

Pathophysiology, Diagnosis, and Treatment of Spinal Meningoceles and Arachnoid Cysts

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Though the nomenclature regarding spinal arachnoid cysts and meningoceles is confusing and sometimes overlapping, spinal arachnoid cysts and meningoceles are distinct entities with different presentations and etiologies. Meningoceles are usually congenital lesions discovered at infancy with a high incidence of associated anomalies. Spinal arachnoid cysts are most often asymptomatic but can cause nerve root and/or cord compression. We review the presentation, pathophysiology, and management of these lesions.

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Spinal cysts refer to cavities that occur within the spinal cord substrate itself or its coverings and can occur throughout the length of the spinal column. This review will focus on two seen anomalies, spinal meningoceles and spinal arachnoid cysts, and will review current classification systems, pathophysiology, diagnostic evaluations, and treatment options.

Before discussing spinal meningocele, a brief review of dysraphism is warranted as spinal meningocele is a form of closed neural tube defect. Spinal dysraphism is characterized by failure of differentiation and/or fusion of dorsal midline structures through the embryonic developmental stages of gastrulation, primary neurulation, and secondary neurulation. Dysraphism is divided into two clinical subsets: open and closed. In open dysraphism the neural placode is exposed to the environment giving rise to entities such as myelomeningocele, hemi-myelocele, and myeloschisis, which are almost always associated with Chiari type II malformations. Closed spinal dysraphism, by contrast, is characterized by neural arch defects covered with intact skin. Closed dysraphism can be further divided into two subsets based on the presence or absence of a subcutaneous mass. Liposchisis, lipomyelomeningocele, meningocele, or myelocystocele may present as closed dysraphism with a mass, whereas closed spinal dysraphism without a mass comprise complex dysraphic states ranging from complete dorsal enteric fistula to

neurenteric cyst, split cord malformation, dermal sinus, caudal regression, spinal segmental dysgenesis, bony spina bifida, tight filum terminale, filum or intradural lipomas, and persistent terminal ventricle.¹

Spinal meningocele is defined as a protrusion of the spinal meninges through a defect in the vertebral column or foramina usually in association with a congenitally dysraphic vertebrae, with the spinal cord remaining entirely confined to the vertebral canal.^{2,3} These lesions are frequently identified in a posterior location over the thoracic and sacral areas at birth and constitute about 10% of all patients with spina bifida.² Much more rarely seen are meningoceles protruding through anterior, lateral, or anterolateral defects. One example of such is the exceedingly rare anterior sacral meningocele. More than three-quarters of cases are found in females. This may reflect, however, the fact that women of reproductive age are more likely to undergo pelvic examination than males. Interestingly, uterine anomalies are frequently found in association with anterior meningoceles.⁴ An anterior sacral mass with an associated sacral defect when present with an anorectal anomaly is known as Currarino's triad.⁵ Another rare form of meningocele is the nondysraphic meningocele, characterized by the absence of a congenital defect of the vertebrae. These are usually located in the anterior thoracic level and are frequently associated with neurofibromatosis, or less commonly with Marfan's syndrome.²

Spinal arachnoid cysts are uncommon arachnoid protrusions that result in a collection of cerebrospinal fluid that can occur in a perineural, extradural, intradural, or intra-extradural site.⁶⁻⁸ Extradural cysts exist as outpouchings of the arachnoid that communicate with the intraspinal subarachnoid space through a small defect in the dura.^{9,10} Intradural

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spinal arachnoid cysts usually consist of multiple lobules.¹¹ For our purposes the discussion here will be restricted to intradural arachnoid cysts. Arachnoid cysts are expanding lesions covered by a lining of arachnoid-like cells.^{6,12} Histologically, cells lining arachnoid cysts do not immunostain with antibodies against GFAP, S-100, transthyretin (prealbumin), and CEA, helping to differentiate arachnoid cysts from epithelial cysts.¹³

These lesions are located posterior to the spinal cord in 80% of cases, but have also been identified anteriorly and, rarely, laterally within the dentate ligaments. Most frequently they occur in the thoracic spine (70%) or at the thoracolumbar junction (12%), but they can also be found in the lumbosacral (13%) and cervical (3%) spine.¹² Men and women seem to be equally affected, usually in the third through fifth decades of life.¹⁴

