Cavernous Malformations of the Brainstem

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In 1966, McCormick categorized nonneoplastic vascular malformations of the central nervous system into four types: arteriovenous malformations, venous angiomas, capillary telangiectasias, and cavernous malformations (CM).[23] In the 4 decades since this original classification, significant work has been done to characterize the epidemiology, pathogenesis, natural history, proper management, and prognosis of each of these lesions. Until recently, CMs proved to be the most elusive diagnosis because they are undetectable by angiography. Before magnetic resonance imaging (MRI), a cerebral hemorrhage in a young, normotensive, angionegative patient was assumed to be a cavernous malformation, a diagnosis that could only be confirmed at autopsy. Presently, the diagnosis is often made with the pathognomonic MRI of a CM and the remaining controversy lies in which lesions to treat as well as when and how to treat them.

A subset of these lesions, cavernous malformations of the brainstem (CMB), pose a challenge to neurosurgeons as a result of their location in highly eloquent brain regions, unique presentation, and distinct natural history. In 1934, Walter Dandy was the first to operate on a CMB. Before his pioneering surgery, the brainstem was rarely operated on for fear of extensive morbidity and a lack of developed approaches. Since then, surgical series have described the incidence and course of these lesions while basic science research has provided insight into the genetics and pathogenesis of the disease. CMBs appear to be a distinct subset of CMs and must be given special consideration in planning a course of treatment. With such a high risk of postoperative morbidity and mortality, surgical intervention must be reserved only for lesions that pose a strong risk of significant, lasting neurological injury.

Epidemiology

CMs of the central nervous system occur in 0.1 to 0.9%[9,23,32,33] of the population and make up 8 to 15% of vascular malformations. CMBs account for 9 to 35%[13,16,38] of all CMs. The actual incidence may be lower than rates cited in case series because CMBs are more likely to be symptomatic than supratentorial CMs, leading to an overrepresented presentation. Brainstem lesions show a strong predilection for the pons.[14] In one study of 137 patients with CMBs, 83 were located in the pons, 20 in the midbrain, and 18 in the medulla.[40]

Pathogenesis

CMs are angiographically occult lesions defined pathologically as thin-walled, dilated capillary spaces devoid of intervening neural tissue. There is extensive collagenous hyperplasia and fibrosis present. They are composed of immature proliferative vessels and abnormal endothelial cells that lack tight junctions.[4,42,48] CMs are similar pathologically to telangiectasias in that they do not contain elastic fibers or smooth muscle.[35] Lesions are characteristically surrounded by a ring of hemosiderin deposits, providing evidence of ongoing microscopic hemorrhages. Except in rare circumstances, CMs are angiographically negative because flow through these lesions is insufficient to be detected by conventional angiography.

Secondary to observations of family clusters of CMs, it has long been suspected that at least a subset of these lesions is inherited. CMs are now characterized as either sporadic or inherited. The inherited form is transmitted as an autosomal-dominant trait and is thought to occur through a "two-hit" model, similar to retinoblastoma, in which an individual receives one normal allele and one abnormal allele and then acquires mutations in the normal allele throughout life to express the disease phenotype.[8] Three genes have been identified through linkage studies in patients with familial CMs: CCM1 (40–50%), CCM2 (10–20%), and CCM3 (40%)[24] (*Table 10.1*). CCM1 is located on 7q and encodes a protein known as KRIT1 whose function is currently unknown.[47] Familial CM is more common in Hispanic populations and is often the result of the same mutation in CCM1, which indicates the existence of a single ancestor.[8] CCM2 resides on 7p and encodes a protein titled MCD4607, better known as malcaverin, also without a known function.[6] It has recently been discovered, however, that KRIT1 interacts with both a protein called integrin cytoplasmic domain-associated protein-1α[46] and malcaverin itself.[47] CCM3 is located on 3q and encodes a protein called programmed cell death 10, which is known to be involved in apoptosis.[2]

It was previously believed that CMs are static, congenital lesions, but recent work has noted that these lesions are likely to be dynamic.[5] Case reports have described de novo

0148-703/08/5501-0088

TABLE 10.1. Genes associated with cavernous malformations[a]

