A Tarlov cyst is a bulbous enlargement of a spinal nerve root cerebrospinal fluid (CSF) space that is distinct from other meningeal cysts, such as dural ectasia or meningeal diverticula. Tarlov cysts most often are found in the sacral spinal canal, where they can produce bone erosion and compression of adjacent spinal nerve roots, resulting in a debilitating sacral radiculopathy syndrome.

Tarlov first described these cysts in 1938 during his autopsy studies of the filum terminale at the Montreal Neurological Institute. Since his seminal report, numerous cases of symptomatic Tarlov cysts have been published in the literature.

With the advent of MRI, our ability to diagnose meningeal cysts, such as dural ectasia or meningeal diverticula, has been enhanced.

The treatment of symptomatic Tarlov cysts has evolved, along with our understanding of their pathophysiology. Various therapeutic strategies have been described over the years, with more recent literature trending toward definitive surgical treatment. In this chapter the pathologic, radiographic, and clinical characteristics of Tarlov cysts are discussed, along with our understanding of their pathophysiology and the current treatment options are presented.

Epidemiology and Histology

Tarlov, or perineurial, cysts are one of the most common forms of meningeal cysts. Estimates of the prevalence of meningeal cysts, including Tarlov cysts, in the general population vary, but generally are in the 5% range. In a study of 500 consecutive patients with back pain undergoing lumbosacral MRI, 5% were found to have one or more meningeal cysts. Among this latter group, the cyst was thought to be the source of the symptoms in 1% of the cases. Tarlov cysts, particularly those that are symptomatic, are more common among women. The reason for this is unclear, and we have postulated that there may be gender-related differences in the fundamental makeup of dura mater or spinal nerve roots that produce this epidemiologic disparity.

Tarlov distinguished perineurial cysts from other meningeal cysts based on several histologic criteria. He defined them as perineurial dilations that develop between the endoneurium and perineurium, typically of the S2 or S3 nerve roots, just proximal to the junction of the dorsal root ganglion and nerve root (Fig. 115-1). Simply stated, each cyst is a dilated spinal nerve root sheath, and the individual nerve fibers of that root are found running within the cyst cavity or its inner lining. Other meningeal cyst subtypes, such as meningeal diverticula and arachnoid cysts, typically are devoid of nerve root fiber elements.

Tarlov cysts can be single or multiple, and can develop anywhere along the spine where nerve roots are present. Progressive cyst enlargement can cause significant bone erosion and impingement of adjacent spinal nerve roots, producing corresponding radiculopathies. For example, a Tarlov cyst in the sacral spinal canal arising from the S3 nerve root can cause symptomatic impingement of the ipsilateral S2 nerve root beside it, and of the S4 or S5 nerve root below (Fig. 115-2). A Tarlov cyst can also produce contralateral symptoms if it is large enough to extend across the midline and compress contralateral nerve roots. Additionally, the nerve root fibers running inside a Tarlov cyst often are attenuated and splayed out over the inner wall of the cyst. This neural fiber alteration and stretching also are suspected of causing symptoms.

Tarlov cysts occasionally can be found in combination with other meningeal cysts. For example, patients with connective tissue disorders, such as Marfan syndrome, can have Tarlov cysts and large ectatic dural cysts so extensive that the distal spinal sac extends out into the pelvis (Fig. 115-3).

The pathogenesis of Tarlov cysts remains unclear. Tarlov proposed that cyst formation could be the result of trauma, ischemic degeneration, inflammation, or hemorrhagic infraction from the subarachnoid space. Some patients with symptomatic Tarlov cysts report a history of sacral trauma, and evidence of old hemorrhage in the form of hemosiderin deposits and dystrophic calcification within Tarlov cyst walls supports prior trauma as an etiologic factor. Other reports have suggested that Tarlov cysts result from arachnoidal proliferation or blockage of perineural fluid flow. Nabors et al. support a developmental origin, although an association between Tarlov cysts and spinal dysraphism is not as strong as that with other types of meningeal cysts. Only two patients with symptomatic Tarlov cysts and spina bifida have been reported, and the relationship could have been coincidental.

Strully et al. and Smith proposed that Tarlov cysts form as a result of increased CSF hydrostatic pressure. They point out that spinal nerve roots are in communication with the thecal sac, and that there is myelographic evidence that spinal fluid flows within the nerve roots and could produce
dilatation due to either higher hydrostatic pressure or inherent, traumatic, or iatrogenic weakness in the nerve root sheath. They also point out that the frequency and size of Tarlov cysts along the spine can be correlated with the rostral-caudal hydrostatic pressure gradient.\(^{17,18}\) Several reports on patients with Tarlov cysts have documented either a history of straining or coughing or an exacerbation of symptoms by these maneuvers.\(^{7,10,11,18}\) We also are aware of two cases of Tarlov cysts in patients with pseudotumor cerebri. However, no criteria have been established to determine who might benefit from CSF shunting for Tarlov cysts, and investigations are ongoing.

