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Electromyography and A Review of the Literature Provide Insights into the Role of Sacral Perineural Cysts in Unexplained Chronic Pelvic, Perineal and Leg Pain Syndromes

Hulens Mieke[1*], Bruyninckx Frans[2], Somers Alix[3], Stalmans Ingeborg[3], Peersman Benjamin[4], Vansant Greet[5], Ricky Rasschaert[6], De Mulder Peter[7] and Dankaerts Wim[1]

[1]Department of Rehabilitation Sciences, Faculty of Kinesiology and Rehabilitation Sciences, Musculoskeletal Rehabilitation Research Unit, University of Leuven, Belgium

[2]Clinical Electromyography Laboratory, University Hospitals, Leuven, Belgium

[3]Department of Neurosciences, Research group Ophthalmology, University of Leuven and Department of Ophthalmology, University Hospitals UZ Leuven, Herestraat 49, 3000 Leuven, Belgium Belgium

[4]Department of Radiology, Regional Hospital, Tienen, Belgium

[5]Department of Social and Primary Health Care, Public Health Nutrition, University of Leuven, Belgium

[6]Department of Neurosurgery, ZNA Middelheim Antwerpen, Belgium

[7]Department of Pain Therapy, Imelda Hospital, Bonheiden, Belgium

*Corresponding Author:
    Mieke Hulens
    Department of Rehabilitation Sciences
    Faculty of Kinesiology and Rehabilitation Sciences
    Musculoskeletal Rehabilitation Research Unit
    University of Leuven, Overwegstraat 14
    3051 Sint-Joris-Weert, Belgium
    Tel: +32-478-338003
    Fax: +32-16-470559
    E-mail: miekehulens@skynet.be

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Abstract

Objective: The clinical entity "Symptomatic Tarlov Cysts" is a highly under reported condition. We aimed to perform an electrophysiologic evaluation in patients with Tarlov cysts to determine whether the cysts create electrical abnormalities that could translate into clinical symptoms. The findings are correlated with the data currently available in the literature.

Methods: Thirty patients with unexplained pelvic, sacral, perineal and/or leg pain who harbored small and/or large Tarlov cysts were selected at an outpatient clinic for physical medicine in musculoskeletal disorders. An MRI of the lumbosacral spine of each patient was reviewed. An experienced physiatrist acquired information related to pain and paresthesia in addition to bladder, bowel and sphincter symptoms. An expert electrophysiologist performed nerve conduction and electromyography studies on the patient's legs and the pelvic floor.

A review of the case reports on Tarlov cysts was performed. The symptoms of the patients in the study were compared with the symptoms reported in reviews and case reports.

Results: In all cases, the presence of Tarlov cysts was associated with sensory neuron symptoms, such as pain and paresthesia, and with bladder, bowel, sexual, and/or sphincter complaints. In all cases, electromyography documented axonal damage in multiple lumbar and sacral nerve root myotomes.

Conclusion: Symptomatic Tarlov cysts clinically and electrophysiologically represent a progressive chronic cauda equine syndrome. In patients with intractable sacral, perineal, pelvic or leg pain, symptomatic Tarlov cysts should be included in the differential diagnosis.

Keywords

Introduction

Tarlov cysts (TCs), or perineural cysts, are spinal meningeal cysts that contain nerve root fibers. They originate from dilations of the nerve root sheath at the dorsal root ganglion and are most frequently identified in the sacrum [1,2]. In 1938, when Isadore Tarlov discovered sacral cysts, he first believed that they were benign incidental findings. However, he later reported that they could be a source of radicular pain [3,4]. Unfortunately, this first erroneous assumption perpetuated, resulting in a worldwide consensus regarding their clinical insignificance [5]. There are also few clinical diagnostic characteristics that can be used to link these cysts to specific symptoms. Hence, it is almost always concluded that a patients' pain originates from other coexisting conditions, primarily degenerative disorders that are observed on imaging [6]. Many physicians are therefore unaware that "sacral symptomatic Tarlov cysts" (STCs) are a well-defined clinical entity that includes debilitating pelvic, perineal and leg pain and urogenital and bowel disturbances. STCs are easily identified when a comprehensive history is obtained [2,5,7,8].

Because this pathology is systematically overlooked, several authors have suggested that the incidence of STCs is significantly underestimated [2,5,8-16].

The purpose of this paper is to report electrophysiological findings in patients with unexplained chronic pelvic, perineal, and leg pain syndromes who harbor lumbosacral TCs to determine whether TCs create electrophysiological abnormalities that could translate into clinical symptoms. We used nerve conduction studies (NCS) and needle-electromyography (EMG) to analyze the lumbosacral nerve root myotomes. To determine the pathogenic activities and mechanisms that may lead to the various symptoms of this debilitating disease, our findings from NCS/EMG were linked to the information available regarding STCs in the literature.

Methods

The electronic database of a physiatrist outpatient clinic for patients consulting for musculoskeletal pain was searched to identify patients with unexplained debilitating or refractory lower back, pelvic, perineal and/or leg pain. An MRI of the lumbosacral spine, the sacrum and/or the pelvis was reviewed to determine whether TCs were present. TCs with a minimal size of 5 mm (or 3 mm for those that were located on the smaller lower nerve roots (S3-S4)) were analyzed [16].

If TCs were detected, the patients were specifically questioned regarding pain and numbness in the leg, perineum, bladder, bowel or sphincter dysfunction, genital symptoms, neck pain and headaches [2]. The painful myotomes in the leg (L3 to S2) and perineum (S3-S4) were recorded. Myotomes S1 and S2 were considered together and defined as pain on the posterior side of the leg because patients usually reported a diffuse distribution.

The first 30 patients found to harbor TCs (27 women and 3 men) were selected for further investigation. NCS and needle EMG were performed by an expert senior neurophysiologist. The neurophysiologist was informed of the location of only the largest TC in each patient but was blinded to the MRI images.

The electrodiagnostic tests consisted of conduction studies performed on the sensory sural nerves (which contain fibers from nerve roots S1 and S2), the motor peroneal nerves and the S1 Hoffmann reflexes (the electrophysiological equivalent of the Achilles tendon reflex). The needle EMG included the L3 to S3-S4 myotomes: L3 (vastus medialis muscle); L4 (vastus lateralis muscle); L5 (extensor digitorum muscle); L4-L5 (tensor fascia lata muscle and tibials anterior muscle); S1 (gastrocnemius muscle medial head); S2 (tibial nerveinnervated intrinsic foot muscles) and the S3-S4 myotomes (external anal sphincter). The diagnosis of a S1 radiculopathy was based on abnormalities during needle EMG and not solely on the S1 Hoffmannreflex latency.

