Arteriovenous malformations of the medial cerebral hemisphere and the limbic system

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In a series of 164 resected arteriovenous malformations (AVM's), 25 were located on the medial aspect of the cerebral hemispheres. These AVM's require special attention because of the following characteristics: 1) their obscure location, making resection more difficult than the usual AVM; 2) a high incidence of intraventricular hemorrhage; 3) their frequent involvement of the choroid plexus of the ventricles and the arteries and veins related to the choroid plexus; and 4) the configuration of their venous drainage, which is often to superficial veins as well as to the deep venous systems. The results achieved with excision of these difficult AVM's were good. There was no mortality and the morbidity rate was under 4%. The surgical exposures, which were tailored to the location of these malformations, are described in detail.

KEY WORDS • arteriovenous malformation • operative approach • limbic system • temporal lobe • hippocampus • seizure • intraventricular hemorrhage

Arteriovenous malformations (AVM's) located on the medial aspect of the cerebral hemisphere and involving portions of the limbic system represent a special group of malformations because of their obscure location and the difficulties encountered in their resection. Heros3 reported three AVM's of the medial aspect of the temporal lobe, involving the parahippocampal gyrus, uncus, and choroidal fissure of the temporal lobe, and bordering on the basal ganglion. These were removed without major neurological deficit. Drake,1 in a review of his experience with 166 AVM's, specifically mentioned those located on the medial aspect of the temporal lobe in close proximity to the trigone of the lateral ventricle, and the various exposures that can be used to resect them successfully. Yaşargil, et al.,11,12 detailed the surgical approach for successful removal of those malformations located in portions of the corpus callosum and cingulate gyrus. Although the cases reported by Martin and Wilson6 were on the medial aspect of the occipital lobe in intimate relationship to the calcarine cortex, this type of case is not included in this discussion except where general principles apply, as in the parafalx approach and retraction of the hemisphere to bring the malformation into view.

In this report these malformations have been classified by region, which is relevant to the surgical approach. The regions have some degrees of overlap (Fig. 1). Region A is the amygdaloid-uncus region or the anteromedial aspect of the temporal lobe. Region B is the parahippocampal and fusiform gyrus region involving the medial aspect of the temporal lobe at its mid and more posterior aspect. The fusiform gyrus lies somewhat lateral to the parahippocampal gyrus and may be identified in an exposure of the former region or may itself be involved by extension of an AVM. Region C is the region medial to the trigone of the lateral ventricle or the parapeduncular region which also incorporates the posterior parahippocampal and fusiform gyri. Region D includes the splenium and posterior third ventricle. Often malformations in this area also involve the posterior aspect of the cingulate gyrus. Region E is the corpus callosum-cingulate region, including the medial frontal or parietal regions, depending upon the anteroposterior site of the lesion. Region F is the septal or medial frontal region.

Among a series of 164 AVM resections, 25 of the lesions were located in the medial hemispheric or limbic region. These represent 15% of the total AVM's. There were four AVM's in Region A, four in Region B, three in Region C, six in Region D, six in Region E, and two in Region F. These AVM's had clinical features characteristic of AVM's in general; 16 presented with hemorrhage and nine with seizures.
Anatomical Considerations

These cases present a unique challenge for the neurosurgeon to obtain an adequate exposure for total removal of the lesion if a good result is to be achieved. The basic problem created by the location of these malformations is that they are often approached tangentially rather than perpendicularly. A tangential exposure to an AVM limits the surgeon’s ability to deal precisely with the feeding arteries while preserving the major draining veins as he circumscribes the lesion by incision. These malformations often have dual venous drainage, with major draining veins extending to the sagittal sinus via the cortex and to the deep venous system including the internal cerebral veins or vein of Galen, and hence to the straight sinus of the tentorium (Fig. 2). The importance of this feature is that the surgeon may sacrifice one of the main venous drainage systems in order to improve the exposure, while relying upon the secondary system to decompress the AVM. The arterial supply to these malformations, unlike the superficial arterial supply to most convexity malformations, comes from deep and often obscure arteries; these include the primary trunks or branches of the anterior and posterior choroidal arteries and the posterior cerebral artery (Fig. 3). Because of their relationship to the choroidal arteries, these malformations are often contiguous with the choroid plexus, which contains part of the feeding and draining vasculature of the malformation.

