

Surgery for a giant arteriovenous malformation without motor deterioration: preoperative transcranial magnetic stimulation in a non-cooperative patient

Case report

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Transcranial magnetic stimulation (TMS) is a noninvasive activation method that is increasingly used for motor mapping. Preoperative functional mapping in vascular surgery is not routinely performed; however, in cases of high-grade arteriovenous malformations (AVMs), it could play a role in preoperative decision making. A 16-year-old male was suffering from a giant, right-sided insular, Spetzler-Martin Grade V AVM. This patient's history included 3 hemorrhagic strokes in the past 3 years, resulting in Medical Research Council Grade 2–3 (proximal) and 2–4 (distal) paresis of the left side of the body and hydrocephalus requiring a ventriculoperitoneal shunt. Preoperative TMS showed absent contralateral innervation of the remaining left-sided motor functions. Subsequently, the AVM was completely resected without any postoperative increase of the left-sided paresis. This case shows that TMS can support decision making in AVM treatment by mapping motor functions.

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KEY WORDS • arteriovenous malformation • brain mapping • eloquent area • microsurgical resection • outcome • pediatric hemorrhagic stroke • vascular disorders

ARTERIOVENOUS malformations (AVMs) are a major cause of pediatric hemorrhagic stroke (HS), accounting for almost half of the strokes in children, with an average annual incidence rate of 0.5 case per 100,000 person-years.^{9,17,29} The etiology of cerebral AVMs remains unclear, but genetic factors have been suggested to play an important role, which is supported by associations with other diseases such as Wyburn-Mason syndrome.¹ Clinical features of this syndrome are AVMs involving the retina, brain, and skin of the face and result from an error in the development of vessels at the 7th week of gestation.²⁵ Patients present with seizures, hemorrhages, or headache.¹⁶ The annual risk of AVM hemorrhage in pediatric patients is estimated to be

2%–4% per year and is even higher in patients with previously ruptured AVMs.^{7,24,26,29} Case fatality rates vary from 5.2% to 54%, with a reported average mortality of 25% in children with HS.^{3,9,11,24}

Treatment options for intracerebral AVMs are microsurgical removal, endovascular embolization, stereotactic radiosurgery, or a combination of these techniques.^{1,29} Large high-grade AVMs are often located in or near eloquent areas, such as the motor cortex. Surgery can be destructive as a result of direct damage to the brain, especially during severe intraoperative hemorrhage, or as a result of ischemia caused by obstruction of arteries, which have en passant feeders combined with a large or essential cerebral perfusion territory.^{7,24,29}

When considering neurosurgical intervention, preoperative motor mapping can be performed.²⁸ A noninvasive instrument that is increasingly used in tumor surgery is transcranial magnetic stimulation (TMS). It can stimulate the cortex by generating a magnetic field passing through the scalp^{21–23,28} and may be used to map the motor cortex by activating the cortex with single pulses and to simultaneously record motor evoked potentials.^{18,28}

Transcranial magnetic stimulation has the advan-

Abbreviations used in this paper: AChA = anterior choroidal artery; AVM = arteriovenous malformation; EVD = external ventricular drain; fMRI = functional MRI; GCS = Glasgow Coma Scale; HS = hemorrhagic stroke; ICA = internal carotid artery; MCA = middle cerebral artery; MEG = magnetoencephalography; MRA = MR angiography; MRC = Medical Research Council; PCA = posterior cerebral artery; TMS = transcranial magnetic stimulation; VP = ventriculoperitoneal.

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tage that patients do not have to lie still as in functional MRI (fMRI) or magnetoencephalography (MEG). We describe a 16-year-old patient with a giant, right-sided, insular AVM in whom we performed preoperative TMS.

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History and Examination. In 2009, a 12-year-old patient presented to our institution with severe headache, vomiting, lethargy, and progressive left-sided paresis (Medical Research Council [MRC] Grade 4).⁴ Since early childhood, our patient had suffered from coordination and fine motor skills disorders of his left arm without evident signs of (hemi)paresis. General examination showed redness of the forehead, nose, and right cheek. Papilledema and a retinal vascular malformation of the right eye were documented during eye examination; neurological examination showed a lethargic child with a slightly dilated right pupil and MRC Grade 4 left-sided paresis. Magnetic resonance imaging demonstrated a space-occupying lesion in the right hemisphere without signs of stroke (Fig. 1A); MR angiography (MRA) and digital subtraction angiography revealed a giant Spetzler-Martin Grade V AVM in the insula with the anterior choroidal artery (AChA) and

lenticulostriate arteries as the major feeding arteries with an additional contribution of the posterior cerebral artery (PCA; Fig. 1B–D). Combining these findings, we diagnosed Wyburn-Mason syndrome.^{16,25} We faced a difficult decision in the multidisciplinary consultation. Treatment with embolization carried a great risk of complications, and complete cure could not be achieved.