Additionally, the S3-S4 ano-anal reflex (the electrophysiological equivalent of the ano-cutaneous reflex) was analyzed. This reflex is meant to prevent fecal incontinence, and it therefore requires an intact reflex arc that consists of both afferent and efferent limbs. When using needle-EMG on the anal sphincter, only the efferent (motor) limbs were evaluated, whereas when analyzing the ano-anal reflex, the afferent (sensory) limbs were also assessed. This perineal reflex also indirectly provided information about the urethral sphincter [17].

During needle EMG, the presence of denervation potentials or the presence of at least 50% polyphasic Motor Unit Potentials was considered abnormal. Most muscles normally show only 5-25% polyphasic MUPs (e.g. about 5% in the biceps muscle and 25% in the extensor muscles of the upper and the lower limbs). The muscle with the highest percentage polyphasic MUPs in most individuals is the adductor longus muscle with sometimes up to 40% of polyphasic MUPs. Therefore, we used ≥ 50% polyphasic MUPs as the absolute cut off value, so as to be sure that we didn't overcall the findings.

In our study, patients with sacral TCs often complained about neck pain. A MRI of the cervicodorsal spine was therefore reviewed, if available. Because the headaches that were often reported in our patient group might have reflected an increase in pressure inside the cerebrospinal canal [2,18], a complete ophthalmologic examination that included optic disc biometry was performed by an expert ophthalmologist. The examination used optical coherence tomography (Spectralis OCT, Heidelberg) and static automated perimetry (Humphrey Field Analyzer, Zeiss or Octopus 300, Heig-Streit) to detect and grade possible papilledema.

To explore STC pathogenesis and identify the typical patterns of symptoms that are caused by STCs, the literature from 1955 through 2016 was searched to obtain relevant reviews and case reports about patients with TCs. The MESH terms Tarlov, perineural, sacral and/or meningeal cysts were used. To gain new insights, the results of the NCS/ EMG tests were correlated with the data obtained from the literature.

Results

The characteristics and symptoms of the patients are shown in **Table 1**. The ages of the patients ranged from 25-74 years old (mean age, 46.0 ± 11.8 y). The size of the largest TC in each patient varied from 3 mm to 36 mm (mean 7.9 ± 3.8 mm). Most cysts were located on the L5 to S4 nerve roots. The patients were symptomatic for between 8 months and 50 years (mean 11.6 ± 12.0 y).

	Cases in the literature	Percentage	Patients in the study

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Total number of cases	507		30
Females	379	74.8%	27
Males	125	24.7%	3
Mean age (y)	49.4 ± 14.0		46.0 ± 11.8
Duration of the symptoms (y)	4.4 ± 6.7		11.6 ± 12.0
Trauma	54	10.7%	5
Heavy lifting/straining history	7	1.4%	0
Onset during or right after pregnancy	6	1.2%	5
Lower back/sacral pain	146	28.8%	25
Buttock pain	46	9.1%	24
Coccygeal pain	52	10.3%	14
Perineal pain	32	6.3%	10
Dyspareunia/genital pain	65	12.8%	18
Lateral hip pain	8	1.6%	24
Leg pain	174	34.3%	25
Pain in the foot/feet	10	2.0%	24
Paresthesia, perineum	10	2.0%	14
Paresthesia, buttocks	0	0.0%	14
Paresthesia, leg(s)	23	4.5%	19
Paresthesia, foot/feet	13	2.6%	24
Foot drop	4	0.8%	1
Acute cauda equine syndrome	3	0.6%	0
Headaches	8	1.6%	22
Abdominal pain	8	1.6%	22

	Cases in the literature	Percentage	Patients in the study
Anal pain	6	1.2%	18
Bladder dysfunction	59	11.6%	23
Urinary incontinence	77	15.2%	18
Bowel dysfunction	27	5.3%	21
Fecal incontinence	17	3.4%	12
Sitting increases pain	17	3.4%	29
Standing increases pain	36	7.1%	26
Weakness, leg/feet	54	10.7%	19
Walking increases pain	29	5.7%	21
Valsalva increases pain	33	6.5%	9
Physical straining increases pain	3	0.6%	25
Restless legs	0	0.0%	11
Persistent genital arousal syndrome	13	2.6%	0
Significant pain, neck/arm(s)	Not reported	Not reported	23

Table 1: Characteristics and symptoms of patients in the case reports and this study.

The EMG results are listed in **Table 2**. NCS revealed sural nerve abnormalities in 5 (16.7%) of the patients and increased latency in the S1 Hoffmann reflexes in 6 (23.1%) of the patients. Of the 28 patients who underwent ano-anal reflex testing, 25 (89.3%) displayed increased latency on one or both sides. None of the patients showed motor peroneal nerve abnormalities.

	Number (percentage) of patients with EMG abnormalities in the nerve root myotomes	Number (percentage) of patients with NCS/EMG abnormalities in the myotomes corresponding to their dermatomal pain and/or paresthesia	Number (percentage) of EMG abnormalities corresponding to the presence of TCs on that nerve root	Number (Percentage) of cysts located bilaterally on that nerve root	Mean size (range) of the cysts on nerve root (mm)

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Nerve conduction studies					
Sural nerve (S1S2)	5/30(16.7%)	4/5(80%)			
S1 Hoffmann-reflex latency	7/30(23.3%)	7/7(100%)			
Ano-anal reflex	25/28(89.3%)	23/25(92%)			
Needle EMG					
L4	11/30(36.7%)	8/11(73.0%)	7/18(38.9%)	18(30.0%)	7.3(5-10)
L5	23/30(76.7%)	14/23(60.9%)	33/49(67.4%)	49(81.7%)	7.6(5-11)
S1	12/30(40.0%)	26/28(92.9%)	25/58(89.0%)	58(96.7%)	8.9(5-17)
S2	28/29(96.6%)	(both S1 and S2)	32/56(57.0%)	56(93.3%)	7.2(3-34)
S3-S4	21/28(75.0%)	17/21 (80.9%)	37/59(63.0%)	S3 37(61.7%)	8.7(3-35)
			S4 22(36.7%)	7.1(3-16)	

Table 2: Results of NCS and needle EMG studies showing the mean number of cysts and the mean size (range) of cysts on each nerve root (Reference values: sural nerve amplitude ≤ 5 µV and/or latency ≥ 3.5 ms at 14 cm [53], S1 Hoffmann reflexes ≤ 30 ms, ≤ 32 ms and ≤ 34 ms for body length ≤160 cm, ≤182 cm and ≤195 cm repectively or a left/right difference of 2.0 ms; ano-anal reflex latency ≤ 50 ms or left-right difference ≥ 5 ms, or amplitude of L-R difference ≥ 500 µV [57]).