Surgical Considerations

Prior to surgery, it is essential that the exact location of the lesion and its anatomical features be delineated. This is accomplished by computerized tomography (CT), with and without contrast infusion, which gives an excellent indication of the relationship of the malformation, especially the thrombosed parts, to the ventricular system and other important portions of the medial hemisphere (Fig. 4). Stereoscopic angiography in the lateral projection with an anteroposterior shift of the x-ray beam is most helpful in displaying the three-dimensional anatomy of these complex lesions.
Exposing these lesions involves the general principles that apply to AVM surgery elsewhere in the brain. Because of the obscure location of most of these lesions, proper positioning of the patient and relaxation of the brain are essential. Spinal drainage is installed before the patient is positioned, and up to 150 cc of cerebrospinal fluid (CSF) is removed before the operation; the usual amount being 60 to 100 cc. Rarely are dehydrating agents required. The patient's head is then positioned securely in an orientation that will place the AVM as high as possible above the heart. In those lesions in which a temporal or parietal approach is used, the patient is placed supine with the area to be approached uppermost. When a parafalx approach is utilized, the patient is placed in a slouched sitting position (Fig. 5).

It is important that a “keyhole” type of surgical exposure not be used. For the parafalx exposure the craniotomy should be made slightly to the opposite side of the midline. An inadequate exposure limits brain retraction and predisposes to cerebral injury. As in all AVM's, the major arteries are secured and divided first whenever possible. One large draining vein must be left intact until the end of the operation. In order to reach the obscure location of these lesions with adequate exposure to permit precise removal, the operating microscope is recommended in all cases. Hypotensive anesthesia can be most useful when a deep artery is being secured, or when the complex vasculature of the choroid plexus is involved with the lesion.

As to the specific details of the various surgical approaches, one may picture these lesions as occupying the various positions on a clock face when the right hemisphere is viewed from its lateral aspect. Briefly, those lesions located in Region A would be approached from a 3 or 4 o'clock direction, those in Region B from a 6 o'clock direction, those in Region C from an 8 o'clock direction, and those in Region D from 10 and 11 o'clock via a parafalx approach; posterior AVM's in Region E would be approached from a 10 or 11 o'clock direction and anterior AVM's in Region E from a 1 or 2 o'clock direction; AVM's in Region F would be reached from a 2 or 3 o'clock direction (Fig. 1).
Operative Approaches

Region A Lesions

Those AVM's involving the amygdaloid nucleus and uncus (Region A) are supplied predominantly by branches of the anterior choroidal, posterior communicating, and thalamoperforating arteries (Fig. 6). There are also minor contributions from the main branches of the middle cerebral artery. These lesions, being located anteriorly, are best approached by a pterional craniotomy, in which the Sylvian fissure is opened widely and the malformation skimmed off the branches of the aforementioned arteries. The anterior choroidal artery is an essential landmark in pursuing the arterial supply to the malformation, and its main division must be left intact. A transcortical incision then circumcribes the malformation with resection of the amygdaloid and uncal regions. These lesions often drain via the basal vein of Rosenthal, running posterior and medial from the malformation. This should be the last vascular element that is occluded.

Region B Lesions

Those AVM's located in the parahippocampal fusiform gyrus region (Region B) are resected via a subtemporal exposure (Fig. 7). This may require a modest resection of the inferior temporal gyrus or, as Heros recommends, a transcortical incision whereby the fusiform or the inferior temporal gyrus is incised to reach the malformation. This requires retraction in two directions, and it is preferable to resect a small area of the inferior temporal gyrus, even in the dominant hemisphere. This will provide exposure to medially placed AVM's that receive blood supply from the anterior choroidal artery or, more often, from a large feeder from the inferior temporal branch of the posterior cerebral artery (Fig. 8). These arteries must be divided before a transcortical incision is made around the malformation. The venous drainage may be cortical around the inferior convexity of the temporal lobe, but may also be medial and deep via the vein of Rosenthal. One major draining vein must be preserved until the AVM is removed.
Region C Lesions

