Intramedullary spinal cord cavernous malformations in the pediatric population

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INTRODUCTION

Intramedullary spinal cavernous malformations (ISCM) make up only 1% of all intramedullary pediatric spinal cord lesions. Children most typically present with acute neurological deterioration characterized by the acute onset of severe motor deficits. Of interest, spinal cord ISCM lesions are considered more aggressive than those occurring intracranially due to the limited space within the spinal cord. Nevertheless, if recognized and treated in a timely fashion, ISCMs have been shown to have favorable outcomes, when compared to the adult population.
Unfortunately, the misdiagnosis rate for ISCM is estimated to be as high as 55%, as the clinical presentation is too frequently misinterpreted as consistent with cerebral palsy, syringomyelia, and transverse myelitis. Here, we present the clinical, radiologic, and surgical features of pediatric ISCM, along with a discussion of the relevant literature.

**PATHOLOGY**

Cavernous malformations are well-circumscribed vascular malformations that are dark blue or reddish-brown. They can often be seen coming to the spinal cord surface. They consist of sinusoidal vascular spaces lined by a single layer of endothelial cells encompassed by a loose connective tissue stroma. As these cavernous malformation lack normal vessel wall components, they are predisposed to hemorrhaging.

**APPROPRIATE DIAGNOSTIC WORK-UP**

Spinal cavernous malformations often occur in association with multiple cerebral or systemic lesions; therefore, patients should undergo full neuroaxis and complete-body imaging. There is a high incidence of these lesions reported in the Hispanic population likely due to an autosomal dominant alteration of chromosome 7q. However, this increased incidence has not been demonstrated in the pediatric population. Further, de novo development of ISCM has been reported following spinal radiation and trauma.

**DIAGNOSIS AND DIAGNOSTIC CHALLENGES**

Magnetic resonance imaging (MRI) is the diagnostic modality of choice as these spinal lesions tend to be occult on angiographic imaging. On MRI, they are well-circumscribed and multilobulated with heterogeneous intralesional T1 and T2 signals reflecting a reticulated core. Their “popcorn-like” appearance is attributed to the presence of blood products in different phases of evolution (e.g., best seen on gradient-echo images). Typically, T2-weighted images reveal a hypointense rim surrounding these lesions, representing hemosiderin. Calcifications can occur in an estimated 34% of patients. Contrast enhancement is usually minimal. Susceptibility-weighted imaging has extremely high sensitivity for even small amounts of blood products and can also aid in the diagnosis of these lesions. Adjacent intramedullary hemorrhage is also frequently present, often eccentric, and bidirectional.

**DIFFERENTIAL DIAGNOSES**

Differential diagnoses for ISCM include intramedullary tumors (particularly hemangioblastomas) or arteriovenous malformations; angiography may rule out the latter. Misdiagnosis or a failure to diagnose ISCM can result in delays in treatment, and risk the sequelae of re-bleeding, especially in pediatric patients.

**HEMORRHAGE AND REHOMORRHAGE RATES**

The annual hemorrhage rate for spinal cavernous malformations in both adult and pediatric patients ranges from 1.4% to 6.8%; the re-hemorrhage rate is 2%. Better outcomes are reported for patients undergoing resection versus conservative management. Ren et al. reviewed a series of 20 pediatric patients with ISCMs and demonstrated that in spite of ISCCM prevalence in the pediatric population being relatively low (7.9%), the rate of hemorrhage (8.2%) and re-hemorrhage (30.7%) is higher than in their adult counterparts. Notably, repeat hemorrhages correlated with more severe permanent neurologic disability.

**NEUROLOGICAL SEQUELAE AND SYMPTOMS**

Neurological sequelae of intramedullary cavernous malformations in the pediatric population is highly variable. Ren et al. looked at pediatric patients with ISCMs and categorized symptomatic presentations to include pain, weakness, sensory deficits, and bowel/bladder dysfunctions; deficits ranged from ASIA scale grades A-D on admission. Badhiwala et al. did not show a statistically significant relationship between ISCM size and neurological outcome. Their results suggested that while ISCM size is important to consider, location was a far better determinant of presentation and outcome. They also demonstrated that symptomatic patients undergoing surgery within <3 months after their initial presentation displayed statistically significant improvements in neurological outcomes versus those having surgery more than 3 months later.

**OPERATIVE TECHNIQUES**

Pin-pointing the location of ISCM in the cord (ventrally, dorsally, etc.) is critical to operative planning. Surgical is typically indicated by the presence of unremitting and/or progressive neurological symptoms/signs, or for lesions larger >5 mm in diameter, and/or growing. Expedited surgery is particularly indicated in the pediatric population to reduce the risk of re-hemorrhage/long-term sequelae. While most spinal cavernomas are found in the cervical spine, they do occur in the thoracic and lumbar levels. Typically, prone positioning is preferred for a posterior approach to the spine, while supine positioning is preferred for an anterior approach. If fusion is required in the thoracolumbar spine, then hip extension may be used to increase lordosis. A lumbar drain can be placed to prevent cerebrospinal fluid leak and pseudomeningocele formation and is typically left in for 1–2 days postoperatively. The surgical resection should be limited the extent of myelotomy to maximize white matter.
preservation and postoperative deficits. Further, all surgical approaches to the spine (e.g., anterior, anterolateral, posterior, and posterolateral), including minimally invasive approaches, have their own unique respective risks and complications.

Intraoperative neuromonitoring, utilizing electromyography, electroencephalography, somatosensory evoked potentials, and motor evoked potentials are a critical adjunct to limit postoperative neurological deficits for the patient undergoing ISCM surgery.[19]

POSTOPERATIVE OUTCOMES

The surgical removal of ISCM in the pediatric population may provide adequate functional outcomes. Although most patients remained unchanged from their preoperative status, over 50% improve over a mean follow-up duration of 4 years.[11] Notably, the preoperative neurological status is the best indicator of clinical outcome, and the more symptomatic the patients are preoperatively (e.g., with delayed treatments) the poorer the outcomes; notably, worsening after surgery occurs in 10% of pediatric patients.[16] Nevertheless, as the annual rate of hemorrhages and re-hemorrhages is higher in this age group, early complete obliteration is preferred to partial endovascular treatment and/or other incomplete management strategies.[11]

CONCLUSION

Rates of hemorrhage and re-hemorrhage are higher in pediatric patients with ISCMs and, thus, should prompt early surgery and complete lesion resections where feasible. Extensive meta-analyses specific to the pediatric population are needed to examine the relationship between specific neurologic sequelae and neuroradiological findings to ultimately decrease the rate of misdiagnosis and hemorrhage rates.

Declaration of patient consent

Patient’s consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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