Following needle EMG, abnormalities were identified in multiple lumbosacral nerve root myotomes in all of the patients, and these abnormalities were bilateral in 80% of the patients. In

25 (83%) patients, the pain was located in two or more dermatomes at the same time. Only 17 (57%) patients predominantly complained about pain in one leg. Ten (59%) of these 17 patients had the largest TC on the same side as the most severe pain. In 15 of the 17 (88%) patients, the most severe EMG abnormalities were found in the most painful leg. In all the patients, TC and/or nerve root dilations were found on all of the nerve roots of the painful dermatomes.

Three patients with symptomatic sacral TCs and EMG abnormalities in the sacral nerve root myotomes also had severe thoracic pain in a dermatomal pattern. Therefore, an EMG of the thoracic paraspinals was performed, and in these cases, EMG abnormalities were detected in the corresponding dermatomes. The MRIs showed nerve root dilations in the corresponding nerves, and the nerve root fibers were splayed out.

Additionally, 23 (76.6%) patients also complained about neck pain and/or pain in the arms. Their MRIs of the cervicodorsal spine were therefore revised. A previous MRI was available in 18 (60.0%) patients. In 15 (88.2%) of these patients, nerve root dilations were identified in the lower cervical and/or upper dorsal nerve roots (C7 to Th4). Fundoscopy and OCT showed no papilledema, and a visual field examination showed no abnormalities in any of the 30 patients.

Discussion

From the literature search, a total of 79 case reports and reviews describing 507 cases (236 women and 100 men) were identified, and the results of these studies were compared to those in our cohort. In the literature, only rare cases of large TCs that cause significant neurologic symptoms are generally reported because they are obvious and cannot be easily overlooked. The results of our study, in combination with a more thorough analysis of the case study-based literature and reviews, demonstrate that even small sacral TCs can cause debilitating sacroiliac, pelvic and genital pain, sexual dysfunction, bladder and bowel symptoms, and urinary and fecal incontinence.

Pathogenesis

Increased intraspinal pressure: TCs emerge as a result of fluid becoming trapped inside a nerve root. The work of Sun et al., who microscopically analysed resected TCs, increased our understanding of the mechanisms underlying cyst formation. The upright position of humans causes the physiological hydrostatic pressure in the terminal dural sac to be high. Under increased pressure, the lumbar and sacral nerve root sheaths, which contain the nerve root fibers that leave the terminal dural sac, may begin to enlarge. Eventually, for unknown reasons, fluid may become trapped between the endoneurium and the perineurium, and some of these cases may evolve into cyst-like structures: the so-called perineural cysts or TCs [19]. This expansion of the nerve root sleeve may be enhanced by congenital weakness of the connective tissue because large TCs have been found to occur more frequently in patients with hypermobility and connective tissue disorders, such as Marfan syndrome [2,20].

Sun et al. [19] compared the mechanisms involved in cyst initiation and enlargement with the formation of aneurysms in arteries. In aneurysms, years of increased pressure force a weakened vessel wall to enlarge. Similarly, nerve root dilation may require a long period of increased hydrostatic pressure in the spinal canal [2,19,21-24].

A ball-valve mechanism has been described in some original reports and reviews on TCs. This valve mechanism ensures that when CSF comes under increased hydrostatic pressure, it enters but does not leave the cyst. This allows pressure to build up inside the cyst. On CT myelography, cysts with a valve mechanism do not immediately fill up with contrast medium. Where the root passes through the foramen, the presence of a valve mechanism can be periperatively confirmed using the Valsalva maneuver [4,6,25,26].

The valve mechanism theory does not, however, cover all cases because several studies have reported evidence demonstrating that non-valved cysts can also cause symptoms [2,6]. Additionally, it is well established that aseptic meningitis can occur when treating STCs with aspiration and fibrin glue injection if the glue is injected into a non-valved cyst from which it can spread into the dural sac [12,27]. Potts et al. [28] proposed that in cases with communicating cysts, the pressure within the cysts should be equal to the pressure in the subarachnoid space.

The literature therefore indicates that TCs without a valve mechanism are capable of producing significant symptoms. However, the mechanisms underlying these symptoms remain unclear.

