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**Review Article** 

# Tarlov cyst. Theme update

#### Mikail Sallé MD1\*, Yurledys Jhohana Linares Benavides MD1, Francisco Félix Goyenechea Gutiérrez MD, PhD1,2

#### Thania Cristina Torres Santana MD<sup>1</sup>, Yasmany Fornaris Cedeño MD<sup>1</sup> and Bianchy González Pérez MD<sup>1</sup>

<sup>1</sup> Department of Neurosurgery, Institute of Neurology and Neurosurgery of Cuba.

<sup>2</sup> Head of the National Neurosurgery Group, Cuban Society of Neurology and Neurosurgery.

\*Corresponding Author: Yurledys Jhohana Linares Benavides, Department of Neurosurgery, Institute of Neurology and Neurosurgery of Cuba, Cuba.

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#### Abstract

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Tarlov cysts or perineural cysts are congenital conditions formed by ectasia of the perineural space of the spinal nerve roots. Despite the fact that there are isolated reports and small series of patients, they remain a little known topic in the medical field and there are few extensive reviews about it. Tarlov cysts are generally rare congenital conditions. Most are asymptomatic. In a group of patients the symptoms lead to invasive treatment. The ideal treatment continues to be a subject of debate.

Keywords: Tarlov cysts, extradural arachnoid cysts

## Introduction

Tarlov cysts or perineural cysts are congenital conditions formed by ectasia of the perineural space of the spinal nerve roots. These lesions have a predilection for the posterior root of the spinal nerve and are more frequent in the sacral region.(1). Its prevalence is 4.6 to 9% of the population and its estimated incidence is 1.4 to 4.65 in radiological studies.(2). Although most are asymptomatic, approximately 1% of cases may present variable symptoms. Despite the fact that there are isolated reports and small series of patients, they remain a little known topic in the medical field and there are few extensive reviews about it. That is why a bibliographic review of the updated literature was carried out with an emphasis on etiopathogenic theories, epidemiology, diagnosis and therapeutic options.

#### Ethiopathogenesis

These lesions have also been called extradural arachnoid cysts or periradicular cysts.(3).

They were first described as an incidental finding in 30 autopsies performed at the Montreal Neurological Institute by Isadore Tarlov in 1938, a pioneer in the field of Neurosurgery, and to whom their name is attributed. (2). Since then, less than 100 cases have been reported in the literature.(4). This author performed a histological examination of these findings and observed that the cystic lesions are located in the perineural space, between the endo and perineurium at the level of the junction of the posterior root and the posterior root ganglion, and concluded that these cysts can completely surround to the nerve root or invade it and compress the nerve fibers. He differentiated perineural cysts from meningeal diverticula based on the following findings: communication with the subarachnoid space, location, covering or wall, and the symptoms they present. He also distinguished perineural cysts from arachnoid processes of nerve roots that have no pathological significance.

They consist of small and occasionally large masses that originate at the expense of the innermost meninges: the pia mater and arachnoid mater. They are located around the nerves of the lumbosacral region and most at the S2 or S3 level (4). Its content is composed of cerebrospinal fluid (CSF) and communicates with the spinal subarachnoid space through a pedicle.

Different mechanisms have been described in their formation, and there are controversies about them. Some authors consider them to be extradural lesions acquired by the presence of inflammatory cells in their interior that suggests an inflammatory origin, or of hemosiderin that alludes to a traumatic origin or complication of anticoagulant treatment.(5). Some authors suggest that they are due to rupture of the venous drainage in the perineurium and epineurium secondary to hemosiderin after trauma.

For other authors, these are lesions of congenital origin due to arachnoid proliferation. However, reports of these injuries in pediatric patients are unusual.(3, 6). However, its eventual association with other congenital anomalies such as connective tissue disorders or dysragraphic disorders supports the theory of its congenital origin.

Recently, a growing number of authors suggest that a one-way valve mechanism occurs secondary to congenital anomalies or connective tissue diseases.

## **Clinical Manifestations**

Most of these lesions are diagnosed as imaging findings and are asymptomatic. However, as mentioned, they can cause variable symptoms of insidious or sudden onset, and of mild, moderate or severe intensity.(7).

The most frequent clinical manifestations include low back pain, lumbosacral pain that is aggravated by sitting, sciatica, lower limb paresthesia or hypoaesthesia in certain dermatomes. These manifestations are attributed to compression of the spinal nerves by the cyst and reduction of blood flow due to arterial and venous compression. They can also cause abdominal pain, especially presacral cysts.(8).

