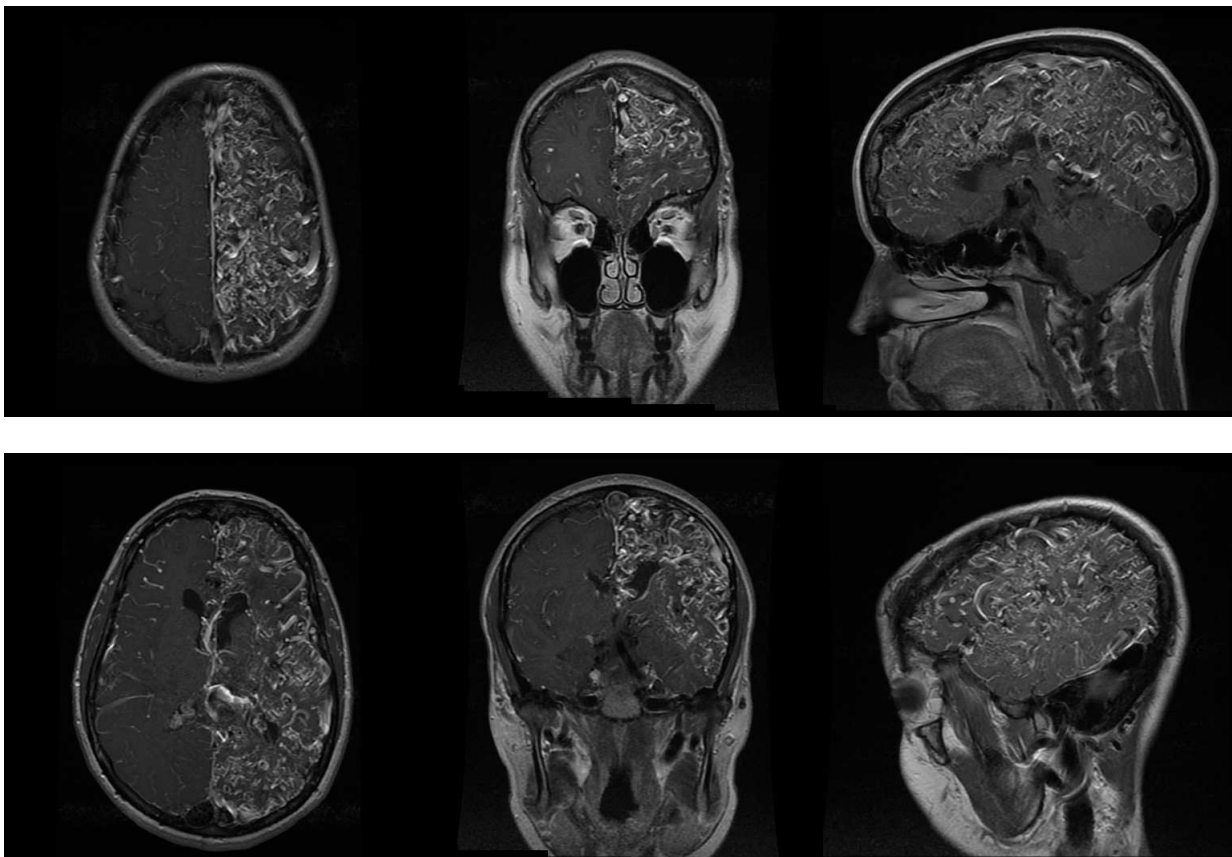
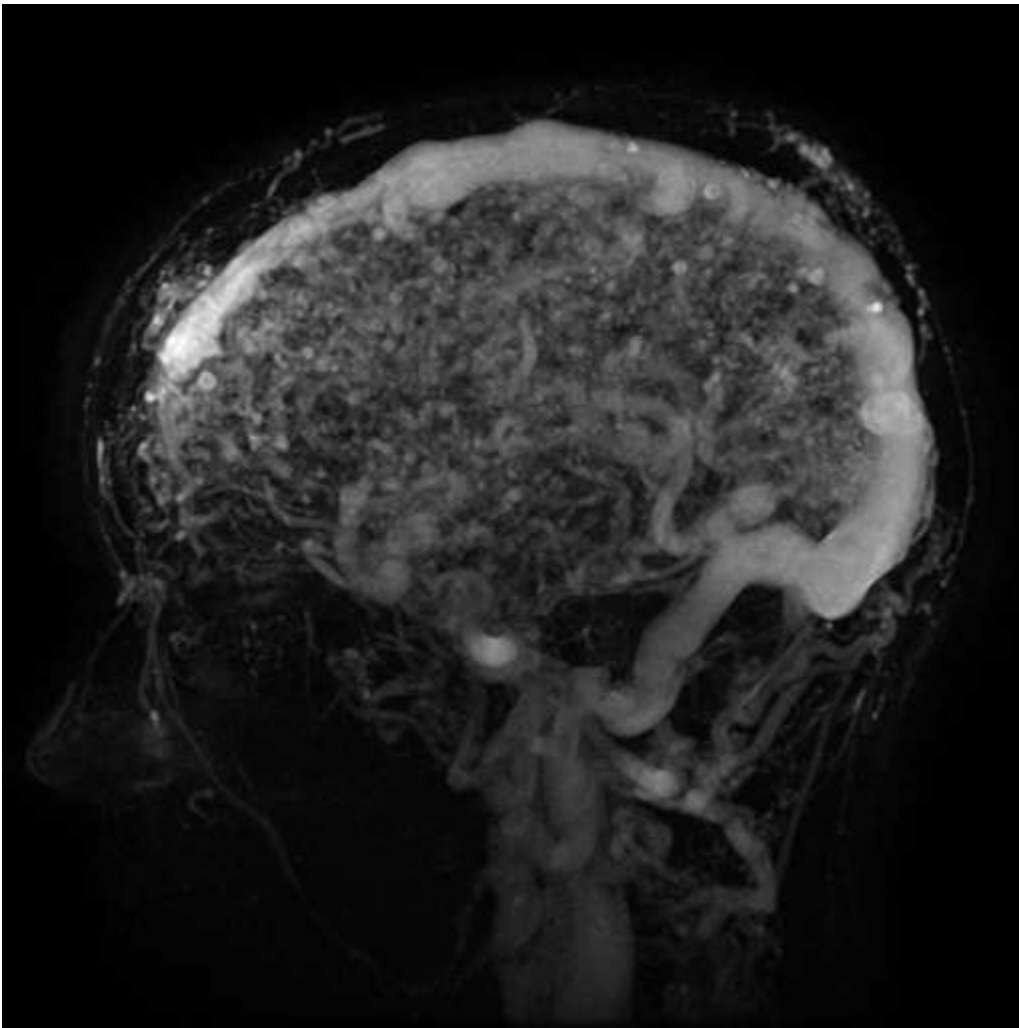