Using CT-myelography, we documented both valved and nonvalved STCs in one patient in our cohort (**Figure 1**). The MRI of this patient showed that there was a large cyst (36 mm) on nerve root S2 that caused significant bone erosion (**Figure 1A**). Smaller TCs were observed on the bilateral L5, bilateral S1, and right S2 nerve roots (sizes: 8 mm-14 mm) (**Figure 1B**). Neither of the S2 cysts immediately filled on CT myelography, and they were therefore categorized as valved cysts. In contrast, the smaller cysts that filled immediately were categorized as non-valved cysts (**Figure 1C**). This patient showed bilateral atrophy in the S2-supplied intrinsic foot muscles in addition to significant bilateral EMG abnormalities in these S2 myotomes, which contained the valved cysts. However, she also developed a partial left foot drop (extension strength 3/5) with significant EMG abnormalities in the left L5 nerve root-supplied myotomes, on which a smaller non-valved TC (8 mm) was located. This case demonstrated that larger valved and smaller nonvalved cysts (which are actually nerve root dilations) can be present in the same patient and can produce neurological symptoms as a result of axonal damage that can be demonstrated using EMG.
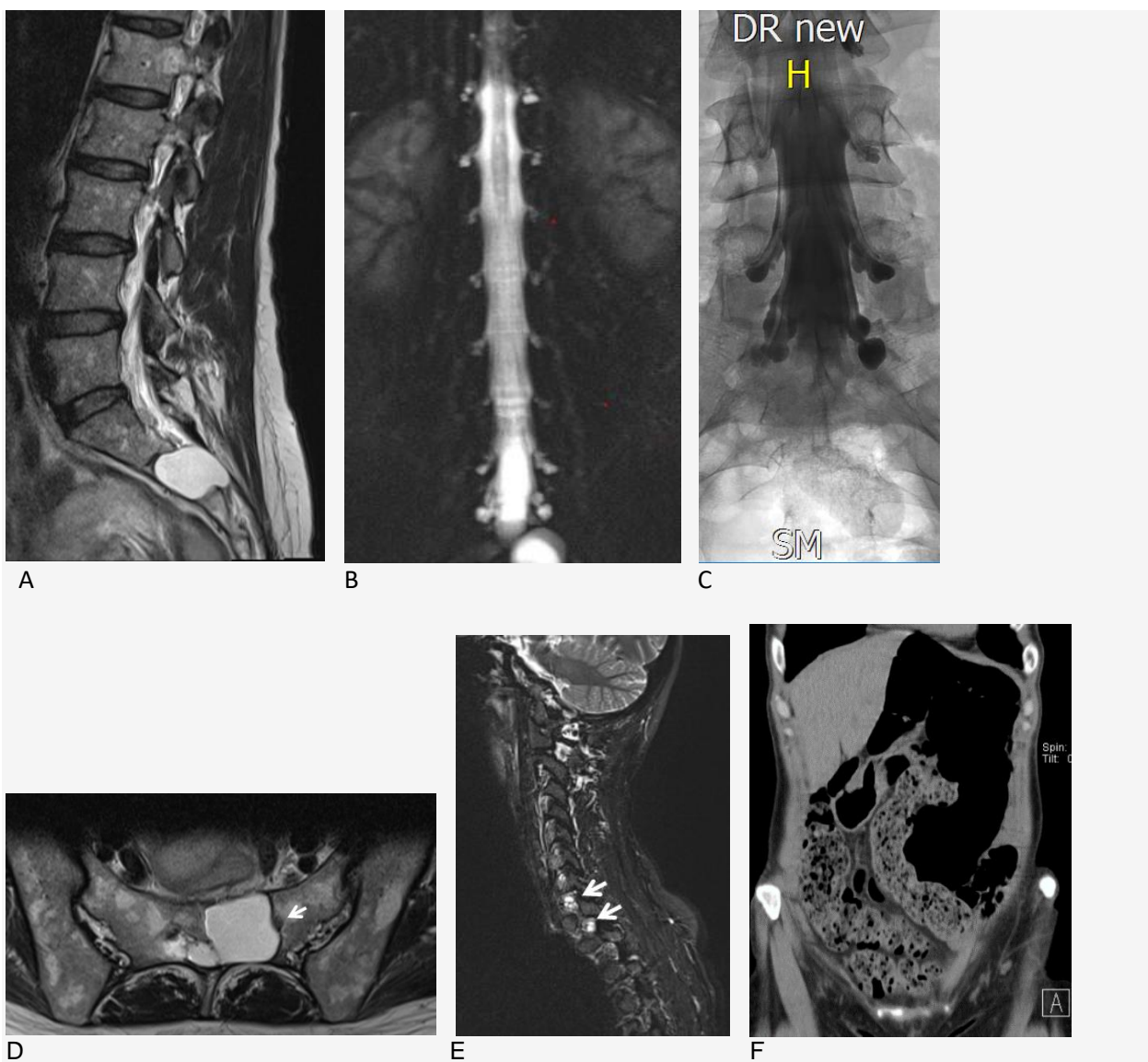

Figure 1: Six images obtained for a single patient who presented with a large STC. (A) Sagittal T2-weighted image of the largest cyst at the level of vertebra S2 showing that it eroded the entire left S2 vertebral body. (B) MRI myelography showing nerve root dilations and a large cyst on the left S2 nerve root. (C) CT myelography showing that the cysts on S2 did not immediately fill with contrast

medium (indicating a valved cyst), whereas the other bilateral nerve root dilations/cysts on nerve roots L5 and S1 filled immediately on both sides (indicating non-valved cysts). (D) Axial T2-weighted view of the large left and smaller right TCs at the level of the S2 vertebrae. The S2 nerve root is compressed against the bone (arrow). (E) Sagittal STIR image of the cervical spine in the same patient showing T1 and T2 nerve root dilations (arrows). (F) Coronal image of a CT scan of the abdomen showing a significantly dilated transverse and descending colon.

Consequently, we conclude that in non-valved cysts, a prolonged increase in hydrostatic pressure in the spinal canal is the crucial factor that contributes to the irritation of the nerve root fibers inside the nerve root sheath. We hypothesize that under a high level of intraspinal pressure, some of the nerve root sleeves become merely dilated, whereas others form true cysts. This mechanism is indeed similar to the mechanism that leads to the formation of a vascular aneurysm [19] because despite a prolonged increase in arterial pressure, only a minority of hypertensive patients develop an aneurysm.

Additional evidence has been reported to support the role of increased intraspinal pressure in patients with STCs. When performing spinal fluid taps, Feigenbaum and Henderson observed persistent increases in opening pressure in 1 out of 10 STCs. They also suggested that increased intracranial pressure may be associated with STCs [2]. Anatomical pathological evidence comes from studies that have analyzed resected nerves that were obtained from autopsies. Large TCs were found caudally on the lumbar and sacral roots, whereas smaller cysts were located on the thoracic nerve roots. These data suggest that in more cranial locations, intraspinal pressure may be elevated at least high enough to facilitate the formation of TCs [21].

.These data are in accordance with the findings reported in our study. Twenty-three patients with sacral TCs complained of significant neck and/or arm pain. A MRI of the cervical spine was available in 18 of the patients, and 15 of these patients also harbored multiple nerve root dilations or smaller TCs (<10 mm) in the cervicodorsal region. An example of this type of nerve root dilation that was observed in the patient who had large sacral TCs is shown in **Figure 1** (**Figure 1E**). Moreover, three patients with unilateral thoracic pain also demonstrated thoracic radiculopathy on needle EMG. They showed no obvious TCs, but the nerve fibers were splayed out inside the nerve sheath.

Several authors have described that patients may report experiencing more pain while in an upright position, such as walking, standing or sitting, or when performing Valsalva maneuvers. This pain is often relieved when the patient is lying down, indicating that it may be affected by the pressure inside the spinal canal [2,10,19,29]. In our study, 30% of the patients reported increased pain while performing Valsalva maneuvers, while 86.7% reported increased pain while standing and 83.3% reported increased pain while straining. In 66.7 % of the patients, this pain was immediately relieved when the patient lied down. However, the majority (86.6%) of the patients could not lie down for long periods of time because of local pain on pressure points (e.g., the sacrum and the greater trochanter). In women, the symptoms may be exacerbated during pregnancy [30]. Among our patients, 12 out of the 16 women who had been pregnant reported pelvic or lower back pain during pregnancy.

Additional evidence of increased intraspinal pressure indicates a higher risk of experiencing a dural leak while performing surgery for STCs [6,11,19,25,31,32]. This is because of the fragility of the tissues and the elevated pressure in the spinal canal [10]. Finally, a high frequency of headaches might also reflect increased pressure in the cerebrospinal canal [1,2,8,11,18,33].

Increased intracranial pressure: It is likely that there is a link between STCs and idiopathic intracranial hypertension (IICH) because a combination of symptoms, including headache with pain in the neck, the back or the limbs, has been reported in patients with benign IICH. This condition occurs primarily in young obese women and is associated with papilledema and visual problems. Bortoluzzi et al. [34] discovered that in these patients, the spinal root sleeves were markedly dilated. Additionally, several other authors have reported radiculopathy in patients with IICH [35-38]. Obeid et al. observed radiculopathy during a needle EMG of the paraspinal muscles in a patient with IICH [39].