Gene	Incidence[b]	Location	Product	Function
CCM1	40–50%	7q	KRIT1	Binds icap1α
CCM2	10–20%	7p	MGC4607 (malcaverin)	Binds krit1
CCM3	40%	3q	Programmed cell death 10	Apoptosis

[a]CM, cavernous malformation; icap1α, integrin cytoplasmic domain-associated protein-1α.
[b]Among familial cases of CM.

formation,[17,29] enlargement, regression, and even a dramatic decrease in size.[43] Although they are widely believed to be benign lesions, there are some reports that suggest a neoplastic basis for CMs. The endothelial cells appear microscopically atypical, they express proliferating cell nuclear antigen,[26] there are multiple reports of lesions occurring in previously irradiated areas, there is a report of a cavernous hemangioma seemingly growing in the biopsy track of a different lesion,[5,27] and the two-hit hypothesis behind inherited CMs is very similar to the genetics underlying multiple cancer syndromes such as retinoblastoma. More research is necessary to clarify the biology behind these lesions and many more discoveries are likely to occur in the near future to elucidate the genetics and molecular biology underlying CMs.

Presentation

CMBs often present differently from supratentorial or cerebellar CMs, making it important to recognize these lesions as a discrete subset of CMs. The most commonly encountered presenting symptom of a cerebral CM is seizure occurring in approximately 60% of patients.[16] Other presenting symptoms include progressive neurologic deficit (50%), hemorrhage (20%), and hydrocephalus. CMBs never present with seizures and unlike supratentorial CMs, even microhemorrhages in CMBs are likely to produce neurological symptoms. Whereas the annual bleeding risk for supratentorial CMs is approximately 0.5% per year for unruptured lesions and 5% per year for lesions that have already bled,[9,33] CMBs are assumed to have an annual risk of hemorrhage of 2 to 6%[37] and an annual rebleeding risk of up to 6 to 35%.[12] Other factors that have been shown to increase risk of hemorrhage in CMs include being male, having a family history of CM, and having a venous anomaly associated with the lesion.[20,34] Hemorrhage from a CMB is more likely to be symptomatic than hemorrhage from a cerebral CM. As well, lack of a universally used definition of hemorrhage leads to a variety of interpretations and a range of values in the literature when attempting to pinpoint bleeding and rebleeding rates.[45] Although CMBs pose a moderately increased risk of hemorrhage and rebleeding compared with CMs, criteria for surgical intervention are far more strict as a result of the high morbidity and mortality of surgical manipulation of the brainstem.

Evaluation

As previously discussed, the CM posed a diagnostic dilemma until very recently when the development of MRI made it possible to make an early, often definitive diagnosis. Even with the aid of angiography, diagnosis had been difficult because such a large percentage of these lesions is angiographically silent. Most often, patients with a CMB present with symptoms of hemorrhage, which manifest in one or more neurological deficits depending on the location of the lesion. Patients often first receive a computed tomography scan, which will miss many small and some large lesions. Signs of hemorrhage on the computed tomography may incur suspicion of a vascular lesion, thereby prompting an angiogram, which will often be negative.[1,21] If a CM is suspected, a T2-weighted MRI is the appropriate test. The image of a CM on an MRI is nearly pathognomonic, and often no further imaging is needed to determine the diagnosis. The image is described as showing a "mulberry-like" reticulated core of mixed signal intensity surrounded by a rim of decreased signal intensity, representing the continuous hemosiderin rim.[14,31] It is important to check rigorously for multiple lesions, which is not uncommon, especially in Hispanic patients and patients with a family history of CMs.[21]

After a CMB is diagnosed, the most important imaging concern relates to whether the lesion is operable. Anatomic criteria that come into play, and therefore must be evaluated radiologically, involve exactly where the lesion is located in the brainstem and how close it is to the pial surface. Poor outcome in surgical treatment of CMBs has been directly correlated with the parenchymal thickness between a CMB and the pia. Often, this distance is evaluated on a T2-weighted MRI, but recently it has been suggested that three-dimensional constructive interference in steady-state imaging allows more accurate visualization of the intervening parenchyma between a CM and the pia.[45]

