Operative management of brainstem cavernous malformations

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Cavernous malformations are vascular lesions of the CNS with an estimated prevalence of 0.4–0.9% in the general population. The proportion of CMs occurring in the brainstem is difficult to determine precisely, as brainstem lesions are more likely to be symptomatic than similar lesions in the cerebral hemispheres. Therefore, it is presumed that a disproportionate number of brainstem CMs come to clinical attention.

Cavernous malformations are believed to cause symptoms in several ways. They are implicated in the etiology of a patient’s new symptoms when there is radiographic evidence of recent hemorrhage (on MR imaging, usually) as well as a correlation between the location of the lesion and the patient’s neurological deficit. Hemorrhages can be entirely intralacional, in which case new clinical findings probably represent increasing pressure on adjacent neural structures by the expanding capsule. Alternatively, hemorrhages can extravasate outside the bounds of the malformation, in which case symptoms may be caused by the direct interaction of adjacent structures with blood and its breakdown products as well as by local mass effect. Over time, because of repeated hemorrhage—many of which are suspected to be clinically silent—CMs develop a hemosiderin-laden margin.

This injured tissue may contribute to the symptomatology; supporting this theory are hemispheric CMs associated with seizures, in which the complete resection of surrounding hemosiderin-laden tissue has been found to yield better seizure control.

The clinical presentation of brainstem CMs often correlates with their anatomical location. Somatic motor and sensory symptoms predominate, as would be expected given the presence of these tracts along the entire axis of the brainstem. Oculomotor abnormalities are more common with lesions of the mesencephalon compared with other portions of the brainstem. Ataxia, meanwhile, has been reported more commonly with lesions situated toward the medulla—although this is not always the case, as cerebellar long tracts and cerebellar peduncles are present in all segments of the brainstem. Vertigo and nausea have been noted in conjunction with pontine lesions. Trigeminal neuralgia has been documented with intraaxial lesions at the root of CN V. Headache, too, is a common associated complaint; in the absence of hydrocephalus, however, the degree to which these headaches are etiologically related to the CM is unknown. Lesions can also be completely asymptomatic; presumably more frequently than previously thought given the increased use of neuroimaging for nonspecific symptoms, such as headache, during screening for metastatic lesions in cancer patients or after a minor head injury.
Clinical Decision Making

The decision to pursue operative intervention for brainstem CMs must take into account the lesion location and size, patient age, the nature of the presenting signs and symptoms, and the potential neurological deficits associated with surgery. Experience at many centers suggests that there are indeed safe ways to proceed. For example, a retrospective review of 87 patients who underwent resection of a brainstem CM revealed that 78 (90%) were neurologically stable or improved at a later follow-up. Note, however, that at least transient postoperative neurological signs or symptoms are very common and occur in the majority of patients.

When a patient presents with a brainstem CM, either incidentally or through the appearance of new neurological signs or symptoms, the decision to recommend intervention depends primarily on 3 core factors: 1) the estimated risk of future hemorrhage, that is, the natural history of the lesion; 2) the form of neurological injury predicted to occur with such a hemorrhage; and 3) the patient’s clinical status, specifically, his or her current neurological condition and age and the size and location of the lesion.

The risk of brainstem CM bleeding is generally reported to be higher than the risk of hemorrhage from a CM situated elsewhere. This phenomenon may be explained by the likelihood that lesions located outside the brainstem generally occur in less densely eloquent areas of the brain and are therefore more apt to be clinically silent when hemorrhage occurs. Nevertheless, lesions occurring in different areas may have different underlying pathophysiology, and variations in local anatomical structure can, in theory, influence their likelihood of hemorrhage. In general, the annual risk of hemorrhage for a brainstem CM is reported to be between 2% and 21%. Finally, hereditary CMs have been linked with increased propensity to bleed.