In a study of 101 adults with IICH, Round and Keane reported observing neck stiffness in 31, paresthesia in 22, and lower back pain in 5 patients. They attributed all of these symptoms to spinal nerve root irritation because all of the symptoms resolved immediately following lumbar puncture [40]. In a recent prospective study by Wall et al. that included 165 patients with IICH, 84% of the patients reported headaches, and half of the patients (53%) had back pain, while 42% had neck pain and 19% had radicular pain [41].

Obesity is associated with increased intra-abdominal pressure, which secondarily increases cerebrospinal pressure to a level higher than that observed in lean patients. One of our patients who lost 20 kg of body weight over 3 years reported marked improvement of pelvic pain and in the intensity and the frequency of headaches. Of the cases reported in the literature, only 8 (1.6%) were reported to have headaches, and this is most likely because the link between STCs and headache remains unclear. The presence of headaches was therefore not investigated.

Thus, we propose that clinical entity "STCs" may essentially correspond to an "idiopathic lmoderate chronic cerebrospinal hypertension syndrome".

To investigate the above hypothesis, we screened our patients for papilledema. We did not observe papilledema in any of our patients. This is probably because the pressure inside the cerebrospinal canal in STC patients is likely to be substantially lower than the pressure in IICH. Further investigating this hypothesis would require invasive cerebrospinal pressure monitoring [42].

Size and multiplicity: Multiple TCs can originate as a result of the above-described increases in intraspinal pressure. This process has been confirmed in many studies in the literature. Smith [21], who microscopically examined nerve roots from 100 autopsies, found cystlike formations in 9 patients. They were all multiple, and most were symmetrical. The number of cysts per patient ranged from 5 to 13. While most of the cysts were macroscopic in size and readily visible at autopsy, several could only be seen in microscopic preparations. Similarly, in his autopsy studies, Tarlov observed predominantly multiple sacral cysts [4,25,26].

Langdown et al. identified multiple and small TCs in peroperative investigations of the majority of patients with lumbosacral complaints. Because they were small, the authors assumed that the patient's radicular pain was attributable to other lesions that were identified on MRI and that the small TCs were therefore clinically irrelevant [6]. However, it was not reported whether these patients suffered from perineal pain or urogenital and bowel problems.

In a retrospective study, Komisaruk and Lee reviewed the MRIs of 18 women with persistent genital arousal disorder (PGAD) and found TCs in 12 (66.7%). The cysts had sizes that ranged from 3 mm to 20 mm (mean 9.6 ± 5.1 mm). The authors concluded that smaller cysts usually produce sensory symptoms [16]. This conclusion was confirmed in a more recent study by Sun et al. in which a negative correlation was found between the size of the cysts and their multiplicity. They also noted that when cysts are small, the pressure inside may still become critical. Therefore, even very small cysts can be symptomatic, and the initial symptoms in such cases are pain and paresthesia, which can be severely debilitating [19,24,43].

These data are in accordance with the findings in our study, which showed that all of the affected patients had either multiple smaller cysts or clear nerve root dilations and EMG abnormalities in multiple nerve root myotomes (**Table 2**) in addition to bilateral pain or paresthesia. The EMG abnormalities corresponded more often to the painful dermatome than the radiologic findings (**Table 2**). According to Naderi, fluctuations in pain severity and a lack of correlation between radiologic findings and symptomatology are the main characteristics of non-operated Tarlov cysts [44]. This result is in accordance with the findings of our study. The pain was usually not in one dermatome, but in several dermatomes at the same time. Moreover, the most severe pain was not always on the side of the largest cyst [8]. To date, the reason for this lack of correlation is not clear.

These findings confirm that it is not the size of the cysts but rather the pressure inside the nerve roots that is responsible for these symptoms [6,19,45]. **Figure 2A** shows a MRI myelography that was obtained from a 64-year-old male in our cohort who suffered for almost 50 years from

unexplained debilitating sacral and leg pain. The needle EMG demonstrated bilateral radiculopathy in multiple lumbosacral myotomes.
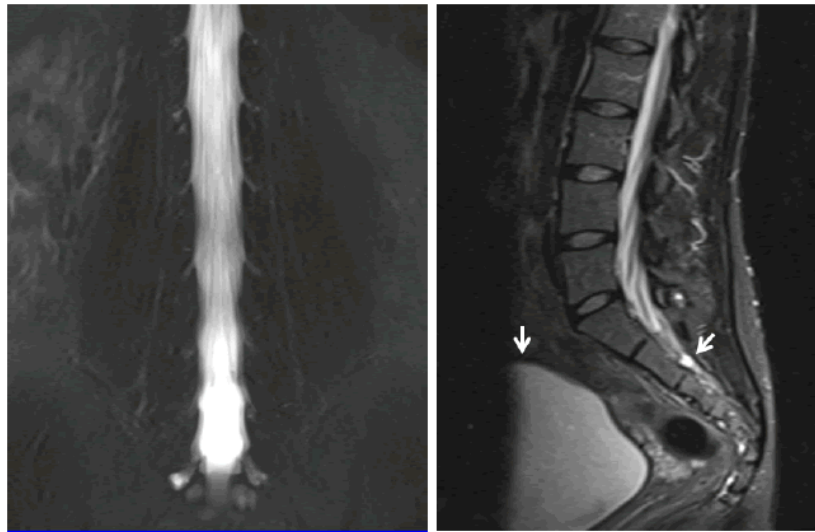

Figure 2: MRI myelography of 2 patients evaluated in this study. The sacral cysts are multiple, bilaterally localized and small. Both patients reported back pain, leg pain, and bladder and bowel symptoms and exhibited EMG abnormalities in multiple myotomes. (A) A 64-year-old male patient who suffered for almost 50 years from unexplained debilitating sacral and leg pain. (B) A 38-year-old female patient who suffered for 18 years from unexplained pelvic and leg pain and bladder issues. She harbored multiple small sacral cysts (one of them is visible under the right arrow) and had a significantly dilated atonic bladder (left arrow).

These results clearly highlight the clinical relevance of small TCs, despite the fact that it remains widely assumed that small, multiple TCs are clinically irrelevant.

Location

Sacral lesions are usually located on the S2 and S3 nerve roots [4,7,23,46]. In our study, all patients harbored TCs on either the S2 or the S3 roots or both, and all patients had EMG abnormalities in the S2 or S3-S4 myotomes or both.