Management

Management of CMBs poses important concerns resulting from the subacute nature of the lesion and the complexity of the brainstem. A number of small case series combined with some recent larger series illuminate our understanding of CMBs' natural history, presentation, and management-dependent out-

TABLE 10.2. Surgical series of cavernous malformations of the brainstem[a,b]

First Author	Year	Number of Patients		
		Total	Surgically Treated	GKS-Treated
Drake	1986	14		
Weil	1990	7		
Bertalanffy	1991	15	13	
Fahlbusch	1991	20	10	
Symon	1991	7		
Zimmerman	**1991**	**24**	**16**	
Fritschi	1994	41		
Zabramski	1994	21		
Sathi	1996	50	23	
Amin-Hanjani	1998	14	14	
Cantore	1999	12		
Porter	**1999**	**100**	**86**	
Steinberg	2000	42		
Vinas	**2002**	**8**	**8**	
Wang	**2003**	**137**	**137**	
Mathiesen	2003	68		
Ferroli	**2005**	**52**	**52**	
Kim	2005	6		6
Liu	2005	49		49
Liscak	2005	33		33
Bruneau	2006	22	22	
Huang	2006	7		7
Taplin	2006	7	7	
Zausinger	2006	13	13	

[a]GKS, gamma knife surgery; CMBs, cavernous malformations of the brainstem.

[b]Studies in bold focused specifically on CMBs. In other studies, CMBs made up a subset of the cavernomas studied.

come (*Table 10.2*). Various case series have reported that approximately 50% of patients present with acute hemorrhage and 40% present with subacute hemorrhage (>6 weeks).[13,29] With such a large number of patients presenting subacutely, surgical removal of the lesion does not need to occur immediately. There has been some controversy in the literature regarding adequate timing for surgery; with some authors favoring early intervention, whereas others have justified delayed treatment, reporting that outcome is independent of surgical timing.[22,36,40] As a result of high rates of postoperative morbidity and the complexity of the procedure, many authors argue for conservative management.[18] One study followed patients with CMBs who did not receive surgical treatment and observed a surprisingly low morbidity rate of 8% over 5 years.[18] Many surgeons, however, still opt to operate soon after presentation, but not necessarily emergently.

At this point in time, most neurosurgeons argue that microsurgery rather than gamma knife surgery (GKS) is clearly the answer in treating CMs, predominantly as a result of lack of evidence that stereotactic radiosurgery is efficacious. In 2005, three case series were published describing a total of 279 patients, of which 88 had CMBs.[15,19,20] The largest of these studies, by Liu et al., reported 125 cases of CM treated with GKS, including 49 patients with a CMB.[20] They reported an overall post-GKS annual rebleeding rate of 6.5% and a postoperative seizure rate of approximately 50%. These results suggest that GKS may play a role in supplementing or delaying microsurgery in certain inoperable cases, but it does not improve the hemorrhage rate significantly enough to justify foregoing microsurgery. In addition, long-term outcome studies are necessary for further evaluation of this potentially useful therapy.

If microsurgical resection is performed, complete resection of the lesion is of the utmost importance, underscored by two case reports in the literature describing fatal postoperative hemorrhage.[3,11] Other reports have correlated worse outcome with patients who have a subtotal resection of their CMB.[22]

Review of the medical literature clearly supports a conservative approach to asymptomatic CMBs as a result of their dangerous location for surgery and relatively low risk of morbidity. Clinically symptomatic lesions, however, pose a more difficult management dilemma. In the vast majority of cases, symptomatic hemorrhages from CMBs produce only a transient neurological disability. Satisfactory recovery is the rule if recurrent hemorrhages do not occur. Recurrent hemorrhages occur in a minority of patients with CMBs. Therefore, surgical series involving acute surgical resection after primary symptomatic hemorrhage subject some patients to the unnecessary peril of open brainstem surgery. Although some patients are described as unchanged or slightly improved after surgical resection, some of these surgical patients may have made better or even complete recovery if treated conservatively.