At that time, in our opinion, the only option was coiling the associated aneurysm. This procedure was performed and resulted in partial stenosis of the AChA. Stereotactic radiosurgery was not indicated because of the size of the AVM. As surgery could result in a significant chance of complete hemiplegia, we adopted a wait-and-see policy. At discharge, the left-sided paresis recovered to an MRC Grade 5.

In the subsequent 3 years, our patient suffered from 2 major intracerebral hemorrhages with breakthrough into the ventricular system, resulting in a poor but slowly recovering neurological state; a temporary external ventricular drain (EVD) was needed at both admissions. At the first HS, he presented with the acute onset of paralysis of his left arm and MRC Grade 4 paresis of his left leg, which recovered to Grade 5 left-sided paresis (identical to his neurological status before the first HS). At the second HS, he presented with Grade 2 acute paresis of his left

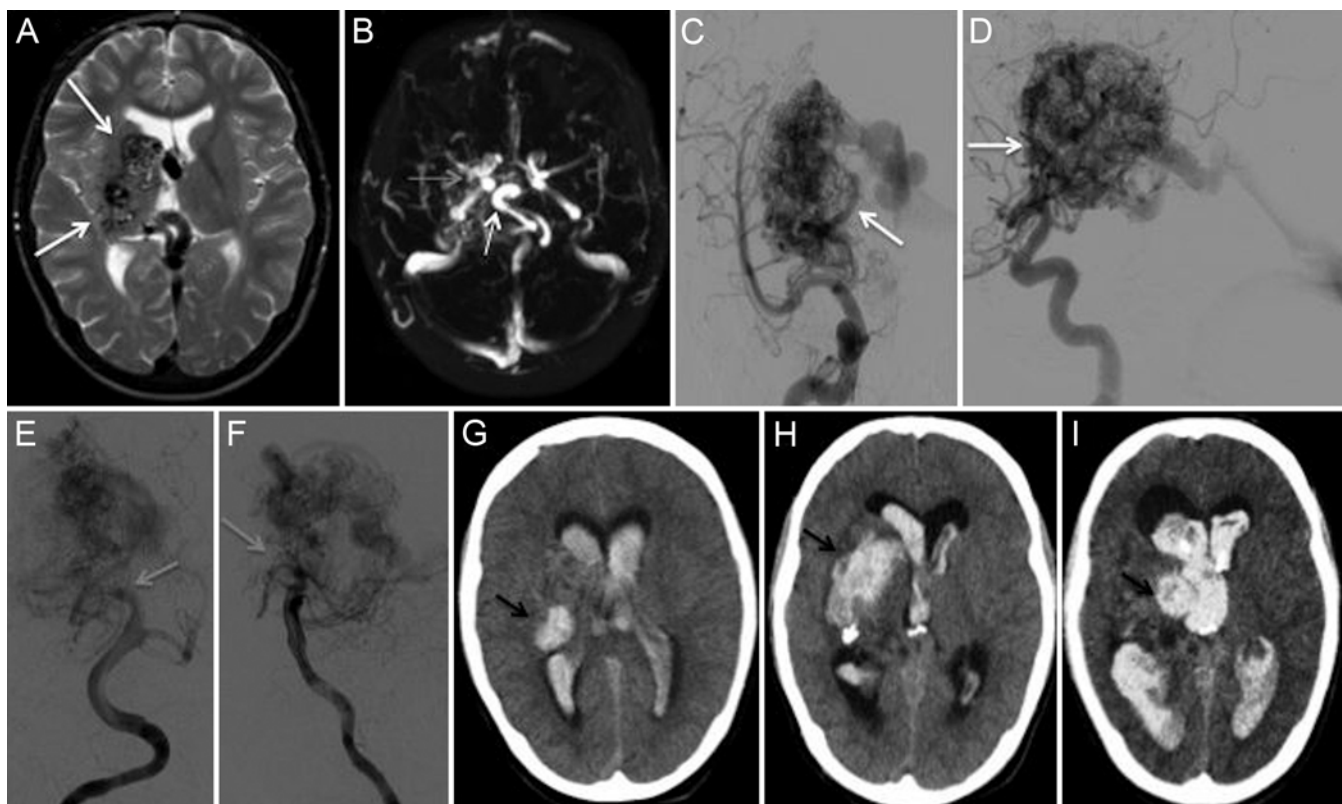


Fig. 1. Axial T2-weighted MR image (A) showing a space-occupying lesion in the right hemisphere suspected to be an AVM (arrows). Axial MRA reconstruction (B) confirmed a giant AVM, with the MCA (gray arrow) and PCA (white arrow) as major feeding arteries. Anteroposterior and lateral angiograms of the right ICA (C and D) and right vertebral artery (E and F) illustrating the complex architecture, with the AChA (white arrows) and PCA (gray arrows) as feeding arteries of the AVM. Axial CT scans demonstrate the first (G), second (H), and third (I) HSs from the AVM; all hemorrhages involved the ventricular system and caused hydrocephalus. Hemorrhage originating from the posterior site (black arrow) of the AVM (G), bleeding from the frontolateral part (black arrow) of the AVM (H), and bleeding from the frontomedial site (black arrow) of the AVM in the basal ganglia (I), with bilateral EVDs inserted.

arm and Grade 3 paresis of his left leg, which recovered to Grade 4 left-sided paresis at discharge.

At the age of 16, he suffered from a third HS. He presented with a Glasgow Coma Scale (GCS) score of 3 and fixed dilated pupils, and bilateral EVDs were emergently inserted. Finally, a ventriculoperitoneal (VP) shunt was placed for shunt-dependent hydrocephalus (Fig. 1G–I). He recovered from his coma slowly but considerably well. Thirty days after the hemorrhage, he recovered to a GCS score of E4M6V4, and neurological examination showed a lethargic patient with left-sided paresis (left arm: Grade 4 proximally and Grade 2–3 distally; left leg: Grade 4 proximally and Grade 2 distally) and hemianopia.