Malformations located medial to the trigone of the lateral ventricle or parapeduncular in relation to the midbrain (Region C) are most difficult to approach, even after resection of the inferior temporal gyrus, because of the upward slope of the tentorium (Fig. 9). Because their blood supply comes from the posterior choroidal and posterior cerebral arteries, where these vessels course around the cerebral peduncle, an interhemispheric approach may be too distant from the arterial sources. This can result in an approach to the venous side rather than the arterial side of the malformation. If the lesion is too large to be reached by a subtemporal approach with a moderate resection of the inferior temporal gyrus, then it must be approached through a transcortical incision. Drake has used this exposure in the majority of his cases with or without hemorrhage. Lesions that have bled usually have a large cystic area around them and are easier to approach transcortically. A short incision must be made in the posterior aspect of the middle temporal gyrus and carried down to the ventricle. The arterial supply will be encountered from the choroidal fissure of the temporal horn and should be visualized and secured first. This exposure has the disadvantage of encountering the malformation at its body rather than at its margins, where the arterial supply enters it. The venous drainage is to the ependymal veins and then deep to the internal venous system. Rarely do these malformations drain over the convexity. However, with difficulty the AVM can be removed via this route. An alternative to the one-stage operation is to carry out the resection in two stages. The approach for the first stage is via the posterior infratemporal route, and as much of the malformation as possible is secured. The second stage involves an interhemispheric parafalx exposure to remove the residual portion of the malformation and its venous drainage.

Region D Lesions

Malformations that are located in the splenial and posterior third ventricular region (Region D) can only be treated by an interhemispheric parafalx approach (Fig. 10). This is carried out with the patient in a slouched sitting position. With adequate relaxation of the brain and division of one or two of the nonessential parietal bridging veins, these AVM’s can be approached with a high degree of safety and adequate exposure. The blood supply to these malformations comes from the posterior medial and lateral choroidal arteries as well as branches of the posterior cerebral artery. In those AVM’s that extend into the splenium, there is often a supply from a major branch of the distal pericallosal artery. This may reach the malformation directly over the corpus callosum under the falx or by a circuitous route through the cingulate fissure and then to the malformation laterally. These important features are determined from the preoperative angiographic studies. Any of the arteries dorsolateral to the brain stem can be reached via this exposure. However, the surgeon can reach little further in a deep lateral direction around the side of the brain stem. These malformations may be confined to the midline or just off the midline. Unfortunately, in a number of cases they extend out to the trigone of the lateral ventricle and thereby involve the choroid plexus (Fig. 11). The limited retraction permitted on the parietal lobe makes access to this aspect of the malformation difficult. It is often at this point that the surgeon is wrestling with the last arterial supply to the malformation and with large arterialized
FIG. 10. **Left:** Right lateral carotid arteriogram demonstrating an arteriovenous malformation (AVM) of Region D, fed primarily by branches of the anterior cerebral artery (*arrow*) and posterior choroidal arteries (obscured by overlying veins). **Right:** Right lateral vertebral arteriogram of the same patient demonstrating the vascular supply of this AVM from the posterior cerebral and posterior choroidal arteries (*arrowheads*) and the involvement of the roof of the third ventricle and splenium. The AVM permeates the splenium at a point identified by the *arrows*.

FIG. 11. Anteroposterior vertebral arteriogram demonstrating a malformation involving the choroid plexus of the lateral ventricle at the trigone fed by branches of the posterior choroidal arteries (*arrows*).

FIG. 12. Right lateral carotid arteriogram demonstrating an arteriovenous malformation (AVM) of the posterior cingulate gyrus and corpus callosum fed primarily by branches of the anterior cerebral arteries (*arrows*). This AVM occupies Region E.
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FIG. 13. Left: Right lateral carotid arteriogram demonstrating an arteriovenous malformation (AVM) involving the mid-anterior portion of the corpus callosum and cingulate gyrus fed primarily by branches of the anterior cerebral arteries (closed arrows). The venous drainage is primarily to the deep venous system and associated with venous varices (open arrows). This AVM occupies Region E. Right: Postoperative arteriogram in the same patient showing the changes of spasm in the preserved branches of the anterior cerebral artery intimate to the AVM (arrowheads).

veins draining into the ependymal region of the lateral ventricle. At best, this is a delicate situation, demanding brain relaxation, hypotension, and the finest of illumination and microsurgical techniques. It may also be necessary to resect a portion of the posterior fusiform and cingulate gyri in order to remove the entire malformation. The venous drainage to the vein of Galen or internal cerebral veins can be separated without injury to these vital structures.

