

# Ventriculus Terminalis Cyst – Case Report And Literature Review

Zygmunt Siedlecki<sup>1\*</sup>, Karol Nowak<sup>1</sup>, Sebastian Grzyb<sup>2</sup>, Bartłomiej Gromadzki<sup>1</sup>, Magdalena Grzonkowska<sup>3</sup>, Mateusz Szostak<sup>2</sup>, Michał Wiciński<sup>2</sup>, and Maciej Śniegocki<sup>1</sup>

<sup>1</sup>Department of Neurosurgery, Neurotraumatology and Pediatric Neurosurgery, The Ludwik Rydygier Collegium Medicum in Bydgoszcz, The Nicolaus Copernicus University in Toruń

<sup>2</sup>Department of Clinical Pharmacology, The Ludwik Rydygier Collegium Medicum in Bydgoszcz, The Nicolaus Copernicus University in Toruń

<sup>3</sup>Department of Normal Anatomy, The Ludwik Rydygier Collegium Medicum in Bydgoszcz, Nicolaus Copernicus University in Toruń

**Abstract:** *We present a case report of a 48-year-old woman with a randomly diagnosed ventriculus terminalis cyst. The cyst was asymptomatic with radicular pain resulting from compression of the nerve roots. Due to the fact that such an anatomical variant is rare, we decided to describe this finding and to review the available literature. We revealed that although such cysts are asymptomatic, in the case of neurological symptoms they usually require surgical treatment.*

**Keywords:** *ventriculus terminalis, medullary cone, cyst, low back pain*

## Background

The ventriculus terminalis (VT) or terminal ventricle of Krause, also known as the 5<sup>th</sup> ventricle, is an ependymal-lined fusiform dilatation of the terminal central canal of the spinal cord, positioned at the transition from the tip of the conus medullaris to the origin of the filum terminale [1]. This differs from a filar cyst which is located within the filum terminale. That the filum contains nerve tissue was soon appreciated by many investigators [1]. The dilatation of the ependymal canal was called the ventriculus terminalis or the fifth ventricle by Krause in 1875 [1]. In turn, Saint-Remy et al. (1887) was unable to find a dilatation of the ependymal canal in adults comparable to that described by Krause [2]. The existence of VT was also postulated by Kernohan et al. (1924), who described its development, growth and clinical course [3]. Tarlov et al. (1938) described the filum terminale as a cylindrical tubule which becomes enlarged at the conus medullaris [4]. Sigal et al. (1991) noted that ventriculus terminalis was small cavity of the conus medullaris that forms during embryonic development as result of canalization and retrogressive differentiation [5].

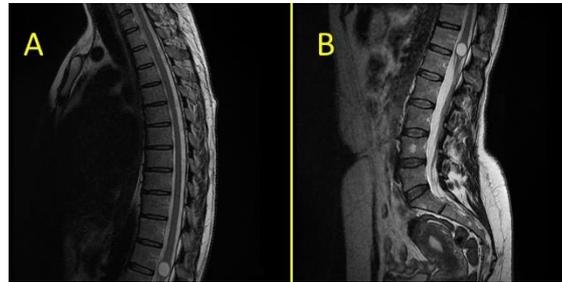
VT dilatation forming spherical cyst is found as incidental finding. It is defined as cystic dilation of the ventriculus terminalis (CDVT) or cystic lesion of the ventriculus terminalis (CLVT). Some authors and clinicians have postulated that CDVT sometimes can cause even severe neurological symptoms requiring surgery [5-15].

In clinical practice, asymptomatic ventriculus terminalis cysts are most often found as an incidental find during magnetic resonance imaging (MRI) diagnostics for spine pain. In most cases, these cysts are asymptomatic and the neurological symptoms result from compression of the nerve roots through the herniated disc. Nevertheless, some authors describe symptomatic ventriculus terminalis cyst and suggest surgical treatment.

## Case presentation

The authors present the case of a 48-year-old female patient complaining of lumbar spine pain, right-sided root pain of the sciatica type and pain of the right sacroiliac joint. Pain was periodically severe and required the use of analgetic drugs. In the neurological examination positive root symptoms were

found on the right side. Lasegue sign was positive at 45 degrees. This indicated irritation of the nerve root. The patient's root pain track corresponded to the L5 neuromer. We decided to perform both MRI of the lumbar and thoracic spine (Figure 1) to comprehensively assess the possible cause of the pain. MRI.



**Figure 1. MRI of spine: A. thoracic, B. lumbar with CDVT**

The experiment was approved by the Bioethics Committee of the Ludwik Rydygier Collegium Medicum in Bydgoszcz (KB 34/2020). A written informed consent was obtained from the patient for publication of this case report and any accompanying images. The case study was retrospectively based on MRI images and available medical data. MRI revealed an incidental find CDVT (Figure 2).



