**Tarlov cyst**

Tarlov cyst is an abnormal expansion of the spinal nerve sleeve, and communicates with the subarachnoid space via a perineural fistula.

Tarlov cysts (TCs) occur most commonly on extradural components of the sacral nerve and coccygeal nerve roots.

**Etiology**

In clinical practice, Xie et al., from the Department of Neurosurgery, Wenzhou Medical University Second Affiliated Hospital, Wenzhou, China and Shanghai jiaotong University School of Medicine, Xinhua Hospital, Shanghai, China, noted that the end section of the thecal sac is apparently different in patients with Tarlov cyst compared with that of the normal population.

They conducted a clinical study based on magnetic resonance imaging (MRI).

The study included 30 patients with Tarlov cyst and 30 healthy volunteers as the control. The L4, L5 cross-section areas, the L4, L5 anteroposterior diameters, and the terminal length of the thecal sac were measured from the lumbosacral spine MRI. Results The L4, L5 cross-section areas and the L4, L5 anteroposterior diameters are larger for the Tarlov cyst patients than the controls, but the terminal length of the thecal sac is shorter.

The Tarlov cyst is correlated with a short broad end of the thecal sac. Possibly, this anatomical variant is a causative factor of Tarlov cyst 1).

Some incidents or conditions that might potentially cause the asymptomatic cysts to become symptomatic are traumatic injuries such as falls, automobile accidents, heavy lifting, childbirth, and epidurals. Trauma to the spinal cord, an increase in the CSF pressure, or a blockage of the CSF can result in cyst formation. Read “What happens in spinal cord injuries?”. It is also noted that the herpes simplex virus can cause the Tarlov cyst symptoms to worsen during a herpes virus outbreak, but there is no known “connection” related to any virus or infection with causing Tarlov cysts to develop.

There is significant clinical evidence that collagen mutations or connective tissue disorders such as Marfan’s, Ehlers-Danlos, Sjogren’s, Loeys-Deitz, Lupus, MCTD (mixed connective tissue disorders), UCTD (undiﬀerentiated connective tissue disorders) are predisposing or contributing to the cysts developing. A number of Tarlov cyst patients have also been diagnosed with a connective tissue disorder, and many more of their biological family members, who may or may not have Tarlov cysts, have been diagnosed with one of the above mentioned connective tissue disorders (CTD).

**Epidemiology**

These lesions are often found incidentally on magnetic resonance imaging, with an estimated prevalence of 4%-9%.

They are located most prevalently at the S2, S3 level of the sacrum.

A significant percentage (11%) of patients have cysts at more than one location of the spine.
Some patients have cysts at all sections of the spine, including cervical, thoracic, lumbar, and sacral.

**Clinical features**

Symptoms of expanding/enlarging cysts occur due to compression of nerve roots that exit from the sacral area. Symptoms may include the following, dependent on the location of the cysts and the section of the spine they occur:

- Pain in lower back (particularly below the waist) and in buttocks, legs, and feet
- Pain in the chest, upper back, neck, arms and hands
- Weakness and/or cramping in legs and feet / arms and hands
- Paresthesias (abnormal sensations) in legs and feet or arms and hands, dependent on cyst locations
- Pain sitting or standing for even short periods of time
- Pain when sneezing or coughing
- Inability to empty the bladder or in extreme cases to urinate at all requiring catheterization
- Bowel or bladder changes, including incontinence
- Swelling over the sacral (or cervical, thoracic, or lumbar) area of the spine
- Soreness, a feeling of pressure and tenderness over the sacrum and coccyx (tailbone), extending across the hip and into the thigh with cysts in the sacrum. Same feelings in upper sections of the spine dependent on cyst locations
- Headaches (due to the changes in the CSF pressure) and sometimes accompanied by blurred vision, double vision, pressure behind the eyes and optic nerve pressure causing papilledema (optic nerve swelling)
- Other sensory system symptoms: Tinnitus/Ear noises (ringing, buzzing, snapping, popping, cricket sounds, etc.)
- Dizziness and feeling of loss of balance or equilibrium, especially with change of position
- The feeling of sitting on a rock
- Pulling and burning sensation in coccyx (tailbone) area, especially when bending
- Sciatica
- Vaginal, rectal, pelvic and/or abdominal pain
- Restless leg Syndrome
- PGAD (Persistent Genital Arousal Disorder)
- Sexual dysfunction and painful intercourse
Diagnosis

Sometimes they are found incidentally with magnetic resonance imaging.

The TCs appear on MRI to be dilated or ballooned areas of the sheaths that cover nerve roots.

They exhibit delayed filling with contrast medium on myelography, which is one of the criteria used by Tarlov to distinguish perineural cysts from meningeal diverticula.