For any individual, the estimated risk of injury from the hemorrhage of a brainstem CM is usually estimated from its specific location with respect to brainstem tracts and nuclei, and—most usefully, perhaps—from that individual’s relevant neurological history. While the effects of a lesion can be predicted with reasonable accuracy on anatomical grounds alone, the surest guide is each patient’s particular experience. Therefore, if prior symptoms consisted entirely of a tolerable deficit such as somatic sensory disturbances, then the low-pressure, low-volume forms of hemorrhage common with CMs make sudden devastating deficits as a result of a rehemorrhage less likely. Nonetheless, there is some evidence that when hemorrhage is accompanied by new neurological symptoms, early surgery to resect the brainstem CM might yield better outcomes.

In addition to the risk of future hemorrhage, an individual’s current functional status is important when establishing an assessment of risks and benefits. Where someone lies on the neurological spectrum—from asymptomatic and fully independent to symptomatic and dependent—determines his or her marginal risk. For instance, someone who is moderately disabled yet retains just enough function to be somewhat independent may have little tolerance for a slight worsening of symptoms. In such a case, intervention would be considered to prevent any further decline. Conversely, persistent symptoms, with or without evidence of hemorrhage, may be a result of mass effect; in these cases, removing the lesion could result in an improvement of neurological functioning. A full assessment of these factors, along with common sense considerations of a patient’s overall health and age as well as his or her individual preferences, informs the decision to observe and follow or to recommend intervention.

Treatment

There is usually agreement that an incidentally discovered brainstem CM in an asymptomatic individual is not a candidate for resection. On the other hand, if intervention is desirable (for example, in a person with multiple symptomatic hemorrhages causing additive deficits), the particular surgical approach depends on the accessibility of the lesion and on the patient’s ability and desire to tolerate an operation and its associated risks. If these factors are unfavorable, radiosurgery is sometimes considered; however, the true efficacy of radiosurgery in
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Reducing future rehemorrhages is difficult to assess. Any radiation-derived benefits are expected to occur in a delayed fashion (beginning many months after treatment), typified by the radiobiological effect following single-fraction therapy precedence seen elsewhere in the human cerebrum. Of note, the natural history of CMs suggests that after the same time period, a patient's risk for rehemorrhage will spontaneously decline. Therefore, studies that document a reduction in the risk of hemorrhage by radiosurgery, particularly after 2 or more years from the preceding hemorrhage, do not necessarily indicate an effect of treatment but may simply chart the natural course of these lesions. The possibility remains that radiosurgery offers a risk reduction over and above that due merely to the passage of time. A prospective study comparing radiosurgically treated patients with untreated controls will therefore be required to measure its true benefit.

In light of the remaining uncertainties regarding the efficacy of radiosurgery and the potential complications associated with radiation administered to the brainstem, operative resection represents the generally accepted method for reducing the probability of future hemorrhage from a CM. The relative complexity of resection for a particular brainstem lesion is dependent on its location and intrinsic characteristics. These factors are each considered in turn.

A lesion that abuts a pial or ependymal surface is often directly accessible without the need for traversing normal brainstem tissue. One that lies deeper entails a potentially increased risk in proportion to its distance from a pial or ependymal surface and in relation to those brainstem nuclei or tracts lying superficial to it. In experienced hands, however, outcomes after the resection of deeper lesions can be comparable with those following the resection of lesions rising to meet a pial surface. Obstacles to the excision of a deep-seated lesion are implicit given the lesion's position within the known standard anatomy of the brainstem, but if necessary can be made more explicit with the aide of MR imaging. While typical MR imaging sequences do not provide discrimination of brainstem structures with significant contrast, newer methodologies, if locally available, can provide useful anatomical detail. Functional MR imaging, for example, is capable of revealing the locations of lower CN nuclei. The complementary technique of diffuser tensor imaging may resolve the positions of axonal tracts within the brainstem and can be used to guide the surgical approach to a CM. While the highly conserved nature of brainstem anatomy across individuals usually renders more exhaustive neuroimaging in any particular case unnecessary, these diagnostic studies might be useful because of their potential to reveal the true positions of structures displaced by larger lesions; knowing whether a motor tract is displaced medially or laterally by a lesion in the medulla, for example, could help determine the most advantageous pial point of entry. The integration of these imaging techniques with intraoperative neuronavigation technologies will likely improve safety during the resection of CMs juxtaposed to white matter tracts and nuclei.