Figure 2. MRI TRICKS sequence of the same AVM.



Questions & Answers

1. Is there any potential treatment indication for this AVM?

The lesion itself is obviously inoperable. However, presence of significant flow-related saccular aneurysms in feeding arteries might indicate prophylactic treatment for them in a young patient. Therefore we felt that DSA examination is justified.

Palliative partial treatment with embolization is not advisable in such a lesion, because it is unclear whether it would relieve any of the neurologic symptoms, and may increase the risk of rupture due to change in hemodynamics. Radiosurgery is also useless in the treatment of a lesion of this size.

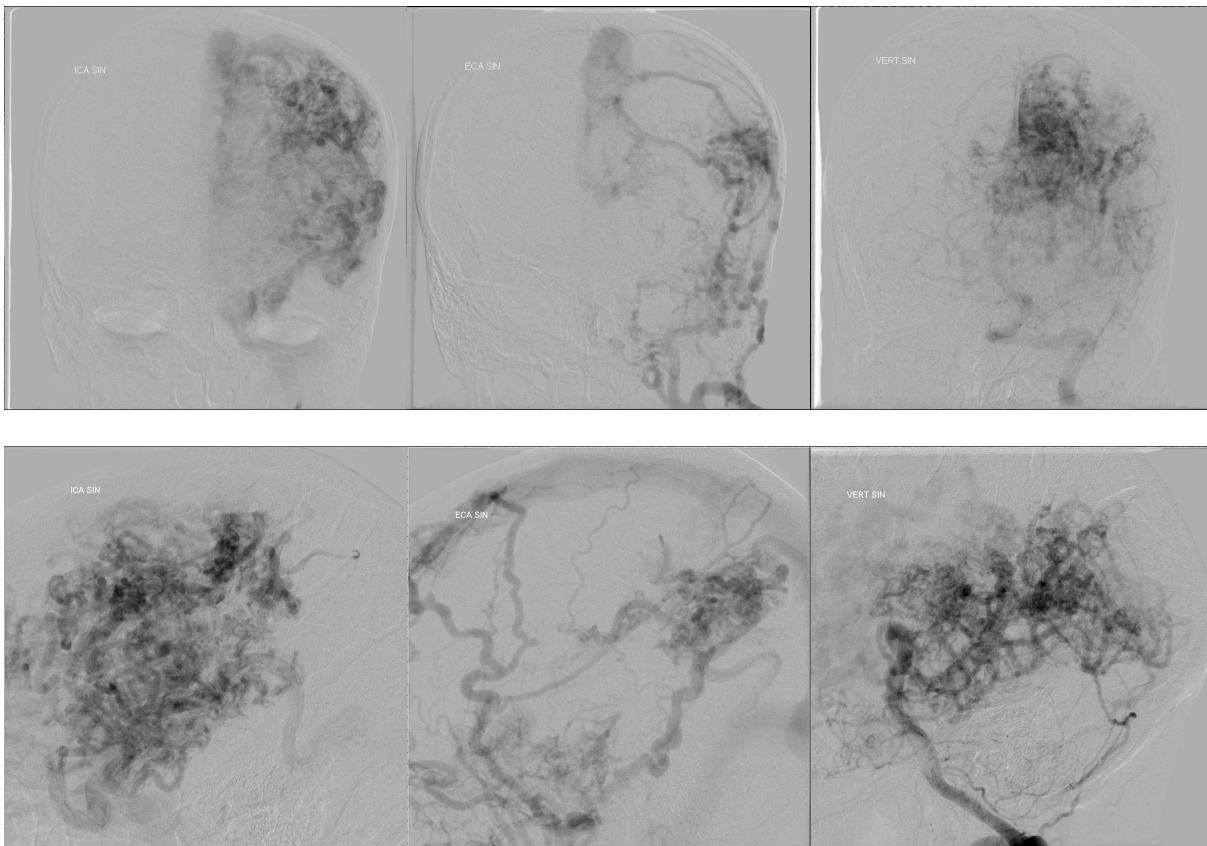
2. What is TRICKS sequence?