Differential diagnosis

It is sometimes confusing to make an accurate diagnosis as to the cause of the symptoms, if there are multiple diagnoses found, such as herniated discs, ruptured disc, DDD (degenerative disc disease). It is sometimes diagnostically conclusive that the cysts are the cause of symptoms, when pain is improved by aspirating the fluid from the cysts. Although using a needle to aspirate CSF from the cysts can temporarily relieve symptoms, eventually the cysts will refill and the symptoms will recur usually within hours.

Treatment

Given the low estimated rates of symptomatic TC and the fact that symptoms can overlap with other common causes of low-back pain, optimal management of this entity is a matter of ongoing debate.

Pain may be also temporarily controlled by aspiration of the cysts and then injecting the cysts with fibrin glue (a substance produced from blood chemicals involved in the clotting mechanism). The aspiration of CSF and injection of fibrin glue procedure theoretically is designed to remove the CSF from the cyst, and to block the entrance or the neck of the cyst with the sealant glue, to prevent return of the flow of CSF into the cyst. Some patients have found immediate relief after the procedure, while others have reported a delayed benefit from the procedure when the nerve irritation has subsided. After the procedure, there are outcomes of both short term relief, as well as longer term relief reported. However, it is considered to be a temporary relief procedure.

Transcutaneous Electrical Nerve Stimulation (TENS) has been proven useful for some in pain management. TENS devices deliver electrical impulses through the skin to the cutaneous (surface) and afferent(deep) nerves to control pain. Unlike medications and topical ointments, TENS does not have any known side effects, other than skin irritation from the electrodes seen in some patients.

Case series

2016

Elsawaf et al., retrospectively reviewed their clinical data archive from 2002 to 2014. Fifteen patients who were operated on due to symptomatic sacral perineural cysts were enrolled in the study. Patients' symptoms, radiographs, intra-operative findings, and clinical results were evaluated. All 15 patients underwent microsurgical excision of the cyst. The literature on this topic available in PubMed was also reviewed.

There were 5 men and 10 women included in the study, with a mean age of 31 years (range 7-60 years). Preoperative symptoms include low back pain, coccydynia, buttock pain, perianal pain and radicular pain. All of the patients underwent surgical resection. The mean follow-up was 54 months.
Cyst excision is an effective and safe technique for symptomatic sacral perineural (Tarlov) cysts. Careful patient selection is vital to the management and treatment of this difficult and controversial pathology

Burke et al., performed a retrospective review of data from consecutive patients who were surgically treated for symptomatic TCs from September 2011 to March 2013. Clinical evaluations and results from surveying pain and overall health were used. Univariate statistical analyses were performed.

Twenty-three adults (4 males, 19 females) who had been symptomatic for a mean of 47.4 months were treated with laminectomy, microsurgical exposure and/or imbrication, and paraspinous muscle flap closure. Eighteen patients (78.3%) had undergone prior interventions without sustained improvement. Thirteen patients (56.5%) underwent lumbar drainage for an average of 8.7 days following surgery. The mean follow-up was 14.4 months. Univariate analyses demonstrated that an advanced age (p = 0.045), the number of noted perineural cysts on preoperative imaging (p = 0.02), and the duration of preoperative symptoms (p = 0.03) were associated with a poor postoperative outcome. Although 47.8% of the patients were able to return to normal activities, 93.8% of those surveyed reported that they would undergo the operation again if given the choice.

This is one of the largest published studies on patients with TCs treated microsurgically. The data suggest that patients with symptomatic TCs may benefit from open microsurgical treatment. Although outcomes seem related to patient age, duration of symptoms, and extent of disease demonstrated on imaging, further study is warranted and underway.

Potts et al., retrospectively reviewed all consecutive cases of symptomatic giant sacral Tarlov cysts treated with microsurgery at the Department of Neurological Surgery, University of California, San Francisco, California, USA between 2003 and 2011. The main outcome measure was self-reported symptom relief. Postoperative imaging, surgical complications, and subsequent treatments were also recorded.

Thirty-five patients were treated. Mean age was 52 years. All patients presented with a chief complaint of sacral-perineal pain. The mean cyst size was 3.6 cm (largest diameter). Follow-up beyond the initial hospital stay was available in 86% (median 8 months). Ninety-three percent reported improvement in pain at some point during the postoperative course but 50% of those developed recurrent pain symptoms. Postoperative imaging was available in 69% of the patients in whom 92% showed complete obliteration (25%) or reduction in cyst size (67%).

The combination of microsurgical cyst fenestration and the use of vascularized muscle pedicle flaps to fill the cystic cavity and the epidural space results in obliteration or reduction in size of the majority of cysts and is associated with initial improvement in pain in most patients. However, delayed recurrence of pain was common with this technique.

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Twenty-two patients with symptomatic Tarlov cysts were surgically treated. An emulsion balloon was placed into the lumbar subarachnoid cistern through a trocar, so as to temporarily block cerebrospinal fluid flow, then the thecal sac was opened and the inlet of the fistula was sealed by suture of a muscular patch and reinforced by fibrin glue. At last the cyst wall was imbricated and the bony cavity was filled with pedicled muscle flaps.