A deep-seated lesion is typically less accessible because it may not be associated with any surface irregularities (for example, distortion of the contour of surface structures or hemosiderin staining) that help reveal its location intraoperatively. The potential difficulty in localizing the lesion intraoperatively must therefore be considered preoperatively and appropriate measures taken in advance to reduce uncertainty during the procedure. In many cases, dead reckoning can be used to correlate a lesion's radiographic position with normal anatomical features that are clearly identifiable on the surface of the brainstem. If dead reckoning proves insufficient or is uncertain for any reason, other strategies are available. Ultrasonography has been shown to be a relatively sensitive methodology to locate CMs, although the surgeon's experience with this technique will determine its specificity. Intraoperative localization of a lesion can be achieved with the use of frameless stereotaxy as well. Because the need to minimize registration error is paramount in the brainstem, and because many brainstem approaches require prone positioning in which the highly contoured face is largely inaccessible, the use of fiducial markers is recommended. With these markers, the relatively featureless occiput can be populated with unambiguous landmarks. These fiducials can be applied prior to imaging and are subsequently used intraoperatively to register the images with the operative field. Finally, intraoperative neuroimaging can be used in cases of brainstem lesions. This process can help guide the surgeon during resection if manipulation causes a change in the positions of critical structures.

Operative Approaches and Techniques

Because the brainstem is a large and diverse structure, the selection of a particular operative approach out of the many that are possible is dependent on the particular location of the lesion and the experience of the surgeon (Table 1). Ideally, the cranial approach should allow for a line of sight in which the surgeon, the pial incision, and the lesion itself are all collinear. This alignment will provide the best view of the lesion while minimizing the need for brainstem retraction. Meanwhile, the pial incision should be chosen to minimize the distance to the lesion while avoiding critical nuclei and tracts.

The intrinsic characteristics of a CM influence the ease with which it is removed and determine the extent of the exposure and the degree to which it will be hemorrhagic. Calcified lesions with a solid, mineralized portion may necessitate a larger surface incision for removal. Meanwhile, previously irradiated lesions are seen to be more friable and hemorrhagic.

Mesencephalic Lesions

In general, operative resection of mesencephalic lesions is considered high risk and is undertaken in only the most compelling circumstances. If an individual is symptomatic primarily because of compression of the cerebral aqueduct by a lesion, supratentorial CSF shunting is the preferred interventional modality. If, on the other hand, an individual is suffering from multiple, progressive neurological deficits attributable to a mesencephalic lesion
abutting a pial surface, resection is undertaken only with the understanding that further neurological deficits are very likely, at least in the short-term.

Lesions of the dorsal mesencephalon in the vicinity of the colliculi can be accessed via a supratentorial, infratentorial, or combined approach (Fig. 1). The patient can be positioned prone or in a semisitting position allowing gravity to aid retraction of the cerebellum inferiorly. If the latter position is selected, precordial Doppler ultrasonography and central venous access are recommended to detect and evacuate potential air emboli. Cavernous malformations that occur next to the pial surface are resected through a small incision precisely over the area of yellowish discoloration. However, if the lesion is deep and not associated with surface irregularities, this operative intervention is treacherous. The direction of the incision should be parallel to the nearest underlying fibers. One must keep in mind that in this region, CN IV, visualized caudal to the inferior colliculi, decussates just beneath the pial surface within the superior medullary velum; an anteroposterior midline incision here would transect both nerves, resulting in bilateral trochlear nerve palsies. Meanwhile, more rostrally and laterally, fibers of the brachium of the superior and inferior colliculi also travel horizontally, carrying afferents from the geniculate nuclei. When resecting a lesion here, care must be taken to remain within the tectum. Medially, one must avoid entrance into the cerebral aqueduct and certainly should not violate the floor of this structure, which contains the third and fourth CN nuclei. More laterally, lying at the border of the colliculi and the periaqueductal gray matter, one will encounter the mesencephalic trigeminal tract, which carries proprioceptive information. This tract is involved in the masseter reflex arc; damage to it does not necessarily lead to loss of facial sensation or motor function.65