TRICKS stands for Time Resolved Imaging of Contrast Kinetics. It is a form of MR angiography that has very high temporal resolution. In addition to AVM imaging, it can be very useful to detect dural and spinal arteriovenous fistulas.

3. In case you feel that DSA examination was appropriate, what vessels should be imaged?

DSA investigation of a large AVM should always be a “six-vessel” angiography, e.g. include ICA, ECA and VA bilaterally, at least if any treatment is planned. Large AVMs are usually multicompartamental, and parts of the nidus are easily missed unless all potential feeders are imaged.

Figure 3. AP and lateral projections of left ICA, left ECA and left VA DSA. Due to extremely high flow in the AVM, it is very difficult to discern angioarchitectural details, but its multicompartamental nature is apparent. The AVM was fed by both ECAs, both ACAs, left MCA and left PCA. No flow-related aneurysms were seen. Right-sided angiographies are not shown here.



Questions & Answers:

4. Giant AVMs, AV fistulas and venous malformations often present in neonatal age with a certain non-neurological symptom. What is that?

Arteriovenous shunting in a massive intracranial vascular malformation or fistula often leads to congestive heart failure in an infant. The treatment is occlusion of the shunting, if possible. However, in this case the patient did not have a history of cardiac problems, which may suggest that her AVM has reached its enormous size after the perinatal period.

Discussion

Truly holohemispheric pial AVMs are exceedingly rare. This patient is the second patient admitted to our department with holohemispheric AVM since 1945, among appr. 800 AVM patients in our database. The angiography images of the 1945 case have not survived to this day, and it is not clear whether it would be classified as holohemispheric using today's imaging methodology.

The patient's symptoms were surprisingly mild compared with the radiological appearance. It is likely that some, but not all, functions of the left hemisphere had been transferred to the right hemisphere during development. Functional MRI is not likely to shed more light on the issue, because extreme flow in the AVM will disrupt the BOLD signal. We have discussed with the patient about the possibility to map some of the functions using MEG, although it is likely to have more academic than clinical interest.

While there is no sensible treatment to be offered to the patient at present (except for medical treatment for her epilepsy), her condition has luckily remained stable for a long time, and she is able to lead a fulfilling life despite her symptoms. The question of treatment may become a real issue in the unfortunate situation that her AVM bleeds. Even if we were able to demonstrate with MEG or other methodology that most or all of the remaining functions originally located in the left hemisphere had been transferred to the right during development, the occlusion of an AVM of this size has potentially catastrophic consequences. The ability of autoregulation of cerebral blood flow in the healthy hemisphere to adapt to the occlusion of the shunting may well be inadequate, even if the lesion itself could be successfully occluded.

The exact etiology of her symptoms is also interesting. Is it because of chronic ischemia due to vascular steal, mass effect, developmental impairment of the left hemisphere, or all of the above? If the symptoms are mostly due to vascular steal, it is intriguing to contemplate whether either palliative or staged treatment would be helpful, especially if the patient would continuously deteriorate due to progressive symptoms. However, partial embolization has actually increased the rupture risk of AVMs, according to experience of us and many other teams. Staged resections have also been proposed by some authors for giant AVMs, but these probably carry even higher risks than staged embolizations. Staged or fractionated radiosurgery could theoretically be suggested as options, but the volume of this lesion is vastly larger than anything treated successfully with these modalities according to the literature. The cumulative radiation dose would also very likely become a problem.

In the future, biological treatments targeting the probably active angiogenic and inflammatory processes in giant AVMs may become the treatment of choice for these lesions, the treatment of which at the moment is extremely challenging - or outright impossible.