Our policy has been to never operate on a CMB after a single hemorrhage. The indication for surgery of CMBs is to prevent rebleeding. Therefore, only lesions that have declared themselves as high risk are suitable for surgical intervention. High risk implies multiple episodes of symptomatic bleeding with each episode producing increasing neurological disability.

We now present three cases recently encountered at our hospital that exemplify the major management decision regarding conservative therapy versus surgical management.

Case 1

At the time of presentation to our hospital, the patient was a 36-year-old woman who was 20 weeks pregnant. She reported that she had no medical problems other than prior

trauma to her face for which she underwent surgery on her mandible. Her symptoms had begun 6 years prior when she suddenly noticed diplopia and presented to the emergency room of her local hospital. An MRI was performed and she was found to have a sixth nerve palsy from a CMB in the dorsal pons. She gradually improved and after a period of 2 months, only noticed double vision if she looked to the right. Two years later, she experienced a recurrence of diplopia, which again recovered after 2 months.

Approximately 18 months later, she experienced a third episode that consisted of difficulty hearing on the right side, balance problems, and numbness in her left arm. An MRI showed hemorrhage of the CMB. These symptoms again resolved over a few months' period and she was well for another year. Two months before presentation at our hospital, she had become pregnant and experienced a fourth episode that included diplopia, right facial weakness, and weakness and numbness of the left side of the body. An MRI was performed that showed a 2.5-cm CMB in the dorsal pons that had erupted into the floor of the fourth ventricle (*Fig. 10.1*).

Her examination on presentation revealed a near complete right facial palsy, a right sixth nerve palsy, a subtle limp, and mild left hemiataxia. At this point, the CMB had presented to the surface of the brainstem, demonstrated a propensity for rebleeding, and caused debilitating neurological deficits. Surgical excision was therefore discussed with the patient. As a result of an increased risk of complications during pregnancy and a low probability of rebleeding over the subsequent 6 months, it was decided that she would be re-evaluated after her pregnancy. When evaluated 11 weeks after giving birth, she had experienced complete resolution of her symptoms except for diplopia when looking in the distance. She again had an MRI, which showed that the CMB had shrunk and no longer abutted the floor of the fourth ventricle (*Fig. 10.2*). The CMB was no longer exophytic and the patient now had no significant neurological deficits. She was managed conservatively with a plan to follow-up on an annual basis.

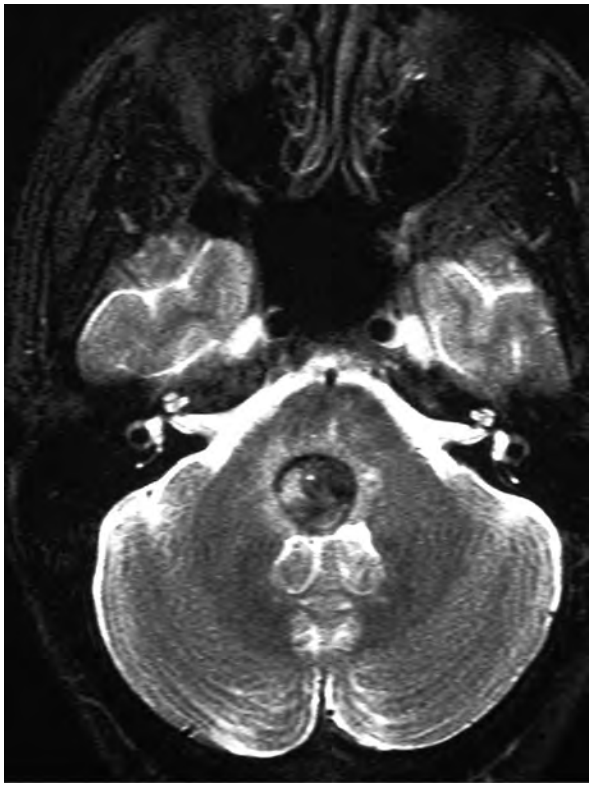

FIGURE 10.1. An axial T2-weighted image shows a 1.8-cm lesion at the pontomedullary junction with surrounding edema, making contact with the fourth ventricle.