Electrophysiological recording of the oculomotor system can be performed to map these important structures prior to resection of the CM.55 However, there is less certainty regarding the predictive ability of mechanically evoked electromyographic activity in the extraocular muscles to signify injury to the brainstem oculomotor system.54 Therefore, the oculomotor system is more easily mapped than monitored. In general, this technique requires placement of an electrode into 1 or more extraocular muscles (usually by an ophthalmologist after the induction of anesthesia) to monitor contraction when the relevant midbrain oculomotor nuclei or tracts are electrically stimulated by the surgeon. This stimulation can be delivered using a bipolar device across a small gap (for example, 1 mm) with low current (approximately 1.0 mA) and low frequency (1–5 Hz). In this way, the oculomotor structures can be mapped and avoided so that postoperative deficits can be minimized.

The ventral mesencephalon is also a high-risk operative region; it contains the cerebral peduncles laterally, the substantia nigra just dorsal to them, and the red nuclei forming the bulk of the intraaxial mass. Lesions here can be accessed via a modified ptetional craniotomy. A modified ptetional craniotomy, in which the posterior arc is exaggerated, extending just beyond the level of the external auditory meatus, will afford better exposure than a standard ptetional craniotomy by allowing the temporal lobe to be retracted posteriorly to reveal the floor of the middle cranial fossa. Meanwhile, an orbitozygomatic modification, which involves removal of the orbital roof, provides good exposure of the interpeduncular and preoptic cisterns via a shallower approach along the floor of the middle cranial fossa.30 A subtemporal transpetrosal route is also available to provide increased exposure of the caudal mesencephalon/rostral pons (Fig. 2).34

At least 1 group has described the successful resec-

**TABLE 1: Various approaches to the brainstem depending on the lesion location**

<table>
<thead>
<tr>
<th>Lesion Location</th>
<th>Anterior</th>
<th>Lateral</th>
<th>Posterior</th>
</tr>
</thead>
<tbody>
<tr>
<td>Midbrain</td>
<td>ptetional orbitozygomatic subtemporal</td>
<td>lateral infratentorial/supracerebellar</td>
<td>lateral infratentorial/supracerebellar</td>
</tr>
<tr>
<td>Pons</td>
<td>ptetional orbitozygomatic subtemporal</td>
<td>far lateral suboccipital retrosigmoid</td>
<td>lateral infratentorial/supracerebellar</td>
</tr>
<tr>
<td>Medulla</td>
<td>subtemporal far lateral suboccipital</td>
<td>lateral suboccipital retrosigmoid</td>
<td>midline suboccipital/4th ventricular</td>
</tr>
<tr>
<td>Transcondylar</td>
<td></td>
<td></td>
<td>midline suboccipital/4th ventricular</td>
</tr>
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![Fig. 1. Drawing showing the approach to the dorsal midbrain.](image)
tion of a CM in this region by utilizing intraoperative monitoring of motor evoked potentials generated by direct bipolar stimulation of the cerebral peduncles as well as by transcranial motor evoked potentials.49 The homunculus is oriented medially to laterally, face to leg, along the surface of the peduncles; the appropriate muscles can be monitored depending on the location of the lesion. If the underlying lesion is sufficiently superficial, the overlying corticospinal tracts may have functional gaps created by the displacement of fibers or by direct damage resulting from compression or chemical irritation by the lesion. Furthermore, based on knowledge of the homunculus, regions corresponding to a patient’s established deficit can be mapped. A myelotomy can then be made in an anterior-to-posterior direction at a medial-to-lateral position chosen to minimize injury to remaining functional tissue.

