Giant arteriovenous malformation

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Spetzler-Martin AVM grading system IV and V grade or Spetzler Ponce classification type-C arteriovenous malformation (AVM)

Management

The management of large and giant arteriovenous malformations (AVMs) in patients presenting with nonhemorrhagic neurological deficits secondary to vascular steal phenomenon is challenging and controversial. In many cases, large AVMs cannot be completely excised or cured, leaving patients with residual or partially treated AVMs, the natural history of which is unknown. Additionally, large, diffuse vascular malformations with multiple, small feeders, slow flow, or so-called cerebral proliferative angiopathy represent a related but distinct clinical and angiographic entity that may require a different therapeutic approach than traditional brain AVMs.

The American Heart Association Scientific Statement and Japanese Guidelines [1] on the management of intracranial arteriovenous malformation AVMs recommend conservative follow up for Spetzler-Martin AVM grading system IV and V grade unless accompanied by massive hematoma, associated aneurysm, or progressive neurological deterioration [2] [3].

Han et al. recommend that no treatment be given for most Spetzler-Martin AVM grading system IV and V. No evidence indicates that partial treatment of an AVM reduces a patient's risk of hemorrhage. In fact, partial treatment may worsen the natural history of an AVM. The authors do not support palliative treatment of AVMs, except in the specific circumstances of arterial or intranidal aneurysms or progressive neurological deficits related to vascular steal. Complete treatment is warranted for patients with progressive neurological deficits caused by hemorrhage of the AVM. This selection process plays a significant role in the relatively low combined morbidity and mortality rates for Grade IV and Grade V AVMs (17 and 22%, respectively) reported by the cerebrovascular group in both retrospective and prospective studies [4].

For giant cerebral AVMs located superficially or not involving critical components, a good outcome can be expected through surgical resection. The obliteration and recurrence rates were satisfying, and the complication rate was acceptable [5].

A large series of 500 intracranial AVMs included 34 occipital AVMs (6.8%), three of which were giant. Surgery in 30 cases including three giant AVMs resulted in favorable outcomes without morbidity or mortality, suggesting that the outcome was independent of size, probably due to the characteristic features of occipital AVMs. The occipital lobe is an eloquent part of the brain, particularly for the optic system. However, optic function has often already been lost in patients with giant occipital AVMs

Giant occipital AVMs are good candidates for microsurgery despite their size [6] [7]

The current management of children with other conditions of chronic cerebral hypoperfusion, such as moyamoya disease, involves consideration of surgical revascularization to enhance blood flow to the compromised hemisphere.

Case report

2014

A 58-year-old man presented with bleeding ulcer of the left arm caused by a high-flow Spetzler Ponce classification type-C arteriovenous malformation (AVM), feed by branches from both the subclavian arteries. He had been previously treated with AVM sclerotherapy, embolization, humeral artery endografting, and open surgery. We urgently performed coil embolization of the left vertebral artery, and we covered the huge subclavian artery by a thoracic endograft. Then, we embolized the right tyrocervical trunk. The result was an immediate interruption of bleeding. At 12 months, the patient had no neurologic complications, and the upper limb continued to decompress [8)].

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 [9)].

2011

Ellis et al. present the case of a young child with a large thalamic vascular malformation who presented with clinical and radiological features of vascular steal and ischemia. In an effort to augment flow to the hypoperfused brain and protect against future ischemia, the authors treated the child with unilateral pial synangiosis. At 12 months, postoperative angiography demonstrated robust neovascularization, and the child has not sustained any further ischemic events. The authors discuss concept of vascular malformation-related hypoperfusion and the utility of indirect revascularization for inoperable vascular malformations presenting with ischemic symptoms [10)].

2009

Alexia et al. report on a case of a 12-year-old girl that was admitted under our care complaining of headache for over a month which was accompanied by vomiting and diplopia over the last 10 days. On neurological examination a right upper limb tremor and cervical rigidity were noted. CT and MRI scan was performed and revealed a giant left frontotemporal lesion. The lesion was heterogenous with calcifications and hemosidirin, surrounded by brain swelling and causing midline shift. There was little enhancement after gadolinium administration. The last finding led us to consider the presence of a vascular abnormality as a possible diagnosis. The patient was operated upon via a left fronto-temporal craniotomy. We managed to excise the lesion totally. Histopathology revealed the presence of an AVM. Postoperatively the patient was neurologically intact but a subcutaneous collection of CSF was noted that was successfully treated by drainage. Although surgical treatment of deeply seated giant AVM's in the dominant hemisphere of speech and motor-sensory area have a relative high proportion of postoperative neurological deficit, careful surgical intervention can produce excellent outcome [11)].

2007

Cerebral AVMs associated with definite or probable moyamoya disease is a very rare situation, and the association between them is unclear. Chen et al. present a rare case of giant AVM-associated with unilateral moyamoya disease, and giant AVM makes planning any aggressive treatments difficult [12)]

1993

A 42-year-old woman suddenly developed headache and nausea on July 26, 1991, and the computed tomography (CT) scan showed a moderate-sized hematoma in the left occipital lobe. After one month's conservative treatment, she had recovered to a neurologically intact state. Cerebral angiography demonstrated a giant arteriovenous malformation fed by enlarged branches of the left posterior cerebral artery as well as small branches arising from the middle cerebral artery, anterior cerebral artery and the meningeal branches of the middle meningeal artery and the occipital artery. Preoperative embolization was planned on February 24, 1992. During an attempt at catheterization of the basilar artery and the left posterior cerebral artery with a balloon catheter and a Tracker-18 catheter, the patient complained of an intensification of her headache, nausea and vomiting. So the embolization procedure was stopped. The CT scan taken immediately at that time showed a severe subarachnoid hemorrhage (SAH). She became comatose about 40 minutes later. CT scan taken next day revealed also a complication of the pontine hemorrhage. Neurologically, she had gradually recovered and could communicate with some simple words 3 months after SAH. The total removal of the AVM was performed on May 26, 1992. Postoperative course was uneventful. She showed rapid and remarkable improvement in her neurological state suggesting that the blood flow in the surrounding brain area had been corrected. A blood deficit had no doubt been caused when blood had been stolen by the giant AVM [13)].

1971

[Total removal of a giant arteriovenous malformation of the vermis cerebelli extending into both cerebellar hemispheres] [14)].

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