Case 2

On presentation, the patient was a 43-year-old woman with no prior medical problems. Eighteen months previously, she began experiencing pain in the back of her head and neck. An MRI of her cervical spine was performed to rule out cervical spine disease. Although her cervical spine was normal, she was found to have a brainstem hematoma in the left pons. The lesion was followed conservatively until 3 weeks before presentation when she suddenly experienced mild left facial numbness. A repeat MRI revealed enlargement of the hematoma (*Fig. 10.3*). Two days before presentation to this hospital, she experienced increased facial numbness and tingling throughout the right side of her body. Her symptoms rapidly evolved over 48 hours to include mild right hemiparesis, right-sided dysmetria, left-sided hemiataxia, a left-sided sixth nerve palsy, and a left-sided seventh nerve palsy.

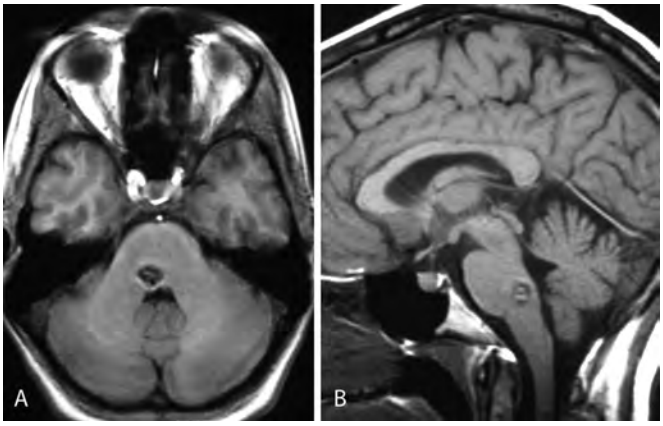

FIGURE 10.2. An axial T2 fluid-attenuated inversion recovery (*A*) and a sagittal T1-weighted image (*B*) show the pontomedullary CMB significantly reduced in size, making only slight contact with the fourth ventricle and showing less surrounding edema than in previous images.

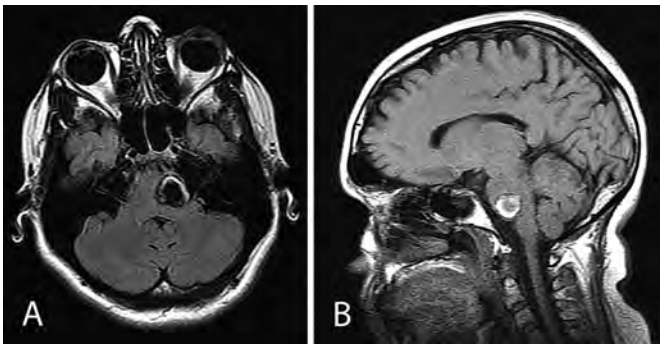

FIGURE 10.3. Axial (*A*) and sagittal (*B*) T2 fluid-attenuated inversion recovery images show a moderately sized CMB at the pontomedullary junction on the left making contact with the surface of the brainstem. A ring of hemosiderin deposits is visible surrounding the lesion.

She was then transferred to our institution. An MRI was performed that showed further expansion of the brainstem hematoma with a significant exophytic component emerging from the left pons displacing the seventh and eighth cranial nerves (*Fig. 10.4*). As a result of the occurrence of multiple bleeding episodes, the progression of the lesion with significant neurological deficits, and accessibility at the surface of the left pons, the patient was taken to the operating room.

The surgery was performed with a left suboccipital craniotomy and a retrosigmoid approach to access the cerebellopontine angle and dorsal pons where the lesion was found to be exophytic. The transverse and sigmoid sinuses were located using the Stealth Navigation Stereotactic System (Medtronic, Louisville, Colorado). Evoked brainstem potentials were used throughout the operation to evaluate brainstem function. On visualization, the seventh and eighth cranial nerves were draped over the lesion. The intraparenchymal hematoma was evacuated and the remaining components of the lesion removed. The lesion consisted of abnormal vascular components and small black areas, highly suggestive of a CM.