In general, even when a lesion appears to present directly to the pial surface in the region of a known motor pathway, preoperative arrangements to perform electrophysiological monitoring are useful because standard anatomical MR imaging alone is an insufficient guide to the extent and nature of the overlying tissue, especially when that tissue consists of axonal tracts, which can be very thin yet functionally significant.

Metencephalic Lesions

Cavernous malformations located in the metencephalon can be accessed either through the floor of the fourth ventricle dorsally or via the pons ventrally. The former is achieved through a midline suboccipital approach, traversing the obex toward the rhomboid fossa. The latter is approached most commonly through a lateral suboccipital craniotomy (Fig. 3).

Lesions within the floor of the fourth ventricle are generally considered resectable if they present to or through the floor’s surface (Fig. 4). Careful planning with respect to the relevant anatomy can minimize the risk of neurological injury, particularly when considering access to deeper lesions causing progressive deficits. The critical structures to avoid are clustered into 2 groups represented on the surface of the floor of the fourth ventricle by the facial and hypoglossal colliculi. The facial colliculi, formed by the sixth CN nuclei and the overlying traversing fibers of the seventh CN, are located in the base of the
rostral triangle of the rhomboid fossa. These structures are within 2–4 millimeters of the surface. More caudally, the hypoglossal colliculi are less prominent than the facial colliculi; they represent the rostral portion of the hypoglossal nuclei, which are long structures (approximately 11–12 mm) crossing the pontomedullary junction. Running parallel to the midline sulcus is the MLF, anteriorly composed of fibers from the sixth nerve nucleus and vestibular nuclei, which mediate coordinated voluntary eye movements as well as the vestibuloocular reflex, and posteriorly composed of fibers that likely mediate head movements associated with gaze shifts.

In addition, there are many important structures not immediately evident externally but whose position can be inferred from external landmarks. In close association with the hypoglossal nucleus is the dorsal parasympathetic motor nucleus of the vagus nerve. This nucleus is just lateral to the rostral head of the hypoglossal nucleus but courses dorsomedially to lie more superficially in the medulla. Lateral to this nucleus is the solitary nucleus. Most laterally, at the ventrolateral angle of the rhomboid fossa and extending caudally into the medulla, the vestibular nuclei are found. They are situated between the fourth ventricle and the cerebellar peduncles more laterally. The motor and principal sensory trigeminal nuclei are also near the lateral angles of the fourth ventricle, just ventral to the superior vestibular nuclei.

With this anatomical knowledge in mind, the 2 visible structures—the facial and hypoglossal colliculi—serve as landmarks for 2 approaches into the dorsal metencephalon.6,62

The first of these approaches is oriented rostral and lateral to the facial colliculi. A paramedian approach is necessary to avoid injury to the ascending MLF. The rostral limit of this approach is defined by the decussation of the trochlear nerves in the superior medullary velum; vigilance is necessary to avoid retraction injury to this structure.64 This approach leads into nuclei of the descending pontine reticular formation. While bilateral imaging abnormalities of this region have been associated with sleep disturbances,43 unilateral damage to these structures during resection of a brainstem CM reportedly does not interfere with level of consciousness or the sleep-wake cycle.17

The second approach enters into the region of the descending reticular formation between the level of the facial and hypoglossal colliculi. A paramedian entry is also necessary here to avoid injury to the descending MLF. The descending reticular formation, through efferent nerves that inhibit the spinal cord, is involved in REM (rapid eye movement) sleep paralysis. While bilateral manipulation of this structure in animal models is associated with altered sleep,23 unilateral damage in humans during surgery has not been reported to cause significant dysfunction. The major structures ventral to the reticular formation at this level but still dorsal to the pontine nuclei are sensory and special sensory in nature. These structures include the medial lemniscus (somatosensory), lateral lemniscus (auditory), trigeminal pathways, and the central tegmental tract (taste).