At this phase, we were concerned about a large AVM located in a part of the brain—a presumed eloquent area—that had bled repeatedly. Untreated, the AVM could rebleed with a high chance of death. Complete embolization was not feasible because of the complex architecture and size of the AVM (Fig. 1E and F). Preoperative embolization was extensively discussed with the intervention radiologists, but the procedure was not performed for the following reasons: 1) access to the AVM was very challenging, 2) rupture risks were expected to be high, 3) partial embolization of this type of AVM could result in difficult intraoperative mobilization and resection, and 4) the AVM had bled at 3 different sites. We concluded that the disadvantages of embolization were greater than the theoretical benefits. Stereotactic radiosurgery was not indicated for this large AVM. Therefore, we finally decided that surgery was the only option. We assumed that surgery would mean a significant chance of complete hemiparesis; therefore, we used a noninvasive tool to determine to what extent we could expect postoperative motor deficits. We used a MagPro Compact TMS round coil (Medtronic) generating biphasic single-pulsed stimuli (protocol described elsewhere¹⁵). Motor responses were recorded with electromyography electrodes from the right and left thenar muscles. Transcranial magnetic stimulation over the left central sulcus produced an intact contralateral motor response, but the contralateral response was absent during stimulation over the right central sulcus. We assumed that the remaining left-sided motor function was innervated ipsilaterally as a consequence of brain plasticity, because of the AVM and repeated right-sided HSs.

Operation. The AVM had 3 main feeding sites: 1) frontal/basal: lenticulostriate arteries of the middle cerebral artery (MCA) and the AChA (Fig. 1C and D), 2) posterior/basal: PCA/posterior communicating artery (Fig. 1E and F), and 3) posterior/cranial: posterior choroidal branches. Our surgical approach consisted of a right-sided pterional approach combined with extension toward a subfrontal and pretemporal route. We achieved this by first meticulously occluding the lenticulostriate feeders from M₁, which led to a significant reduction in flow through the AVM and consequently its size, allowing access to the tip of the basilar artery and the proximal right PCA, seen through the triangle formed by oculomotor nerve and internal carotid artery (ICA). Subsequently, feeders arising from the basilar tip and the proximal PCA were coagulated. The AVM was then exposed in the medial wall of the right ventricle through a frontal corticoto-

my, which allowed occlusion of the anterior and posterior choroidal feeders and complete resection. No complications occurred during surgery. The total duration of the procedure was 11 hours.