Sun et al. reported that nerve root L5 is frequently involved [24]. In our study, TCs and EMG abnormalities were also frequently detected in the L5 myotomes. Although 96.7% of the patients had TCs on the S1 roots, only 40% had EMG abnormalities in the S1 myotomes; however, 81.7% had TCs on the L5 nerve and as much as 76.7% had EMG abnormalities in the L5 myotomes (**Table 2**). It is likely that the S1 root has more space to expand into the lateral recess than the L5 root has. This increase in available space may compensate for the increased pressure inside the root. It is worth noting that the L4 nerve root was also involved in 36.7% of our patients.

These percentages of EMG-abnormalities in the L4 to S1 myotomes would be high in the population being referred for radiculopathy. However, this is an extraordinarily elective group of patients with a longstanding history of unexplained pain. In these patients, the EMG abnormalities have developed over many years. As in other neuropathies, a longer compression of the nerve fibers (axons) in the nerve root(s) is associated with a higher probability that more severe and more widespread EMG abnormalities will occur.

Onset of symptoms

The onset of symptoms usually occurs in patients in their 4th or 5th decade [23]. However, symptoms can appear in teenagers [45,47].

In our cohort, the first symptoms developed at 14-19 years old in 3 patients, in their 20s in 11 patients, in their thirties in 7 patients, in their forties in 4 patients, in their fifties in 4 patients, and at the age of 64 in 1 patient. The mean age of onset (34.4 ± 12.9 y) in our patient group that had smaller cysts (on average) was approximately one decade younger than the age of onset in patients with large TCs in the case studies reported in the literature (46.0 ± 11.8 y). However, in our study, the two patients with the largest cyst sizes (34 mm and 36 mm) were also in their fifties when they first developed symptoms, in accordance with the average age of onset reported for large cysts in the literature.

Onset may occur when there is a history of trauma or heavy straining [30]. In our study, 16.7% of the patients reported that trauma was the trigger that elicited or aggravated their symptoms. The first symptoms are usually present long before an official diagnosis of STCs is established. In case reports in the literature, the time before diagnosis was 4.4 ± 6.7 y (range 1 week-25 y), whereas in our patient group, it was 11.6 ± 12 y (range 1 y-50 y). The apparently longer interval between initial symptoms and diagnosis in our study may have been because we inquired after less obvious sensory symptoms associated with smaller STCs.

Prevalence

Previous studies that analysed the MRIs of patients with lower back pain have reported a prevalence of 1.5%-4.6% [6,22,48,49]. However, because lower back and leg pain are almost always attributed to lumbar degenerative changes or disc problems, these MRIs did not routinely include axial or coronal sequences of the sacrum. Therefore, small TCs may have been overlooked. MRI studies of the lumbosacral spine that accounted for small cyst sizes have reported a prevalence of 9.1%-10.6 % [15,50].

When considering TCs that cause symptoms, it has been estimated that approximately one-fourth of such TCs are symptomatic at the time of discovery [6,22]. However, an unknown number of non-symptomatic cysts may also become symptomatic later in life [5].

TCs were found to be more prevalent in specific patient groups. For example, Tani et al. reviewed the pelvic MRIs of 102 women with otherwise unspecified gynecological problems and found that 10 (8.9%) had sacral meningeal cysts [46]. Likewise, Van de Kelft and Van Vyve [9] showed that of 17 patients with perineal pain, 13 had TCs (75%), while Komisaruk and Lee [16] found TCs in 12 out of 18 women (66.7%) with PGAD.

It has been reported that 61.2%-87% of patients who harbor TCs are women [5,6,30,48,50]. We found that this number was 74.8% in case reports and 90% in our EMG study. The reason for this female predominance is unknown, but there may be gender-specific differences in the structure of the dura mater or spinal nerve roots [2].

Symptoms and associated EMG abnormalities

The lumbosacral nerve roots supply the sensory and motor innervation of the lower extremities, the perineum and the sphincters in addition to the autonomic innervation of the colon and bladder. According to a review by Orendáčová et al., in fully developed cauda equine syndrome, multiple signs of sensory disorders may appear, including lower back pain, bilateral sciatica, saddle hypoesthesia or anesthesia, in addition to motor weakness of the lower extremities, bilateral impairment of the anal reflexes, rectal and bladder sphincter dysfunction and sexual impotence [51]. Reviews that discuss STCs have described most of these cauda equine symptoms [2,7,52]. However, in most case studies, only a subset of these symptoms were reported because not all authors inquire about bladder, bowel, or sexual functions [5]. For instance, lower back or sacral pain was reported in 38.2% of patients, whereas bowel symptoms were mentioned by only 8.1% and dyspareunia/genital pain by 13.0% of patients.

In STC patients, the intensity of the pain fluctuates, and the range of disability is variable. Patients with small cysts may have severely debilitating pain, whereas patients with large cysts may suffer only mild pain and be able to lead a normal life. In our cohort, three patients who were 33, 35 and 43 y old who had multiple bilateral cysts with a maximum size of 12 mm were housebound as a result of severe pain.

Pain, numbness, paresthesia, and hypoesthesia

In our study, all of the patients experienced lower back and/or sacral pain, and almost all of the patients reported buttock and/or leg pain. Pain and paresthesia in the feet was also common (80%) because the plantar dermatomes and myotomes are S2-innervated [5,53].

According to Orendacova, in cauda equine syndrome, the local pain is a deep, boring and aching pain, whereas radicular pain is usually a sharp stabbing pain that is often associated with paresthesia [51].

Because of their location near the dorsal root ganglion and pressure inside the nerve root, sensory nerve fibers are the first to be affected. TCs therefore primarily cause pain and sensory disturbances [24,52]. Hence, EMG may reveal abnormalities in conduction studies of the sensory sural nerve, which contains the fibers of the S1 and S2 nerve root [53]. A delay in the S1 Hoffmann reflex may be a sign of sensory radiculopathy. In 3 patients in our cohort, both sural nerve conduction and the Hoffmann reflexes were delayed, and in 4 patients, only the Hoffmann reflexes were delayed. The EMG abnormalities in patients with leg pain correspond more often to the respective painful dermatomes than to the radiologic findings. This result agrees with Naderi, who stated that the lack of correlation between the symptoms and the radiologic findings is one of the main characteristics of non-operated Tarlov cysts [44]. Additionally, 36.7% of our patients confirmed experiencing restless legs, and 63.3% reported weakness in the legs while walking or climbing stairs.

Neurogenic claudication

Almost all of the patients (80.0%) in our cohort reported to walk slower than before the onset of symptoms. If they attempted to walk faster, they experienced neurogenic pain.