Electrophysiological monitoring and direct stimulation can be applied to identify and preserve many of these important neural structures and pathways.12,20,39,40 As previously mentioned, oculomotor monitoring can be used to identify the location and extent of relevant structures. Here, the electrically evoked activity of the abducens nucleus can be tested using recording electrodes in the lateral rectus muscles. Localization of the overlying facial nerve fibers can be achieved by recording motor evoked potentials from the orbicularis oris and the orbicularis oculi. Importantly, as appears to be true for the other oculomotor CNs, there is no clear predictive relationship between electromyographic activity in the lateral rectus muscle and preserved CN VI function;18 interestingly, activity of the lateral rectus did suggest damage to the facial nerve. When working laterally, the motor trigeminal nucleus can be monitored with recording electrodes in the masseter. Meanwhile, stimulation in the region of the hypoglossal nucleus can be detected by electrodes placed in the tongue. Here, care must be taken with regard to possible current spread into the nearby dorsal motor nucleus of the vagus, causing bradycardia. In all cases, one must keep in mind the potential to directly injure stimulated tissue. In general, higher currents will be required if using monopolar (>1–2 mA) versus bipolar (<1 mA) stimulation. However, it should be remembered that the risk of injury is a function of not just current intensity, but also the charge delivered per unit of time and per unit of electrode surface area.

Pontine lesions can also be approached ventrolaterally at times. Either a far lateral suboccipital craniotomy or a larger craniotomy for a combined supra-/infratentorial approach can be used to gain access to the ventrolateral surface of the lower brainstem.6 In addition, a subtemporal approach is possible for anteriorly situated lesions that may not be accessible from a ventrolateral angle;53 however, this approach involves drilling into the petrous temporal bone to expose and then mobilize the petrous carotid artery. Furthermore, a transpeduncular approach to the rostral pons has also been described (Fig. 5).24

Within the pons, the descending motor fibers of the corticospinal and corticobulbar tracts lie clustered within the interspersed and surrounding pontine nuclei. These fibers do not rise to the surface at the level of the pons; rather, pontocerebellar fibers arise ventrally and curve dorsolaterally along the ventrolateral surface. A paramedian incision into the ventral pons is therefore generally considered optimal unless the lesion rises to the pial surface elsewhere.31 Indeed, the occurrence of significant or lasting new postoperative deficits has been found by some to be lowest when the location of a CM allowed for this particular anterolateral approach.18 The use of transcranial motor evoked potentials may help further minimize risks to the motor pathways.48 Nonetheless, there are risks to the interspersed motor fibers as well as to corticocerebellar pathways that traverse this region. For instance, there is a report of transient postoperative mutism in a child with a pontine CM resected via this anterolateral approach.19 (Others have reported postoperative mutism after the resection of dorsal mesencephalic CMs, suggesting that the observation of this deficit may reflect 1 of multiple forms of damage possible to cerebellar input/output.60,68)
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Medullary Lesions

Medullary CMs can be resected using the same principles described above. However, the higher density of long tracts and lower CN nuclei within this region makes the resection of deep lesions more treacherous. This region can be approached dorsally from a midline suboccipital craniotomy (with or without a C-1 laminectomy depending on the extent of required exposure) or ventrolaterally from a far lateral suboccipital craniotomy (craniotomy from the transverse sinus superiorly to the ventrolaterally from a far lateral suboccipital craniotomy depending on the extent of required exposure) or craniotomy from the transverse sinus superiorly to the sigmoid sinus laterally and to the ipsilateral occipital cortex from the transverse sinus superiorly to the ventrolaterally from a far lateral suboccipital craniotomy depending on the extent of required exposure).