Postoperatively, the patient's neurological status significantly improved with increased strength throughout her right side, increased strength in the left side of her face, decreased ataxia, and resolution of her gaze palsy and hearing deficit. One month after surgery, she continued to experience right-sided weakness and left facial weakness with full recovery of her ataxia. At 3 months after the operation, she continued to improve in strength on the right side of her body and left face. An MRI performed at that time revealed complete resection of the CM with no residual enhancement (*Fig. 10.5*). Considerable scarring and hemosiderin deposition remained in a 1.5-cm lesion, significantly reduced in size from the preoperative 5-cm lesion. At follow-up 1 year later, the patient had experienced no new symptoms, but the mild right hemiparesis and left facial weakness had not resolved and were considered at this time to be permanent.

Case 3

At the time of presentation, the patient was a 44-year-old man in good health with new-onset diplopia. An MRI was performed and he was found to have a cavernous malformation of the quadrigeminal plate. At the time, he was treated with stereotactic-guided radiation. Since the operation, follow-up imaging has shown a gradual but progressive increase in the size of the lesion. One month before admission, the patient noticed ptosis, worsening balance, and retrospectively noted a 5-month history of slurred speech. Physical examination revealed a left third nerve palsy and mild dysarthria.

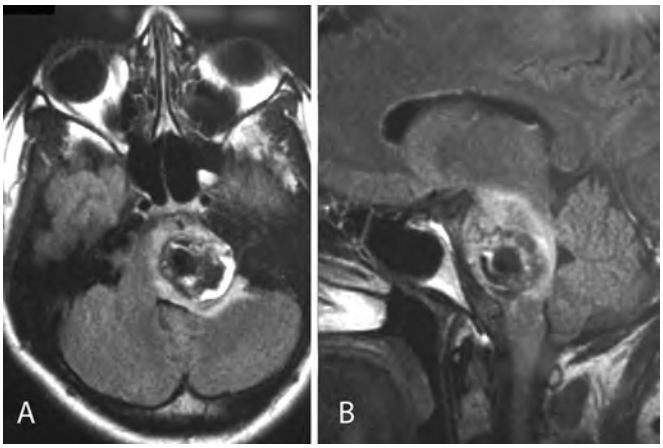

FIGURE 10.4. Preoperative axial (*A*) and sagittal (*B*) T2 fluid-attenuated inversion recovery images show an enlarged, heterogenous, exophytic lesion with significant surrounding edema.

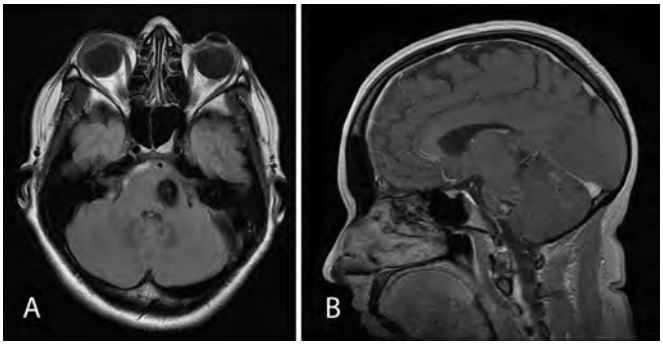

FIGURE 10.5. Three months postoperatively, an axial T2 fluid-attenuated inversion recovery image (*A*) shows a significantly reduced, homogenous, hypointense lesion that no longer abuts the surface of the brainstem. A sagittal T1-weighted image with contrast (*B*) shows no residual enhancement.

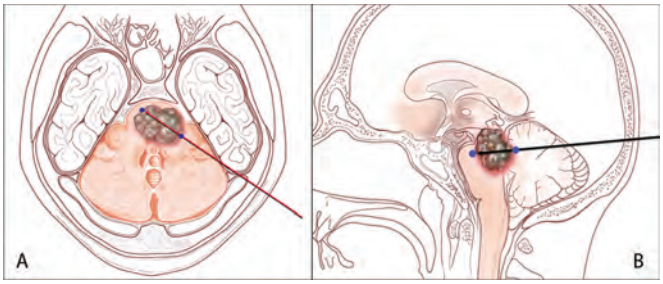

FIGURE 10.6. The "two-point" technique is demonstrated in these axial (*A*) and sagittal (*B*) illustrations.

