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Review Article

Intramedullary spinal cord cavernous malformations in the pediatric population

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Received : 04 August 2020
Accepted : 22 August 2020
Published : 05 September 2020

DOI
10.25259/SNI_494_2020

Quick Response Code:

ABSTRACT

Background: Intramedullary spinal cavernous malformations (ISCM) account for just 1% of all intramedullary pediatric spinal cord lesions. Pathologically, they are well-circumscribed vascular malformations that typically appear dark blue or reddish-brown, often coming to the spinal cord surface. With regard to the histopathology findings, ISCMs are comprised sinusoidal vascular spaces lined by a single layer of endothelial cells within a loose connective tissue stroma. As these lesions are often misdiagnosed in the pediatric population, appropriate treatment may be unduly delayed.

Methods: The authors performed an extensive review of the published literature (PubMed) focusing on ISCM in the pediatric age group.

Results: The search yielded 17 articles exclusively pertaining to ISCM affecting the pediatric population.

Conclusion: Here, we reviewed the clinical, radiographic, surgical, and outcome data for the treatment of ISCM in the pediatric age groups. Notably, over 50% of pediatric patients with ISCM experienced an improvement in their neurological status after a mean postoperative follow-up duration of 4 years. Future meta-analyses are needed to highlight the potential presence of ISCM and, thereby, decrease the rate of misdiagnosis of these lesions in the pediatric population presenting with recurrent intramedullary spinal cord hemorrhages.

Keywords: Cavernoma, Cavernous malformation, Spinal cord cavernous malformations, Spinal cord, Vascular malformation

INTRODUCTION

Intramedullary spinal cavernous malformations (ISCM) make up only 1% of all intramedullary spinal lesions in the pediatric population. Children most typically present with acute neurological deterioration characterized by the acute onset of severe motor deficits.[5] Of interest, spinal cord ISCM lesions are considered more aggressive than those occurring intracranially due to the limited space within the spinal cord.[13] Nevertheless, if recognized and treated in a timely fashion, ISCMs have been shown to have favorable outcomes, when compared to the adult population.[5]

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.[15] 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.[1] They consist of sinusoidal vascular spaces lined by a single layer of endothelial cells encompassed by a loose connective tissue stroma.[8] 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.[17] 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.[12]

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 T1and 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).[7,9] 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.[3,9,14] 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 REHMORRHAGE 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.[2,6,9,10] Ren *et al*. reviewed a series of 20 pediatric patients with ISCMs[15] 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.[15] 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.[15] Badhiwala *et al*. did not show a statistically significant relationship between ISCM size and neurological outcome.[2] 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.[2]

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.[18] Expedited surgery is particularly indicated in the pediatric population to reduce the risk of re-hemorrhage/long-term sequelae.[4] While most spinal cavernomas are found in the cervical spine, they do occur in the thoracic and lumbar levels.[19]

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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How to cite this article: Fiani B, Reardon T, Jenkins R, Covarrubias C, Sekhon M, Soula M, *et al*. Intramedullary spinal cord cavernous malformations in the pediatric population. Surg Neurol Int 2020;11:275.