Buttock pain

Buttock pain elicited by sitting is another typical characteristic of STCs. The inability to sit comfortably is socially debilitating and has been associated with avoidance behaviours. Additionally, employment requiring sitting is laborious, and these patients are therefore at increased risk of losing their job [2,7]. In our study, all but one patient confirmed that they experienced pain while sitting, 60.0% had to stop working, and 76.7% avoided social activities.

Perineal pain and numbness

The S2 to S5 roots supply sensory innervation to the perineum, the clitoris, and the penis. It has been suggested that female patients are unlikely to discuss their perineal symptoms with their physician, and this omission may contribute to the impression that TCs are clinically insignificant [5,11,53].

In the case studies reported in the literature, only 6.3% of the patients reported perineal pain, while 12.8% of the patients reported genital pain and/or dyspareunia. In our cohort, one-third of the patients reported genital pain, and two-thirds reported dyspareunia. It is important to note that these symptoms were specifically investigated in our study.

Coccygodynia

Nerve roots S4 and S5 supply the coccygeus and levator ani muscles and the sacrococcygeal joint. Case studies of patients with STCs commonly report coccygodynia (10%), and in our cohort, half of the patients (46.7%) reported this symptom.

Urinary and fecal incontinence

Nerve roots S2 to S4 supply sensory innervation to the internal urinary sphincter. Compressing these nerve roots results in urinary sphincter dysfunction and irritative voiding symptoms, such as frequency and incontinence. Additionally, compressing the motor fibers that innervate the anal sphincter and the sensory fibers from the perineum may lead to fecal incontinence [51]. Fecal and urinary incontinence can therefore originate from either a disturbance that leads to weakness in the sphincter or sensory loss in the rectum or urethra mucosa, which can lead to a delay in reflexive contraction.

Murphy et al. identified bladder sphincter impairments in 43.2% and bowel sphincter tone impairments in 28.6% of 213 patients. He reported that mild fecal incontinence was common in his patients [7]. This is in accordance with the findings in our study, in which 66.7% of the patients suffered from urinary stress incontinence and 40% suffered from fecal incontinence (30% discrete and 10% significant).

One patient in our study experienced severe fecal incontinence despite a normal sphincter tone and a normal needle EMG of the anal sphincter. She demonstrated a significant delayed ano-anal reflex (latency 87.4 ms), indicating that the sensory fibers of the reflex arc were affected.

Additionally, 60% of our patients experienced anal discomfort, anal pain or sphincter muscle cramps. All but one of the patients who reported anal pain also showed abnormalities on needle EMG of the anal sphincter.

Neurogenic bladder

Nerve roots S2 to S4 contain the parasympathetic fibers that stimulate the contraction of the detrusor muscle. Compression of these fibers results in urinary retention. The bladder therefore empties slowly, and a Valsalva maneuver is required to empty the bladder. Other associated symptoms include an inability to discern when the bladder is full, hesitation in emptying and an inability to completely empty the bladder (residu). Irritative symptoms include frequency, urgency and loss of bladder control [2,52,54]. Wallach and Schrot used urodynamic testing and documented urological disturbances in 32 women with STCs [13].

In our study, all but one patient experienced bladder dysfunction. All but one reported urinary frequency, 50% reported hesitation, 60% reported slow emptying, 53.3% reported urgency, and 56.7% reported a need to use a Valsalva maneuver to empty the bladder. The denervation of the detrusor muscle can eventually lead to a dilated atonic bladder, as demonstrated in **Figure 2C**.

Bowel dysfunction

Nerve roots S2 to S4 contain the parasympathetic fibers that innervate the descending colon and rectum. Some of these fibers also innervate the transverse and ascending colon [55]. In constipated patients suffering from cauda equine injury, difficulty with bowel evacuation and prolonged colonic transit are caused by subtle differences in phasic and tonic motor activity in the colon [51].

In STCs patients, bowel dysfunction presents as constipation, diarrhea, urgency and abdominal pain [2,52]. In our study, all but one patient reported symptoms related to bowel dysfunction. Of these, 73.3% reported constipation, and in 70% of these patients, constipation alternated with diarrhea. Additionally, 73.3% reported abdominal cramping or pain. **Figure 1F** shows an axial view of an abdominal CT scan of a patient with large STCs. This image shows a significantly dilated and atonic megacolon that was caused by a loss of contractility in the transverse and descending colon that was itself caused by denervation.

Sexual disturbances

The genital pain and/or loss of sensibility of the genital organs in STCs can cause dyspareunia and/or anorgasmia in women [56]. Sienaert reported finding unexplained vulvodynia in 3 women who harbored TCs [33]. Some women also experience hyperesthesia of the genitals or may develop PGAD. This is a painful and debilitating paresthetic condition that is probably underreported figure because patients are too embarrassed to declare it [2,16]. Feigenbaum Bperformed TC surgeries in 8 women with PGAD, resulting in a significant improvement in symptoms [56]. In a cohort of 157 STC patients, Marino reported that sexual issues were identified in 29% of the patients [30]. Of the 23 sexually active female patients in our study, 16 reported dyspareunia.

In men, prostate pain, unilateral or bilateral pain and/or numbness of the penis or the scrotum and impotence can occur [2,10,14,23]. Two of the male patients in our study reported genital pain. One of these patients reported impotence and left leg pain that was associated with pain in the left testicle and in the left side of the penis.

Foot drop and atrophy of the foot muscles

Motor neuron symptoms are less frequently observed in patients with STCs. As previously reported, in our cohort, a 54-year-old patient (**Figures 1A-F**) with multiple large STCs developed a foot extension strength deficit (3/5) as a result of compression of the left L5 nerve root in addition to atrophy of the intrinsic foot muscles on both sides that resulted from compression of the S2 nerve roots. This observation is in accordance with the findings reported by Sun et al. Large cysts are usually associated with a long symptom-free interval, and if they become symptomatic, the symptoms are more severe and more progressive. The authors used microscopy to identify these large cysts and characterized them as "paraneural cysts" [24]. **Figure 1D** shows an axial view of S2 cysts with the S2 nerve roots compressed against the bone. Large cysts may also compress the neighboring nerve roots and cause bone erosion.