As a result of the presence of serious neurological dysfunction and rapid progression of the patient's neurological symptoms, surgical resection was considered. A current MRI was used to evaluate the accessibility of the lesion and most appropriate surgical approach using the "two-point" technique (*Fig. 10.6*). This technique consists of drawing a straight line through the lesion from the most exophytic portion to the deepest portion. If the lesion is exophytic meaning that the line does not pass throughout any brainstem tissue before reaching the lesion, then the lesion is accessible and able to be resected. In addition, the best surgical approach is the approach that best approximates the line drawn. In this case, an occipital craniotomy was performed with a transtentorial approach to the dorsal midbrain.

DISCUSSION

In the first example case, the patient initially sustains severe neurological disability and presents with a surgically accessible lesion. Her pregnancy, however, delayed surgery and at re-evaluation her CMB had reduced in size, making her no longer a candidate for surgery. Her case illustrates the principle of conservative management when the risks of surgery outweigh the current neurological deficits and future risk of hemorrhage. As a result of the low risk of rebleeding in these lesions, any time a CMB is not accessible from the surface of the brainstem, the risks of surgery exceed the risks of conservative management. The second case illustrates the importance of surgery if a lesion is accessible from the surface and if a patient experiences multiple bleeds with significant progressive neurological deficits. After surgery, this patient sustained permanent neurologic damage that was, however, an improvement from the deficits she experienced after hemorrhage of the CMB. The third case illustrates the "two-point" technique for assessing the accessibility of a lesion and determining the most appropriate approach.

Operative Technique

Intraoperative Neuronavigation and Neurophysiological Monitoring

As a result of the dense functional anatomy of the brainstem, it is vital for the surgeon to plan a precise surgical approach to the malformation and associated hematoma, two crucial operative technologies that are extremely helpful in safe surgical excision of CMBs are neuronavigation and neurophysiological monitoring. Frameless stereotactic guidance is absolutely necessary for lesions that have not ruptured through the pial surface and therefore the brainstem surface may appear normal when visualized under the microscope.[45] In these cases, the surgeon can use MRI guidance to identify the safest route to a lesion with the least amount of brainstem injury.

Another modality that significantly aids the surgeon intraoperatively is neurophysiological monitoring. This modality makes use of evoked somatosensory potentials, evoked brainstem potentials, and cranial nerve stimulation to safeguard central nervous system function in real time during the operation.[30] Although accessing and manipulating the lesion, the surgeon can monitor effects on essential brain structures, allowing him to modify his technique accordingly to avoid damage to essential structures.

Approaches

There are five major approaches to the brainstem: posterior, posterolateral, lateral, anterior, and anterolateral[10,11,39–41,44,50] (*Table 10.3*). The principal factor determining which approach to take is the location of lesion. Ideally, the surgeon should select an operative approach that visualizes the pial point that is closest to the surface of the hematoma cavity and allows a perpendicular "line of sight" approach from that point to the deepest part of the lesion. The

TABLE 10.3. Surgical approaches to the brainstem

Approach	Lesion Location
Posterior	
Supracerebellar/Infratentorial	Dorsal midbrain
Occipital interhemispheric transtentorial	Dorsal midbrain
Midline transvermian	Floor of the fourth ventricle
Subtonsillar	Medulla
Posterolateral	
Retrosigmoid	Cerebellar–pontine angle
Combined subtemporal/suboccipital presigmoid	Lateral midbrain and pons
Far lateral approach	Anterior–lateral medulla and pons
Lateral	
Subtemporal	Lateral midbrain and upper pons
Subtemporal transpetrosal	Lower lateral pons
Anterior/anterolateral	
Pterional, orbitozygomatic osteotomy	Ventral midbrain and upper pons

major exception to this principle would be with lesions that lie below the fourth ventricular surface. An approach through the floor of the ventricle should be selected only in cases in which it is absolutely clear on preoperative imaging that the hematoma cavity has completely dissected to the floor of the ventricle and there is some point where no overlying functional tissue prevents direct entry into the hematoma cavity.