Region E Lesions

Malformations that are located in the cingulate gyrus and corpus callosum (Region E) are divided into anterior and posterior locations. The AVM's located in the posterior region are not dissimilar from those already described in Region D, except that these extend into the cingulate gyrus and the medial aspect of the parietal lobe (Fig. 12). Their blood supply is primarily from terminal branches of the anterior cerebral artery, occasionally from anastomotic branches of the middle cerebral artery over the cerebral convexity, and from medial hemispheric branches of the posterior cerebral artery. Their venous drainage is to the convexity via the medial hemisphere and frequently also to the vein of Galen; therefore, they have the potential for dual major venous drainage. Occasionally there may be drainage into the ependymal veins, especially with lesions that extend laterally toward the ventricle. Relaxation of the brain and retraction to bring the medial surface of the hemisphere into perspective are essential. A fair amount of retraction of the parietal lobe may be necessary. The arterial supply is secured first and the deep venous drainage left intact, while sacrificing the more superficial draining veins.

Lesions that are located in the anterior cingulate gyrus and corpus callosum may involve the medial frontal lobe and are supplied almost exclusively by large branches of the anterior cerebral artery with occasional collateral branches of the middle cerebral or transcallosal arteries, the latter originating from deep perforating arteries. It is important to resect these lesions without damaging the anterior cerebral arteries (Fig. 13). Again, relaxation of the hemisphere and sufficient retraction to expose the surface of the lesion are essential. The veins draining the Rolandic area should be left intact, and therefore veins anterior or posterior to these are sacrificed in order to effect the exposure. Profound motor deficits may ensue after interruption of the Rolandic veins; fortunately, these generally clear after 2 to 3 weeks. The venous drainage of these lesions is often to the sagittal sinus, and occasionally to the ependymal veins. Since the arterial supply enters anteriorly, and the venous drainage is generally posterior and superior, these lesions may be approached easily from the arterial side via an interhemispheric approach. Extension around and deep to the main trunk of the anterior cerebral artery into the corpus callosum or frontal horn of the lateral ventricle makes the resection more difficult.
Region F Lesions

Malformations located in the septal or medial frontal region (Region F) are approached by a low frontal craniotomy using the principles already stated for interhemispheric parafalx surgery with the patient in the supine position (Fig. 14). Care is taken to preserve the main trunks and branches of the anterior cerebral and perforating arteries of the anterior perforated space. The venous drainage of these lesions is often dual: deep to the ventricular region and superficial to the sagittal sinus.

Operative Results

Of these 25 cases, 18 had primary surgical resection, and seven underwent embolization followed by surgical resection. There were no incomplete resections. Embolization was reserved for the more complex cases and then used in accordance with previously established principles. Using techniques developed at this institute by Hilal, et al. (unpublished data), it is now possible to embolize into the anterior cerebral artery. Twenty patients had an excellent result, with normal neurological status. Two patients had a good result; that is, although they might have minimal paresis, they returned to their previous employment, or remained modestly handicapped by preoperative neurological deficits secondary to hemorrhage. Two patients had a fair result with a moderate degree of paresis or other neurological deficit but were able to care for themselves independently. One patient had a poor result with neurological deficits that prevented independent existence. All patients had postoperative arteriography, and there was no evidence of residual AVM in any of these patients.