**Figure 2. Axial MRI with CDVT**

There was no evidence from the study that the cyst was causing these neurological symptoms. The cyst walls showed no contrast enhancement, while the cyst fluid looked like cerebrospinal fluid (CSF). An analgesic block of the sacroiliac joint was performed in the patient, and ventriculus terminalis cyst, as a rare anatomical find, was decided to be described in this manuscript.

## **Discussion**

The authors reviewed the literature on the CDVT and analyzed 11 references by various authors from 1991 to 2020. Below are the individual reports and their conclusions in chronological order. The presented authors described both asymptomatic CDVT, as in our case, as well as symptomatic, with guidelines for surgical treatment. On the basis of the available literature, we stated that most often CDVTs are asymptomatic, found by chance, have no clinical significance and do not require treatment.

Sigal et al. (1991) on the basis of MRI reviewed retrospectively four patients with evidence of congenital cystic dilatation of the ventriculus terminalis. According to Sigal, the dilated ventriculus terminalis appeared on MRI as a small ovoid cavity with regular margination and intracystic fluid resembled CSF [5]. Sigal noted that after contrast injection, no enhancement of the cyst and its wall was observed [5]. He emphasized that no enhancement differentiated congenital dilatation from cystic tumors, which occur more frequently in this anatomical region [5].

Matsubayashi et al. (1998) reported MRI findings in two patients with cystic dilatation of the ventriculus terminalis. In both cases the markedly dilated ventriculus terminalis was seen as a rounded cavity with regular margins, the content of which gave the same signal as cerebrospinal fluid with all MR pulse sequences [6]. No contrast enhancement was seen [6].

De Moura Batista et al. (2008) emphasized that ventriculus terminalis is a small cavity inside the conus medullaris that is formed during the embryonic development. He reviewed other references and proposed dividing the patients into 3 groups by clinical presentation as follows [7]:

- CLVT Type I, patients with nonspecific neurological symptoms or nonspecific complaints
- CLVT Type II, presence of focal neurological deficit
- CLVT Type III, presence of sphincter disturbances and bowel or bladder dysfunction

De Moura Batista noted that only 1 case in his study was handled conservatively and no improvement was documented [7]. Based on his research, it can be concluded that surgery is the treatment of choice for symptomatic CDVT.

Ciappetta et al. (2008) reported two cases of VT in elderly patients and data from a histological and ultrastructural study. Results of Ciappetta suggested that the site, age, and histological characteristics of the lesion allowed defining VT dilation as a nosological entity distinct from other cystic dilations of the conus medullaris [8].

Ganau et al. (2012) reported 13 new cases of CDVT treated in the Department of Neurosurgery at University Hospital in Verona, Italy. Treatment modalities and clinical and radiological outcomes, both early and at follow-up, were analyzed and compared with a preoperative classification of clinical presentation as established by de Moura Batista et al. (2008) [7]. According to Ganau, surgical treatment seemed to guarantee the resolution of CDVT. Dorsolumbar laminotomy, myelotomy, and cystic drainage were performed in 10 patients [9].

Ganau proposed a revision of the classification [9] of de Moura Batista [7]. According to Ganau, Type Ia with nonspecific symptoms with no relation to CDVT was best treated conservatively, thus Type Ib with rapid onset and invalidating unspecific complaints without comorbidities might benefit from surgical evacuation [9].

Dhillon et al. (2010) presented a 40-year-old man with conus medullaris syndrome secondary to CLVT and as other authors reviewed the relevant literature. The patient presented with 4 years of worsening right leg weakness, and examination showed bilateral fasciculations and hyporeflexia [10]. MRI showed a cystic lesion at Th11–Th12. In study of Dhillon, patient was managed with a Th11–Th12 laminectomy and fenestration of an intramedullary cyst [10].

Dhillon reviewed in 2010 that total of 32 patients have been described since 1968: 24 were female with a mean age of 46.6 years. It is similar epidemiological data to our case. In thus, all patients presented by Dhillon were symptomatically in contrast to our patient. Dhillon concluded that symptomatic patients were best managed surgically, although percutaneous aspiration was an emerging technique [10].

Kawanishi et al. (2016) presented symptomatic case of 66-year-old female suffered from disturbance while walking evolving for the past 2 years. MRI revealed a cystic dilatation of ventriculus terminalis [11]. In study of Kawanishi, patient experienced marked improvement of lower extremity strength by a fenestration of cyst and cyst-subarachnoid shunt [11].

