Modern Management of Brainstem Cavernous Malformations

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- Cavernous malformations • Brainstem • Surgical approaches
- Radiosurgery • Neuronavigation
- Neurophysiological monitoring

Once considered terra incognita, the brainstem has been more commonly and successfully operated on in the past 2 decades.[1] About 20% of cerebral cavernous malformations (CMs) occur in the brainstem, leading to a prevalence of about 1 in 200 in the general population.[2] In 1990 and 1991, case series of 11,[3] 10,[4] and 16[5] surgically treated brainstem cavernous malformations (BCMs), also called brainstem cavernomas, were published. A decade later and in recent years, case series of more than 100 patients have been reported.[6–8] Much of this evolution has mirrored the increasing use and widespread acceptance of magnetic resonance imaging (MRI) as the mainstay of brain imaging. Computed tomography (CT), previously one of the main imaging modalities for the brain, was hampered by poor resolution and beam-hardening artifacts in the posterior fossa.[9] Angiography was (and remains) of little use because BCMs are typically not visualized. Before the popularization of MRI, these angiographically occult entities were frequently misdiagnosed as neoplastic, demyelinating, or occlusive vascular lesions.[10]

Fundamental to any discussion of cerebrovascular disease is an intimate understanding of the natural history risks. For cerebral CMs in general, these data are less well characterized and more controversial than for arteriovenous malformations (AVMs), aneurysms, and extracranial carotid artery disease, all of which have well-designed large studies providing reliable guidelines for annual risk.[11–15] It is against these natural history risks that the risk of treatment of any neurovascular lesion must be weighed. For the rare subset of BCMs, the natural history risks are even less well understood than CMs in general. At the same time, this information is

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more important considering the risks of treating lesions in this most eloquent of brain tissue.

More than 50 surgical series regarding BCMs have been published,[16] and this collective experience underscores several well-accepted principles in neurosurgical circles. The most important is patient selection, which is largely based on ensuring the aggressive nature of an individual lesion. Second, anatomic accessibility within the complex structure of the brainstem is paramount. Radiosurgical series present conflicting data on the treatment of BCMs.[16] Hemorrhage clustering may contribute to the favorable results noted in some radiosurgical series. Surgical and radiosurgical treatments carry significant risks of permanent morbidity and disability.

CMs of the brainstem in particular, once considered untreatable, are now characterized better radiographically with modern imaging techniques; are resected more safely with advances in intraoperative monitoring, frameless stereotactic neuronavigation, and the refinement of skull base techniques; and are increasingly being treated with radiosurgery with uncertain efficacy.

NATURAL HISTORY

Although the article covers the management of BCMs, as with other cerebrovascular diseases, management depends greatly on the interpretation of the natural history risks for any given patient. Natural history studies vary in their reported annual hemorrhage rates for BCMs. Variation can be explained by the relative rarity of the disease, selection bias and referral patterns, differing definitions of hemorrhage (intralesional vs extralesional, which is often difficult to discern), use of symptomatic and asymptomatic bleeding, hemorrhage clustering, and initial versus rebleeding rates. Knowledge of the natural history of BCMs is more ambiguous than that of other vascular malformations.

In 2001, Kupersmith and colleagues[17] published a study on a series of 37 patients followed for a mean of 4.9 years. In their series, low rates of bleeding (2.46%/yr) and rebleeding (5.1%/yr) were reported, with low rates of neurologic complications related to hemorrhage. Some investigators have noted serious flaws in this report with respect to selection bias (a presumably relatively benign subgroup of patients referred to a neuro-ophthalmologist for follow-up monitoring) and the retrospective study design.[8,18] Even so, certainly some BCMs appear to have a benign course and warrant conservative management.[19,20]

Numerous other reports have suggested a more aggressive and devastating natural history course for BCMs, especially when considering rebleeding rates and neurologic event rates. Looking at the largest of all of the series of surgically treated BCMs (with >30 treated), the rates of annual hemorrhage range from 21% to 60%.[6,7,21–23] While surgical series may also represent selection bias, prospective natural history studies have confirmed significant bleed and rebleed rates.[24] Clearly, the manifestations of brainstem hemorrhage are clinically and functionally significant. Cranial nerve deficits, sensory deficits, and motor weakness are all common complaints, occurring alongside ataxia, alterations in consciousness, hydrocephalus, and certain pain syndromes.[7,8,23] Similar to cerebral aneurysms, the female gender confers a greater natural history risk, whereas, unlike with aneurysms, size is irrelevant.

MICROSURGERY

Surgical treatment of BCMs famously and inadvertently began when Dandy[25] evacuated an upper brainstem hematoma attributed to a CM from a 31-year-old man. At