Difficulties have arisen in the classification of spinal arachnoid cysts because the cause and pathogenesis of these lesions are still debated. The nomenclature for this entity is also confusing as intradural arachnoid cysts are variously referred to as subdural arachnoid cysts, subarachnoid cysts, leptomeningeal cysts, arachnoid diverticula, and meningeal hydroids.^{15,16} Arachnoid cysts may be classified as primary or secondary. Primary cysts most likely arise during development and may expand throughout life and progressively displace spinal tissue. Secondary cysts follow a variety of spinal insults including head injury, hemorrhage, chemical irritation, meningitis, and tumors.^{17,18} In 1988, Nabors and coworkers classified spinal cysts simply into three major categories on the basis of surgical, radiological, and histological criteria. Type I are spinal extradural meningeal cysts without spinal nerve root fibers (two subclassifications are defined as Type IA, extradural meningeal cyst, and Type IB, sacral meningocele). Type II are spinal extradural meningeal cysts with spinal nerve root fibers (Tarlov cyst), and Type III are spinal intradural cysts, which are intradural spinal arachnoid cysts.¹⁹ Intradural spinal arachnoid cysts should be differentiated from *Tarlov* cysts, which are extradural perineurial dilations of the posterior spinal nerve root sheath. They have potential or restricted communication with the spinal subarachnoid space and usually occur distal to the junction of the posterior nerve root and dorsal root ganglion.²⁰ Tarlov cysts are discussed in another article in this publication.

Pathophysiology

Spinal meningoceles are usually a form of dysraphism and are thought to arise during development. Development of the spinal cord is traditionally divided into three periods: gastrulation, primary neurulation, and secondary neurulation. During gastrulation (weeks 2 to 3), the embryo undergoes conversion from a bilaminar to a trilaminar structure with establishment of the nuchal cord. This is followed during gestational weeks 3 and 4 by primary neurulation where the upper 9/10th of the spinal cord is formed. Finally secondary neurulation and retrogressive differentiation occur in weeks 5 to 6 with formation of the conus medullaris and filum terminale.¹ Lumbar meningoceles are believed to occur as a

disorder of secondary neurulation. While patients with meningoceles are usually neurologically normal, 90% of patients with meningocele have associated occult spinal lesions such as tight filum terminale, split cord malformation, and epidermoids. Subsequent tethering of the spinal cord may lead to progressive neurological deficit.²¹

Anterior sacral meningoceles result from failure of fusion of the sacrum with subsequent herniation of the sacral meninges into the sacral hollow. Because of their occult location, they typically present later on in life with protean neurologic, urologic, or gastrointestinal complaints.⁴ Anterolateral meningoceles are very rare lesions that have been reported in association with neurofibromatosis and Marfan's syndrome. They are postulated to occur because of congenital mesodermal dysplasia and hypoplastic bone changes.^{2,22}

Arachnoid cysts likely result from alterations in the meningeal layer of the spine and the arachnoid trabeculae that overlie the spinal cord. Whether this is the result of a congenital malformation or from a causative factor such as previous trauma, inflammation, surgery, or subarachnoid hemorrhage, or are completely idiopathic is a greatly debated issue.^{13,14,21,23-26} The rare association of these lesions with neural tube defects may support the idea of a congenital etiology.²⁷ Perret and coworkers theorized that these lesions develop from the septum posticum of Schwalbe, an arachnoid membrane dividing the midline posterior cervical and thoracic subarachnoid space.²⁸ However, problems with this theory have arisen as primary arachnoid cysts have been identified anterior to the spinal cord. Fortuna and coworkers have suggested that incarceration of arachnoid granulations may produce cerebral spinal fluid that becomes entrapped in arachnoid diverticula. These sequestered pockets of fluid lead to further disruption of normal pulsatile cerebrospinal fluid (CSF) flow and thus are capable of expanding and developing into cysts.^{16,29} The finding of melanocytes in an arachnoid cyst by Morioka and coworkers led to the idea that emigrating melanocytes from the neural crest migrated selectively to the original tissue of the arachnoid membrane, which developed an anomalous proliferation in the arachnoid diverticula or septum posticum, resulting in an arachnoid cyst.²³ This theory is based on congenital melanocytic nevi having nevus cells in the dermis congenitally, which emigrate from the neural crest to the dermis at 10 weeks of gestation in primates. Spiegelmann and coworkers reported that the histological examination of an intradural arachnoid cyst revealed hemosiderin containing macrophages trapped in the cyst wall, along with the findings of scarred arachnoid and adhesions at surgery, which led them to consider the arachnoid cyst to be a delayed consequence of the previous injury sustained by the patient.³⁰ A hydrodynamic theory in which normal CSF pulsations dilate weak areas of arachnoid and progressively enlarge into a cyst by the everyday variations in the cerebrospinal fluid hydrodynamics suggests that the mechanism of cyst enlargement and the degree of communication with the subarachnoid space is possibly related to a ball valve effect at the neck of the diverticulum.^{10,12,18} Both spinal meningoceles and arachnoid cysts have also been described in patients with Marfan syndrome, neurofibromatosis, autosomal-dominant

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