This region can be approached dorsally from a midline suboccipital craniotomy (with or without a C-1 laminectomy depending on the extent of required exposure) or ventrolaterally from a far lateral suboccipital craniotomy (craniotomy from the transverse sinus superiorly to the sigmoid sinus laterally and to the ipsilateral occipital condyle ventrally).

As is generally the case for operative resection of intrinsic brainstem lesions, new or worsened neurological symptoms will develop in many patients in the immediate postoperative period. Fortunately, experience with brainstem glioma surgery has shown that many of these lower CN deficits will improve with time. There is no wide consensus regarding the safest ways to violate the surface of the medulla, and few surgeons are comfortable doing so if the lesion does not itself contact the surface. Externally salient features of the medulla include the midline sulcus between the protuberances of the fasciculi gracilis, the shallower protuberances of the cuneate fascicles more laterally, the bulges of the inferior olivary nuclei laterally, and the corticospinal tracts with their pyramidal decussation ventrally. Less clearly evident are the spinal trigeminal tract immediately ventrolateral to the cuneate fasciculus below the level of the inferior cerebellar peduncle and the dorsal and ventral spinocerebellar tracts at the lateral margin of the medulla.

Favorably placed dorsal medullary lesions may be resected with the aid of electrophysiological monitoring. Monitoring of lower CN function is believed to be a reliable guide to postoperative function. Cranial nerve IX can be monitored with electrodes in the posterior wall of the pharynx, and one can assess the status of CN XII with electrodes in the tongue. To prevent damage to the nucleus ambiguous and avoid vagal-mediated deficits, particularly dysphagia, evoked movements of the vocal cords can be monitored with electrodes on the surface of the endotracheal tube. Alternatively, the cords can be directly visualized with a laryngoscope; however, it is not yet known whether continuous observation of the spontaneous vocal cord movements accompanying respiration are a reliable sign of intact CN X function.

Ventral lesions are more difficult in terms of both the cranial approach and the details of the local anatomy. Access to the ventral medulla is achieved using the same approaches described for the pons—although a transoral transclival approach to ventrally situated CMs has been described in 2 patients. The general utility and safety of this technique is not yet clear, especially as it relates to the potential incidence of meningitis. Once the ventral medulla is exposed, the choice of an entry point is critical. While it is preferable to avoid entering into the long tracts directly, especially the motor tracts, working ventrally between these tracts is generally not feasible given the presence of exiting nerve rootlets. Therefore, entering directly into lesions that rise to the ventral surface and either displace or damage these tracts is the least likely route to lead to further deficits (motor deficits, if working ventrally; or ataxia, if working more laterally). There is at least 1 case report of entry into the ventrolateral medulla through the inferior olive to resect a superficially located CM without subsequent observation of a new deficit.

The goals of operative intervention, regardless of the lesion location, are complete resection of the CM and preservation of any associated venous anomaly. The former is achieved by entering the lesion to internally decompress it and then working outward toward the margins. Using low rather than high bipolar currents is generally preferred. The incomplete resection of a CM is likely to be a risk factor for recurrence; any residual endothelial cells are believed able to proliferate and recreate the lesion. Indeed, some authors have reported worse overall outcomes when lesions have been only partially resected. However, in contrast to the resection of supratentorial lesions for which a further goal may be to remove the surrounding rim of gliotic tissue, it is important to remain within the vascular portion of a brainstem lesion given the dense opposition of critical structures. Whether this results in the persistence of neurological deficits that could otherwise be alleviated by removing this pathological tissue is not known.

The presence of an associated venous anomaly must be noted to preserve that structure. The experience of many surgeons has shown that these veins drain normal tissue, and their obliteration can lead to venous infarcts. In many cases, these venous anomalies can be preoperatively studied with MR venography or traditional catheter angiography. While venous anomalies are often observed radiographically or intraoperatively in association with a CM, one group has reported a 100% correlation (86 of 86) based on intraoperative assessment.