### Diagnosis

The treatment of a symptomatic Tarlov cyst first requires a correct diagnosis. Unfortunately, many patients languish with undiagnosed or untreated symptomatic cysts due to the incorrect "rule" that Tarlov cysts always are asymptomatic, regardless of the presence of blatant compression of adjacent nerves or extensive bone erosion. Such patients often are relegated to an escalation of narcotics, injection procedures, and neuromodulatory medications as they become progressively more symptomatic. It is not uncommon to encounter patients who have developed narcotic dependency after management with extended-release morphine, transdermal fentanyl, or implanted pain pumps before they are finally referred for meningeal cyst evaluation.

Even more unfortunately, we have encountered patients with symptomatic Tarlov cysts that were misdiagnosed with a variety of other ailments and treated unsuccessfully with a variety of procedures, such as hysterectomy, laparoscopic exploration, endometriosis surgery, oophorectomy, appendectomy, surgery for piriformis syndrome, sacroiliac joint fusion with implanted cages, fusion of degenerative discs in the adjacent spine, coxectomy, and urinary bladder procedures (Fig. 115-4).
Symptoms

Tethered Cord Syndrome

The presence of a tethered spinal cord sometimes may be overlooked in the setting of a radiographically impressive sacral Tarlov cyst. Such patients typically present with very similar symptoms, including pain and numbness in the low back or sacral region that radiates down the backs of the legs. They also can have overlapping bowel, bladder, and sexual dysfunction. As with Tarlov cysts, the onset of tethered cord symptoms may occur at any time in life, particularly following trauma, childbirth, or hyperflexion of the spine. Patients with connective tissue disorders such as Marfan or Ehlers-Danlos syndrome can have meningeal cysts, including Tarlov cysts, and occult tethered spinal cord even though the conus is at level L1-2 or above.

Unlike Tarlov cysts, however, tethered cord syndrome is associated with toe walking, flat feet or pes cavus, enuresis, and scoliosis. A spinal neurocutaneous marker sometimes may be present. The pain associated with tethered cord syndrome usually is more moderate, exacerbated by straight leg raising or other maneuvers that stretch the cauda equina, and is not affected by upright posture, as with Tarlov cysts. Muscle atrophy is prominent, and especially in the L5- and S1-related muscles. Imaging studies may reveal syringomyelia, descent of the conus to the L2 level or below, and a thickened fatty filum or a lipomeningocele.

Spinal Stenosis and Occult Spondylolisthesis

Stenosis of the central canal or neural foramina can mimic Tarlov cyst–related symptoms and should be carefully ruled out with appropriate high-resolution imaging and dynamic radiographs. Overlapping symptoms common to both include radicular pain and numbness and urinary symptoms.

Cervical and thoracic stenosis symptoms not typical of Tarlov cysts include long-tract findings, such as spasticity and pathologic reflexes like the Babinski and Hoffman signs, and absence of the abdominal reflex. In cases of lumbar stenosis, patients usually describe pain in the lumbar area, whereas patients with symptomatic sacral Tarlov cysts complain more specifically about pain well below in the sacral region.

Other Masses

Schwannomas and neurofibromas can produce a radiculopathic pattern of symptoms similar to those of Tarlov cysts. They also can share imaging characteristics, such as a cystic appearance, lateral location in the spinal canal, and production of bone erosion and foraminal expansion (Fig. 115-5). However, Tarlov cysts do not enhance on MRI following the administration of gadolinium contrast, which is a characteristic typical of schwannomas and neurofibromas. Instead, they have signal characteristics similar to spinal fluid on all sequences, with the possible exception of differing signal when there has been hemorrhage or accumulation of stagnant, more proteinaceous, spinal fluid within a cyst.
FIGURE 115-5. A, This left S1 foramen mass appears hyperintense on T2-weighted imaging. B, Unlike a Tarlov cyst, however, it enhances on T1-weighted imaging following gadolinium administration. At surgery, the lesion was found to be a schwannoma.

FIGURE 115-6. A, The cystic lesion seen centrally and to the left in the spinal canal has imaging characteristics similar to those of a Tarlov cyst on T2-weighted imaging. B, However, on T1-weighted imaging, the lesion is hyperintense to cerebrospinal fluid and has some areas of internal heterogeneity. Further workup with CT myelography, needle biopsy, and angiography revealed the lesion to be a large venous angioma.