Discussion

These malformations, occupying a position on the medial aspect of the hemisphere in relation to the limbic system, deserve special attention but have rarely been singled out for detailed discussion. Martin and Wilson discussed the difficulties in approaching the medial aspect of the hemisphere, but limited their review to 16 cases involving the occipital lobe, especially the region of the calcarine cortex. They stated that adequate exposure could be obtained in order to minimize visual field defects. A number of neurosurgeons have described the technique of resection of anteromedial deep temporal AVM's. Heros reported three malformations in obscure locations on the medial aspect of the temporal lobe involving the choroidal fissure and lying adjacent to the basal ganglion and internal capsule. He specifically noted the difficulties in exposing these lesions and the necessity (in his estimation) of transcortical incisions in order to minimize brain retraction. He underscored the importance of preserving the principal arteries that gave branches to these malformations, while removing the entire malformation cleanly. His results were excellent in terms of preservation of the visual pathways and neurological function, and the lesions were totally removed.

Drake emphasized the difficulties in approaching these medial hemisphere AVM's and divided them into regions similar to those that we have described. He did not specifically indicate how many lesions were involved in each area, although he did report six medial temporal-lobe AVM's and five corpus callosum AVM's. Those lying on the medial hemisphere close to the corpus callosum were exposed by an interhemispheric parafalx approach with the patient in the semi-sitting position. Malformations located in the hippocampal region or in the medial temporal-occipital region were reached by a subtemporal approach. On some occasions, in order to preserve the vein of Labbé, the incision was carried out in front of and behind this vein, while preserving it as an island. Drake also takes note of the position of the AVM so as to avoid the visual radiations in the surgical approach. Lesions located in the trigonal region were approached via a transcortical incision in the posterior temporal region. The results for the medial AVM's were not specifically singled out for review; however, among 159 cases of AVM's located in all regions, 16 patients died and 16 patients had poor results.

Yasargil, et al. described 10 lesions of the posterior corpus callosum and splenium. They pointed out...
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the difficulties entailed in resection of lesions in this area and the high incidence of rupture of these lesions into the ventricular system. They also emphasized the need for preoperative stereoangiograms, operating in the sitting position, and the operating microscope. There were no deaths among their 10 patients. Two had hemianopsias, and one had a residual AVM that had to be removed at a second operation. These authors pointed out that medial hemisphere lesions might be hidden in the substance of the corpus callosum, and often extend to receive blood supply and give venous drainage to the choroid plexus of the trigone. In describing eight cases of the anterior and middle corpus callosum, they emphasized the intimate involvement that these lesions have with the ventricular system, by the fact that their major blood supply is from the anterior cerebral arteries and they drain into the ependymal vein of the lateral ventricle.

Luessenhop and Gennarelli 5 presented a grading scheme based on more than 300 angiograms of AVM's, and correlated the grade with the outcome following surgery. Although many varieties were classified, none of the grades included the group of malformations that have been discussed here. This seems paradoxical, since the purpose of the grading system was to pinpoint those malformations which were most difficult to remove and which might be expected to be associated with the highest risk of morbidity and mortality. Considering that lesions located on the medial hemisphere are some of the more difficult malformations to remove, it seems only fitting that they should be included as a specific group in a comprehensive grading scheme.

Summary

A group of 25 AVM's located on the medial aspect of the hemisphere in intimate relationship to components of the limbic system is presented. These AVM's are unique in terms of the difficulty they pose in obtaining an adequate exposure to permit resection. Most of them are located in obscure areas and are therefore approached tangentially. Special techniques must be utilized, in terms of brain relaxation, retraction, and exposure of the lesion, in order to effect a clean removal with minimal risk of morbidity. These lesions are also associated with a higher incidence of intraventricular hemorrhage, since their arterial supply and venous drainage often involves the lateral ventricles and choroid plexus. In other aspects they present in a similar fashion to malformations located elsewhere in the cerebral hemisphere.

The basis of surgery of these lesions includes the appropriate position of the patient's head, and the use of brain relaxation, hypotension, and the operating microscope. Embolization techniques that are tailored to meet the demands that these malformations present have been successful in reducing the volume of the larger malformations preparatory to surgery.

Considering the obscure location of these lesions and the difficulty encountered in surgical exposure, the results of excision are good. Of the 25 cases, results were excellent or good in 22 cases. Two patients had a fair outcome, and one a poor result. No patient died. The rate of serious morbidity therefore was under 4%.

References


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