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Fig. 5. Drawing of approaches to the ventral pons and medulla.
Furthermore, some authors have suggested that the presence of a venous anomaly is mechanistically related to the development of a CM. Therefore, given these considerations, the inspection of all CMs, preoperatively and intraoperatively, for the presence of a nearby venous anomaly is warranted. When resection is undertaken, great care is used in trying to avoid manipulation of the venous anomaly. In such cases, the CM is separated from the venous anomaly as the resection proceeds.

Illustrative Case

History and Examination. A 27-year-old man presented with a 3-day history of double vision on right gaze and symptoms of sinus pressure with frontal headaches. Convergence insufficiency had been diagnosed 1 year earlier for the intermittent blurriness he noticed while looking left. The patient denied any history of trauma or infection, and a review of his systems was nondiagnostic. Evaluation at the outside hospital emergency ward revealed a left internuclear ophthalmoplegia. A noncontrast CT scan of the head and an MR image of the brain demonstrated a left pontine lesion, which did not enhance. A CT angiogram and formal angiogram were obtained, although neither revealed any vascular pathology. The patient was transferred to our institution for further evaluation.

On presentation to our institution, he was noted to have left internuclear ophthalmoplegia, impaired adduction of the left eye, limited abduction of the right eye, and diplopia on right lateral gaze. He was evaluated by the neuroophthalmology service, which confirmed that these findings were consistent with damage to the left intramedullary fascicle of the right sixth nerve and involvement of the left MLF. An MR image of the brain with contrast revealed a T1 and T2 hyperintense nonenhancing lesion in the left upper dorsal pons extending into the left midbrain, notable for a surrounding hemosiderin ring and mass effect as well as displacement of the fourth ventricle without hydrocephalus (Fig. 6 left). The patient elected to undergo resection of the lesion.

Operation. A suboccipital craniotomy with microsurgical removal of the CM through the floor of the fourth ventricle was performed using frameless stereotactic guidance (BrainLAB VectorVision2 navigation system). Complete resection was achieved (Fig. 6 right).

Postoperative Course. Postoperatively, the patient was noted to have a left facial palsy and slight improvement of his diplopia. At the 1-month follow-up, his facial weakness improved such that he was able to completely close his eyes. A neuroophthalmological evaluation during follow-up revealed marked improvement in the diplopia, with mild and slight left fascicular nerve palsy.

Conclusions

A brainstem CM is a potential candidate for operative resection if there is a history of neurological deficit attributable to previous hemorrhage and if the risk of future hemorrhage is unacceptable. The acceptability of risk in any particular case is a function of patient preference, his or her neurological status, and lesion characteristics (size, location, and intrinsic features). Operative resection of brainstem CMs has been demonstrated to reduce the risk of rehemorrhage and can be achieved relatively safely in experienced hands. The possibility of at least transient focal deficits should be anticipated. Once resection is elected, careful consideration of the local anatomy and possible surgical approaches is necessary to obtain the most useful preoperative studies and to arrange for the proper intraoperative monitoring. During the operation, identification and preservation of abnormal venous structures is a goal on par with gross-total resection of the malformation. In the future, as the aggregate experience of neurosurgeons accrues and as surgical technology improves, the range of patients for whom surgery is a viable option to prevent devastating hemorrhage may expand.

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: Asaad, Ogilvy. Acquisition of data: Asaad, Ogilvy. Analysis and interpretation of data: Asaad, Ogilvy. Drafting the article: all authors. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Study supervision: Ogilvy.

References

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10. Campeau NG, Lane JJ: De novo development of a lesion with the appearance of a cavernous malformation adjacent to an existing developmental venous anomaly. *AJNR Am J Neuroradiol* 26:156–159, 2005
45. Porter PJ, Willinsky RA, Harper W, Wallace MC: Cerebral...


65. Thömke F, Hopf HC: Abduction paresis with rostral pontine and/or mesencephalic lesions: pseudoabducens palsy and its relation to the so-called posterior internuclear ophthalmoplegia of Lutz. BMC Neurol 4, 2004