We also have encountered other lesions with imaging characteristics similar to those of Tarlov cysts. For example, in one case, a large cystic lesion filling the left S1 foramen had the T2-weighted imaging appearance of a Tarlov cyst, but differed on other sequences (Fig. 115-6). Further workup revealed the lesion to be a large venous angioma within the spinal canal.

Radiography

MRI currently is the gold standard for imaging meningeal cysts.\(^\text{21,22}\) Not only is it useful for differentiating them from other lesions, but it also can help distinguish among the different types of meningeal cysts, including Tarlov cysts. For example, Tarlov cysts typically are lateral in the spinal canal and arise from an individual spinal nerve root. Nerve root fibers often are identifiable inside the cyst on T2-weighted images (Fig. 115-7). In contrast, meningeal diverticula are found centrally in the spinal canal, and arise from the tip of the spinal sac, not from an individual nerve root. When they are large, Tarlov cysts can be seen to erode and expand the spinal canal or neural foramina and extend into the retroperitoneal pelvis (Fig. 115-8).

MRI also is superior for defining anatomic relationships with surrounding structures. For example, a careful review of imaging studies often reveals symptomatic cysts to be blatantly compressing adjacent nerve roots and displacing the spinal sac. Understanding these relationships preoperatively is critical for surgical planning.

CT myelography previously was used preoperatively to distinguish Tarlov cysts from other forms of meningeal cysts based on the premise that Tarlov cysts filled poorly and in a delayed fashion.\(^\text{1,2,16,19}\) Other meningeal cysts, such as meningeal diverticula, were believed to fill more rapidly. However, these criteria are not reliable, because the extent of dye penetration into a cyst depends on the degree of its communication with the spinal sac, not on the cyst type. For example, we have encountered Tarlov cysts that communicate quite freely with the spinal sac, and filled readily on CT myelography (Fig. 115-9). Such cysts would have been erroneously categorized as non-Tarlov cysts using the more antiquated radiographic criteria.

Despite the fact that meningeal cysts, particularly Tarlov cysts, can be radiographically impressive, a careful search should be conducted to rule out other pathology that might explain the patient's symptoms. For example, a complete workup of a sacral Tarlov cyst should include not only a sacral

FIGURE 115-7. The nerve root fiber bundle inside this sacral Tarlov cyst is clearly seen on an axial T2-weighted MRI.

FIGURE 115-8. A, Two large intrapelvic Tarlov cysts are seen on this axial T2-weighted MRI (black arrows). Also seen is a separate Tarlov cyst within the sacral spinal canal abutting a nerve root to the right of midline (white arrow). B, On this sagittal image from the same patient, the left intrapelvic cyst can be seen extending out through the S1 neural foramen ventrally into the retroperitoneum.

FIGURE 115-9. A, Side-by-side Tarlov cysts are present on this axial CT myelogram image (arrowheads). One fills with dye but the other does not, demonstrating that CT myelography is unreliable as a diagnostic tool for this pathology. B, Unlike CT, both Tarlov cysts are clearly seen on T2-weighted MRI, the preferred imaging modality.
Unfortunately, a significant number of patients with symptomatic Tarlov cysts undergo percutaneous needle aspiration procedures as an attempt at treatment, not for diagnosis. Such procedures are ineffective, because aspirated fluid within a Tarlov cyst typically is replaced rapidly with spinal fluid through the proximal nerve root in communication with the spinal sac. One study that assessed the effectiveness of percutaneous drainage of Tarlov cysts found that four of the five patients in that series suffered a recurrence of symptoms. A separate report in 2001 described patients who underwent percutaneous aspiration of their cysts preoperatively. None of those patients improved, and, in fact, some experienced marked worsening of their symptoms. This deterioration may have been the result of hemorrhage in the cyst wall, or nerve root injury.

In general, the use of aspiration for treatment of a Tarlov cyst is inconsistent with an understanding of the fundamental underlying pathology involved and exposes the patient to the risk of spinal fluid leakage, meningitis, hemorrhage, and nerve root injury. We therefore restrict the use of needle aspiration, using it only as a diagnostic tool in rare situations, as described in the preceding section.

Needle Aspiration and Fibrin Glue Injection

Treatment with percutaneous aspiration followed by fibrin glue injection also has been described. Authors of one report of four patients with symptomatic Tarlov cysts found that percutaneous fibrin glue therapy was effective in alleviating symptoms, although three patients developed postprocedural aseptic meningitis. Blind percutaneous introduction of multiple needles into a Tarlov cyst also increases the probability of causing nerve injury.