Less frequently, patients may present with sphincter alterations (fecal or bladder incontinence). Other less frequent manifestations include migraine headaches, visual disturbances, or vertigo. The manifestations are generally mild although in a group of patients they are unfortunately severe and disabling. Intense pain and neurological deficits, especially incontinence, affect their quality of life because no postural change provides relief. Neurological deficit can lead to severe disability(7).

# **Diagnostic Criteria**

These lesions are usually diagnosed incidentally during the course of a Magnetic Resonance Imaging (MRI).

Plain radiographs show areas of bone erosion of the lumbosacral structures. Although small cysts are difficult to visualize on Computed Tomography, bone erosions, thinning and even sacral pathological fractures can also be observed. Large cysts are usually visualized as single or clustered cystic masses with CSF density(6).

MRI is the study of choice to diagnose these lesions with high sensitivity and specificity. They are observed as solitary or multiple cystic masses with a signal sequence similar to CSF (hypointense in the T1-weighted sequence and hyperintense in the T2-weighted sequence)(9).

A useful study to determine the presence or not of communication with the CSF and thus establish the differential diagnosis with other similar cystic lesions consists of myelography. This test consists of injecting an iodinated contrast medium into the intra-spinal subarachnoid space and obtaining sequential radiographs at different times of administration: immediately, at two hours, at six hours, and at 24 hours. If it is a Tarlov cyst (communicating with the spinal subarachnoid space), the cysts fill with contrast. This technique allows to know with precision the morphology of the cyst, the type of communication and the filling speed of the cyst, data that are very useful in the diagnosis and prognosis(10).

Neurophysiological tests are indicated in these conditions. A decrease in the amplitude of the sensory action potential of the sural nerve is observed in the neuroconduction study, as well as a reduction in recruitment in the musculature of the affected myotome and a slowing of the reflex in the electromyography.(9).

Nabors et al. in 1988 they classified intra-spinal cysts into three categories: Type I: extradural cyst without nerve roots; Type IA: extradural meningeal cyst or extradural arachnoid cyst; Type IB: sacral meningocele; Type II: extradural cyst with nerve roots or Tarlov's perineural cyst and Type III: spinal cyst(10).

# Treatment

The treatment of Tarlov cysts is controversial due to the low frequency of symptomatic patients, which prevents the performance of studies with a high level of evidence and the recruitment of large series. To date, none of the procedures described has completely prevented relapses.(eleven). It is very important in these lesions to correlate the clinical manifestations of the patient with the location of the lesions, in order to determine if they are really symptomatic lesions and to avoid unnecessary invasive treatment. When the manifestations are predominantly pain and paresthesia, pharmacological treatment with opioid analgesics and antineuritics such as Carbamazepine, Gabapentin or Pregabalin may be indicated. Physical therapy has also been described(7).

There is no consensus on the criteria for invasive treatment. Invasive treatment modalities include bundle branch blocks or root blocks. Peripheral nerve stimulation and spinal cord stimulation have also been described.(12).

Within the surgical treatment options, different variants have been described. Among them, needle puncture with tomographic guidance with or without filling of the cystic cavity with steroids or fibrin.(2). This treatment option has the advantage of being less invasive than open surgery and rehabilitation is started early, but aseptic arachnoiditis and recurrence can occur. Even Zhang et al.(2) have reported percutaneous endoscopic puncture.

Laminectomy with excision of the cyst has also been described with the use of intraoperative neurophysiological monitoring. In these cases, fenestration or subtotal resection or en bloc resection has been described. Other authors recommend lumboperitoneal or cystoperitoneal shunt(2) and even the ligation of the neck of the cyst or the sacrifice of the nerve root from which it originates has been recommended. Rotation of the cyst walls has even been recommended to prevent their collection by valve mechanism.(3).

Microsurgery is superior to traditional surgery to separate and protect the nerve root, resect the cyst walls, and perform the nesting. However, the risk of CSF leak or cyst recurrence is high. Some authors recommend postoperative spinal drainage to avoid CSF fistula or to keep the patient in the prone position for several days(eleven).

Surgical resection of the coccyx (coccygectomy) is considered in case of failure of other treatments. However, it is not recommended by many neurosurgeons due to the poor response to treatment and the high percentage of complications, mainly infectious due to its proximity to the perianal region.(13).

## Forecast

It has been suggested that Tarlov cysts larger than 1.5 cm present a better response to surgical treatment. Spontaneous rupture of a Tarlov cyst and the disappearance of symptoms have also been reported.(13).

# Conclusion

Tarlov cysts are generally rare congenital conditions. Most are asymptomatic. In a group of patients the symptoms lead to invasive treatment. The ideal treatment continues to be a subject of debate.

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