Postoperative Course. Our patient recovered well from surgery with no additional deficits except for complete right-sided oculomotor paresis. Revision of the VP shunt was required 1 week postoperatively because of blood clots in the pump. The patient was discharged to a rehabilitation center 2 weeks after surgery. Computed tomography angiography documented complete resection of the AVM (Fig. 2). At the 6-week follow-up, neurological examination revealed stable residual hemiparesis as had appeared preoperatively. The oculomotor nerve paresis improved, with a lateral position of the right eye and disappearance of the vertical gaze paresis.

Discussion

We described a 16-year-old patient with a giant, right-sided, insular AVM. During the preoperative work-up, cortical motor function was mapped using TMS and showed absent contralateral innervation of the remaining left-sided motor functions. A longer wait-and-see policy was not an option, and aggressive treatment seemed justified because of the size of the AVM and the apparent high chance of hemorrhage.

Because most AVMs are congenital, the developing brain is able to take over behavioral functions for the affected hemisphere, resulting in ipsilateral innervation, especially in children with the plasticity of the maturing CNS in which reorganization of motor control occurs, in particular in the context of early developed lesions.^{5,15,24} Preoperative counseling is beneficial in managing patient expectations and influences neurosurgical decision making. Transcranial magnetic stimulation revealed that our patient's motor function was not served by the contralateral hemisphere. Although TMS did not fully demonstrate ipsilateral innervation, the postoperative course fits the assumption that there was ipsilateral innervation.

Brain mapping in children is challenging because of the reduced level of cooperation, which is required in procedures like fMRI. Multiple types of functional brain mapping have been reported (mostly applied in tumor surgery), and several advantages specific to each mapping

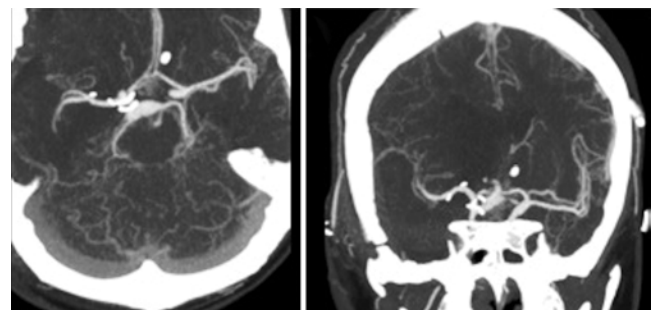


Fig. 2. Postoperative axial (left) and coronal (right) reconstruction CT angiograms showing complete resection of the AVM in the right hemisphere.

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type have been described.^{2,5,6,8,12,13,21–23,27,28} Regardless of the suggested benefits of other mapping methods, TMS offers the advantage of being the only noninvasive, reversible inhibition technique and is more commonly used in the neurosurgical field for motor mapping in patients incapable of fully cooperating.^{13,28} Transcranial magnetic stimulation results on lateralization of motor innervation were confirmed using fMRI in a recent study.¹⁵ An important limitation of fMRI is its inability to show the complete area involved in motor performance.¹³ Mapping using this method can be inaccurate, defining functionally active brain as inactive as a result of the altered hemodynamics surrounding an AVM.¹³ Furthermore, TMS recordings have been shown to be more accurate than fMRI in preoperative mapping, providing information on cortical neurons without being influenced by cerebral hemodynamics.^{10,13,28} Our patient was insufficiently cooperative and could not lie completely still for successful fMRI and MEG, and TMS seemed the optimal and most reliable mapping method, because it does not rely on a patient's full participation nor does it require strict immobility. Clear estimations of the positive and negative predictive value of TMS in neurosurgical motor lateralization are lacking. In recent studies, preoperative navigated TMS results correlated well with direct cortical stimulation data in identifying the primary motor cortex; the negative predictive value of navigated TMS, as compared with direct cortical stimulation, in preoperative language mapping was 83.9%.^{19,20} To definitely establish the accuracy of TMS in preoperative mapping, prospective trials are needed to relate the results of TMS with intraoperative or postoperative findings.¹⁴

The age of and the hemiparesis and hemodynamic problem in our patient made for difficult decisions in which TMS helped as a feasible and robust mapping method. This technique enabled us to manage patient expectations and perform a microsurgical removal that resulted in a satisfying outcome.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Han, Kronenburg, van Doormaal, van Eijnsden. Acquisition of data: Kronenburg. Analysis and interpretation of data: Han, Kronenburg, van Doormaal, van Eijnsden, Leijten. Drafting the article: Kronenburg, van Doormaal, van Eijnsden. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Han.

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