Comparing the preoperative and postoperative pain scores according to visual analog scale, 2 patients were slightly improved; 18 patients were substantially improved, including 3 completely pain-free cases. Only 2 patients were unchanged in pain, and both of them were multiple cysts. As a whole, the postoperative pain score is much better than the preoperative score (2.4 vs 7.5, P<0.01). Bladder weakness was slightly improved, and bowel dysfunction was almost unchanged after operation. During follow-up, cyst recurrence was found in one patient.

The balloon-assisted fistula sealing procedure is safe and effective for Tarlov cyst, especially for the single cyst. It's a good complement to the cyst wall imbricating procedure.

2011

In 13 consecutive patients (10 female, 3 male), MRI revealed sacral perineural cysts and excluded other pathologies. Micro-communication between the thecal sac and the cysts was shown by delayed contrast filling of the cysts on postmyelographic CT. Surgical fenestration achieved free CSF communication between the thecal sac and cysts in all patients. The patient histories, follow-up examinations and self-assessment scales were analyzed. Symptoms at initial presentation included lumbosacral pain, pseudoradicul symptoms, genital pain and urinary dysfunction. Mean follow-up was 10.7 ± 6.6 months.

Besides one CSF fistula, no surgical complications were observed. Five patients did not improve after surgery; in four of these cases multiple cysts were found, but small and promptly filling cysts remained untreated. Seven patients reported lasting benefit following surgery; three of these had single cysts, and all had cysts >1 cm. One patient initially benefited from cyst fenestration but experienced recurrent pain within 2 months postoperatively. Re-myelography revealed delayed contrast filling of the recurrent cyst; however, surgical revision did not lead to an improvement despite successful fenestration and collapse of the cyst revealed by postoperative imaging.

Microsurgical fenestration of sacral perineural cysts to the thecal sac is a surgical approach that has shown success in the treatment of lumbosacral pain, pseudoradicul symptoms, genital pain and urinary dysfunction associated with sacral perineural cysts. The analysis, however, shows that mainly
patients with singular large cysts benefit from this treatment.

2009

Over a seven year period 4000 patients underwent surgery for lumbar disk herniation. In three patients neurological symptoms were caused by large sacral perineurial cysts. Methods of choice for diagnosis of Tarlov cysts are lumbosacral magnetic resonance imaging and computerized tomography myelography. The majority of Tarlov cysts are asymptomatic. In case of large (> or = 1.5 cm) and symptomatic perineurial cyst, as in three patients reported in this article, microsurgical treatment was successful. Although rare, perineurial (Tarlov) cysts must be taken into consideration when approaching to patient with low back and radicular pain. Authors review the medical literature, pathological and pathophysiological features and treatment options of sacral perineurial cysts.

Case reports

2017

Iwamuro et al. present a case of multiple thoracolumbar perineural cysts, one of which was considered the cause of intermittent intercostal neuralgia with atypical findings on postmyelographic computed tomography seen as selective filling of contrast medium.

A 61-year-old woman presented with intermittent pain on her left chest wall with distribution of the pain corresponding to the T10 dermatome. Magnetic resonance imaging showed multiple thoracolumbar perineural cysts with the largest located at the left T10 nerve root. On postmyelographic computed tomography immediately after contrast medium injection, the largest cyst and another at left T9 showed selective filling of contrast medium, suggesting that inflow of cerebrospinal fluid to the cyst exceeded outflow. Three hours after the injection, the intensity of the cysts was similar to the intensity of the thecal sac, and by the next day, contrast enhancement was undetectable. The patient was treated with an intercostal nerve block at T10, and the pain subsided. However, after 9 months of observation, the neuralgia recurred, and the nerve block was repeated with good effect. There was no recurrence 22 months after the last nerve block.

They concluded that intermittent elevation of cerebrospinal fluid pressure in the cyst caused the neuralgia because of an imbalance between cerebrospinal fluid inflow and outflow, and repeated intercostal nerve blocks resolved the neuralgia. Our case demonstrates the mechanism of cyst expansion.

2009

A 28-year-old man presented with back pain and retrograde ejaculations resulting in infertility. After microsurgical excision of large perineurial cysts, back pain resolved, but semen quality showed only marginal improvement. Later, the couple successfully conceived by intrauterine insemination. To the best of our knowledge, this is the first reported case of Tarlov cyst associated with retrograde ejaculation and infertility.

Despite being mostly asymptomatic and an incidental finding, Tarlov cyst is an important clinical entity because of its tendency to increase in size with time. Tarlov cysts of the sacral and cauda equina region may be a rare underlying cause in otherwise unexplained retrograde ejaculations and infertility. Microsurgical excision may be a good option in a select group of patients.
Links

Tarlov Cyst Disease Foundation

http://www.tarlovculdfoundation.org/

Clinical Trials

NCT02595190