In our experience, fibrin glue treatment fails in a large number of patients, and those patients require subsequent surgery. Unfortunately, the introduction of fibrin glue or other substrates into a Tarlov cyst makes subsequent surgical treatment more difficult because it produces scarring or coating. The neural elements within the cyst are then much harder to identify and protect during surgery. We have encountered injured nerve roots intraoperatively in several patients following prior failed fibrin glue treatment (Fig. 115-10). The fibrin glue technique is falling out of favor, both in the United States and abroad, with only a few centers with a significant experience continuing its use.

Surgical Treatment

Patient Selection

As with most spinal pathology, the selection of patients for surgical treatment is based on the correlation among symptoms, physical examination, and radiographic findings. Additionally, the size of a Tarlov cyst is an important factor in determining the probability that it is symptomatic. In general, the larger the cyst the more likely it is to produce symptoms. One study found that patients with neurologic deficits that can be radiographically correlated with Tarlov cysts greater than 1.5 cm in diameter enjoyed substantial improvement following surgery. Furthermore, there was a very strong association between the presence of radicular symptoms and excellent outcome.

However, we also have encountered patients with symptomatic cysts smaller than 1.5 cm. The location of a Tarlov cyst and the extent to which it is compressing adjacent neural structures also are important factors, in addition to its size. For example, an intraforaminal Tarlov cyst in the cervical or thoracic spine arising from one spinal nerve ramus can compress the adjacent ramus of the same nerve root and produce symptoms, even though the cyst is small.

Surgery

The treatment of a symptomatic Tarlov cyst must take into account that the cyst is actually a dilated nerve root. Therefore, simple excision usually is not an option, because it can produce a critical deficit, particularly when in the lumbo-sacral region. This results in a treatment quandary: how does
one eliminate a symptomatic Tarlov cyst without injuring the nerve root fibers inside and producing a neurologic deficit?

In the past, surgical treatments such as decompressive laminectomy, cyst fenestration, and complete cyst excision have fallen from favor due to a lack of success and unacceptable complication rates. More recent surgical techniques have targeted the underlying pathology causing nerve root dilatation, that is, the process that allows spinal fluid to accumulate within an affected nerve root.

An initial step in this direction was the description of fenestration and imbrication techniques that involve opening a Tarlov cyst. The cyst is then reduced in size, either by imbrication or partial wall excision, thereby reconstituting a more normal caliber nerve root, which no longer compresses adjacent structures. However, such techniques do not prevent the continued flow of spinal fluid into the affected nerve root cyst, so they do not eliminate the risk of cyst reexpansion and spinal fluid leakage. Additionally, nerve fascicles in a Tarlov cyst often are found within the cyst wall itself, or so extensively splayed out over the internal surface of the cyst wall that they are not easily seen, even with the aid of an operating microscope. Cyst wall excision to decrease the overall size of a Tarlov cyst therefore increases the risk of producing deficits due to inadvertent nerve root fiber sectioning.

Current surgical techniques are focused on resolving the quandary of how to prevent spinal fluid flow into a symptomatic Tarlov cyst without injuring its nerve root fibers. To this end, we have focused on treatment of the ostium, where spinal fluid and nerve root fibers enter the cyst. We also have made efforts to confine cysts to prevent further compression of adjacent structures (Fig. 115-11).

Intraoperative electrophysiologic monitoring is an integral component of Tarlov cyst surgery. During the process of exposure and cyst dissection, it aids the surgeon in identifying specific nerve roots and gives feedback on the tolerance of nerves to manipulation. Thus, intraoperative monitoring can be used to assess the status of a Tarlov cyst–involved nerve root throughout the process of cyst treatment, and gives the surgeon a baseline to assess nerve function before and after cyst treatment. For example, the nerve root sleeve proximal to a treated cyst can be stimulated following cyst exploration and treatment to determine whether it is still in continuity and conducting as it was before treatment.

Summary

Tarlov cysts are an important clinical entity in the differential diagnosis of spinal radiculopathy, sacral pain syndromes, and sacral spinal insufficiency fractures, particularly in women. The symptoms they produce can be crippling. Although the relationship remains unclear, elevated CSF pressure and trauma may play a role in their pathogenesis. MRI has enhanced our ability to diagnose symptomatic Tarlov cysts and preoperatively assess their anatomic relationships.

The management of symptomatic Tarlov cysts has progressed significantly in the last decade. Trends in the literature have favored surgical treatment in experienced hands to reduce the risk of nerve root injury and spinal fluid leakage. Treatments with percutaneous needle techniques such as aspiration or fibrin glue injection are falling out of favor due to lack of symptomatic improvement and high cyst refilling rates. In our experience, patients with large cysts and corresponding radicular symptoms are more likely to experience substantial relief from surgery. However, patients with smaller cysts also can benefit if focal nerve root compression is identified on preoperative imaging.

KEY REFERENCES


REFERENCES

The complete reference list is available online at expertconsult.com.