Over the past decades, numerous approaches have been designed to access the brainstem, which remains one of the most challenging brain areas to treat surgically. These operations are difficult and must be carried out by a surgeon, who is highly trained in accessing and manipulating the brainstem and related structures.

The posterior approach to dorsal midbrain lesions is best through an occipital interhemispheric transtentorial approach. Small truly exophytic lesions can be resected through a supracerebellar infratentorial approach. For lesions in the floor of the fourth ventricle and dorsal medulla, a midline suboccipital craniotomy and transvermian approach is the most direct route. Lesions in the dorsal medulla can usually be accessed with minimal or no vermian transection merely by superior or lateral retraction of one or both cerebellar tonsils.

From the posterolateral direction, mostly for pontine lesions in the cerebellar–pontine angle, the surgeon can take a retrosigmoid approach for superficial lesions. Deeper lesions in the pons or lateral midbrain require a more lateral approach as achieved through a combined subtemporal and suboccipital presigmoid approach. Anterolateral lesions of the medulla and lower pons can also be accessed through the far lateral transcondylar approach.

The anterolateral approach to the midbrain and upper pons is through some variation of the pterional craniotomy. The pterional craniotomy along with an orbitozygomatic osteotomy and wide Sylvian fissure split provides exposure to the anterior aspect of the midbrain and upper pons. More lateral lesions may be accessed through a subtemporal approach with splitting of the tentorium or even partial resection of the petrous apex (Kawase approach). At this point, it seems that transoral approaches to the ventral medulla and pons are contraindicated because of the high associated morbidity of those approaches for intradural lesions.

Prognosis

CMBs carry a higher morbidity and mortality than supratentorial or cerebellar CMs.[25] Despite difficult surgical approaches into highly eloquent brain regions, reported mortality remains low. Samii et al.[36] and Wang et al.[40] each reported a mortality rate of 0%, whereas Porter et al. reported 4% in their series of 100 patients.[29] Rates of long-term morbidity have been reported as high as 70%,[3] but recent, larger series cite rates of 11[40] and 12%.[29] Immediate postoperative morbidity is naturally two to three times higher[29] as a result of complications related to cranial base surgery that resolve within days to weeks such as pneumonia, cerebrospinal fluid leak, wound infection, and others.

Lesions have been described as intrinsic (with parenchyma between the lesion and pial surface) or exophytic (abutting the pia) with intrinsic lesions carrying higher rates of postoperative injury. These intrinsic CMs are more often treated nonsurgically. Reports of CMBs treated conservatively describe high rates of disability. Fritschi et al.[13] described 30 conservatively treated patients, reporting a 36-month rate of moderate to severe disability or death at 34%, whereas Porter et al.[29] reported 42% in their 14 nonoperative patients. The natural clinical course of these lesions is often described as progressive neurological decline, but many authors have described sudden, debilitating hemorrhage.[1,7,23,28,49] Our own experience with these lesions, however, shows that most follow a relatively benign course. Although brainstem malformations may cause a relapsing and remitting clinical course, most patients, even with severe brainstem problems, make an excellent recovery without surgery. Most CMBs hemorrhage in a symptomatic fashion only once or twice before becoming quiescent. It is only the malformations that bleed repeatedly causing progressive neurological decline that require surgical resection. Our data are in line with Kupersmith et al., who observed approximately only 8% morbidity over 5 years in conservatively treated patients with a CMB.[18]

CONCLUSION

CMBs pose a unique challenge to the neurosurgeon because of their distinctive course and dangerous location in highly eloquent brain regions. They present differently from supratentorial CMs, carry different rates of hemorrhage, and often require highly challenging approaches for excision. As a result of the high risks of open brainstem surgery, surgical intervention should be reserved for cases that show multiple hemorrhages, progressive and significant neurological deficits, and present to the surface of the brainstem for surgical access. Improvements in molecular biology, imaging, and surgical technique will continue to aid in better understanding and treatment of this lesion.

Disclosure

The authors did not receive financial support in conjunction with the generation of this article. The authors have no personal or institutional financial interest in the drugs, materials, or devices described in this article.

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