In the multiple "perineural cysts" described by Sun et al., the nerve fibers are splayed out inside the cavity or along the wall. **Figure 3** shows a T2-weighted image of an MRI of a patient with left sciatica. The S1 nerve root is dilated, and the nerve root fibers are splayed out inside it. These small perineural cysts usually cause pain symptoms very early [24]. The longer pain-free intervals that have been reported in patients with paraneural cysts may result from weakened connective tissue, such as that observed in hypermobile patients, which could allow the root sleeves to expand to compensate for the pressure. Conversely, in smaller nerve root dilations or cysts, it is most likely that the tissues are less capable of expanding and that the pressure inside increases from the time of onset, thereby inducing pain very early.
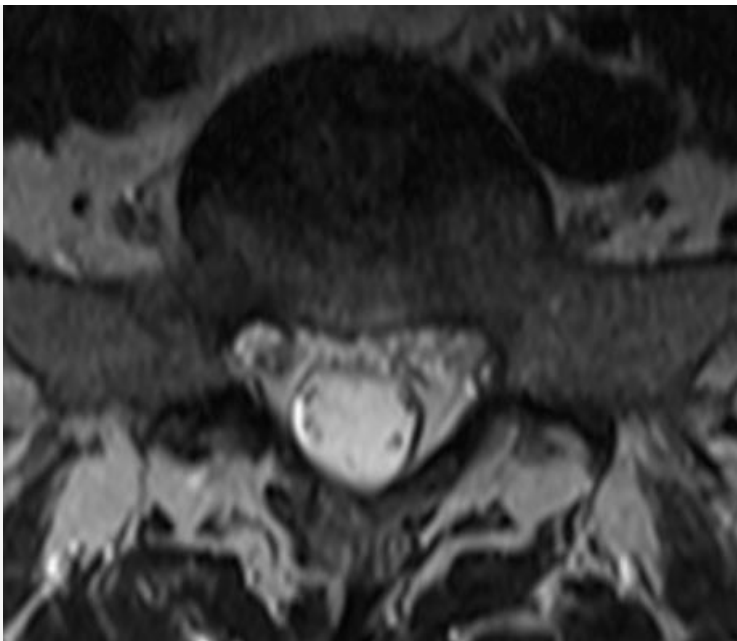

Figure 3: T2-weighted image of the MRI of a patient with left sciatica. The left S1 nerve root is dilated, and the nerve fibers are splayed out inside it.

Large cysts have an obvious clinical presentation and result in progressive motor dysfunction, and these are the cysts that are usually described in reviews and case reports. However, large cysts may be just the tip of the iceberg because nerve root dilations and smaller perineural cysts can also cause pain, paresthesia and bowel, bladder or sexual dysfunction that usually goes unnoticed and is therefore underdiagnosed. These cysts may therefore be a cause of chronic unexplained pain syndromes.

Table 3 lists and compares the most common characteristics of both large and small TCs. In the literature, we found 5 cases that were reported to exhibit foot drop resulting from a TC on the L5 nerve root. A study by Murphy included 213 patients with, on average, large cysts. They reported that absent S1 ankle reflexes were observed in 61% of the patients and weakness of the plantar flexion (S1) in 40.8% of the patients [7]. In the literature, no weakness or atrophy was reported in

the intrinsic foot muscles that are supplied by S2. Foot drop is a more obvious symptom than atrophy of the intrinsic foot muscles, and it is therefore easier to recognize. Hence, it is more likely to be reported.

Sacral Tarlov cysts	Large cysts	Nerve root dilations and
Microscopic (Sun [24]**)**	paraneural	perineural
Valved	usually	usually not
Size	>15 mm	3 mm-15 mm
Multiplicity	associated with smaller cysts and nerve root dilations	Always multiple cysts and nerve root dilations
Onset of symptoms	long pain-free interval	onset early in life
Mean age at onset symptoms	± 5th decade	± 2-3rd decade
Evolution	rapidly progressive	slowly progressive
Pain and paresthesia/numbness	usually	y
Neurological motor symptoms	usually	sometimes
Urinary and fecal incontinence	often	often
Cauda equine syndrome	acute	chronic
Prevalence	rare	common
In the literature	commonly reported	rarely reported
Clinical appearance	easily diagnosed	commonly overlooked

Table 3: Characteristics of small and large STCs.

Depression

Depression is a common comorbidity in STC patients [2,11,55]. Longstanding, unexplained, and debilitating neuropathic pain is often misunderstood not only by a patient's family, friends, and peers but also by their physicians. Additionally, patients with STCs often lose their employment and subsequently become housebound and socially isolated [2]. These patients' depressive symptoms may be attributed to the mistaken perception that the patients' pain, in addition to sexual disturbances, such as PGAD, are primarily caused by non-organic (psychological) factors.

Natural course

Only two papers reported cases of spontaneous remission in a total of two patients. In these studies, conservative measures, such as caudal block and physiotherapy, were applied [6,15]. However, spontaneous clinical improvement is likely to be only temporary. Xu et al. followed two patients who chose not to undergo surgical treatment for 4 years. During this follow up period, their cysts grew significantly and their symptoms worsened [32].

Chronic pain

Possibly, STCs may be the cause of several chronic pain syndromes, including some forms of chronic pelvic pain, chronic perineal pain, pudendal neuralgia, bladder pain, vulvodynia, nonspecific lower back pain and failed back surgery syndrome. However, TCs are never mentioned as a possible cause in more recent reviews of patients with these syndromes. Additionally, because the cervical and dorsal spine are frequently involved, it is likely that STCs or nerve root dilations account for at least a portion of the cases that involve widespread pain and fibromyalgia.

Conclusions

We revisited the MRIs of the lumbosacral spine in patients with longstanding unexplained debilitating pelvic, perineal and leg pain, and we found lumbosacral nerve root dilations and TCs. After integrating NCS/EMG into what is currently known in the literature, we determined that the clinical entity STCs may be a progressive neurological disorder that presents as a chronic cauda equine syndrome. This hypothesis is supported by the results of our study, in which the involvement of multiple lumbar and sacral nerve root myotomes was indicated in all cases. When axonal damage is documented in the S2 to S4 sacral nerve roots, it cannot be attributed to lumbar structural changes, which can simultaneously be observed on MRI.

Clinically, STC syndrome is a specific sensory neuron syndrome that presents with pain and paresthesia. This condition is therefore difficult to recognize. However, taking a comprehensive patient history that includes urologic, bowel and sexual symptoms in addition to urinary or fecal incontinence could guide the diagnosis toward STCs.

Hence, instead of looking for compressive structures around the nerves, we should instead focus more on the nerves themselves because the pressure may be coming from the inside [47]. The smaller size and multiplicity of STCs, in combination with the data obtained from both observations of EMG abnormalities and our search of the international literature, suggest that idiopathic elevations in cerebrospinal pressure could play a crucial role in the pathogenesis of STCs. Lowering this pressure might therefore be a target for therapeutic strategies.

This study has its limitations because it is a retrospective clinical report based on case studies without a control group. The symptoms and EMG findings should be compared to a similar group of patients without TCs on imaging. More prospective controlled studies are needed to further explore the relationship between the presence of TCs and patient symptoms.

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