Kawanishi noted, that treatment for asymptomatic patients (like in our case) was best conducted conservatively, whereas patients with focal neurological deficits was best handled surgically [11].

Lotfinia et al. (2018) described three case reports of symptomatic fifth ventricle cystic dilations. All of them were female and their mean age was 59 years [12]. Lotfinia treated them surgically and all three patients were improved based on clinical and imaging assessments. Cases described by Lotfinia suggested that surgical decompression was a safe and effective treatment in symptomatic patients and the neurosurgeons should be aware of such rare situations [12]. A complete list of differential diagnosis about other cystic dilations of the conus medullaris should be emphasized to select the correct clinical approach [12].

Zeinali et al. (2019) reported that 61 cases of CDVT have being reported to 2019. Symptomatic dilatation of VT in children has not been reported till now [13]. Zeinali presented a 5 year-old-boy with a sphincteric and walking disorder. The patient was assessed by clinical, electrophysiological and urodynamic investigations as well as MRI of the lumbar–sacral segment with and without gadolinium enhancement [13]. Lumbar MRI demonstrated the presence of a cystic lesion containing CSF, which did not enhance after gadolinium, compatible with the diagnosis of the ventriculus terminalis dilation [13]. The patient described by Zeinali underwent laminectomy and the cyst wall was fenestrated with a

midline myelotomy and in 6-month of follow-up, urinary problems and gait disturbance improved [13].

Domingo et al.(2020) described the case of a 54-year-old woman with a long-lasting history of left lower-extremity weakness and recent onset of bladder dysfunction. On further assessment, MRI of the thoracic and lumbar spine showed a Th10-L3 intramedullary cystic lesion [14]. Surgical fenestration of the cyst was rendered, but no biopsy was taken due to the highly functional tissue along the full extension of lesion [14]. Domingo concluded that cystic lesions of the ventriculus terminalis are rare entities with a common presentation of severe, progressive neurologic impairment [14]. Domingo described novel surgical techniques used to achieve successful fenestration of the cyst wall with remarkable neurologic symptom improvement [14].

Tuleasca et al. (2020) noted that asymptomatic patients with no radiological progression could benefit from regular clinical and radiological follow-up. Tuleasca also emphasized that symptomatic cases with posterior cysts could be offered laminectomy, myelotomy and marsupialization. In patients presenting with additional focal arachnoiditis, which might be subject of further cyst recurrence, a “T” shunt can be placed and further attached to piamater [15]. According to Tuleasca, physiological role of VT was currently undiscovered. The most common theory placed VT at the cross point between ending of Reissner fibers, which extend from subcomisural organs of the epi-thalamus to VT [15]. Tuleasca noted, that in persistent asymptomatic cases, similar to case described in our study, other abnormalities of craniospinal axis, such as spinal cord tethering, tumors should be excluded. In our study these abnormalities were excluded and CDVT was isolated and sporadic finding [15]. Tuleasca noted that, in theory, in other associated particular conditions, such as syringomyelia, dilatation was usually present in upper parts of spinal cord and is frequently associated with Chiari type I and myelomeningocele [15].

### Conclusion

CDVT is most often asymptomatic and is found incidentally on MRI as exemplified in this case report, however symptomatic CDVT usually requires surgery. Therefore, while CDVT is rare, it cannot be underestimated in clinical practice.

### Abbreviations

- CDVT - cystic dilation of the ventriculus terminalis
- CLVT - cystic lesion of the ventriculus terminalis
- CSF - cerebrospinal fluid
- MRI - magnetic resonance imaging
- VT - ventriculus terminalis

### Declaration

- Competing Interests: The authors declare that they have no conflict of interest.

### Acknowledgement

The authors thank technicians and doctors of the radiology department for professional and perfect imaging of the images analyzed in this case report. The authors thank physiotherapists and everyone involved in the patient's diagnosis and treatment.

### References

1. Krause, W. (1875). Der ventriculus terminalis des Rückenmarks. *Archiv für mikroskopische Anatomie*, 11(1), 216-230.
2. Saint Remy, G. C. A. (1887). *Recherches sur la portion terminale du canal de l'épendyme chez les vertébrés* (Doctoral dissertation, Impr. Nancéienne).
3. Kernohan, J. W. (1924). The ventriculus terminalis: its growth and development. *Journal of Comparative Neurology*, 38(1), 107-125.

4. Tarlov, I. M. (1938). Structure of the filum terminale. *Archives of Neurology & Psychiatry*, 40(1), 1-17.
5. Sigal, R., Denys, A., Halimi, P., Shapeero, L., Doyon, D., & Boudghene, F. (1991). Ventriculus terminalis of the conus medullaris: MR imaging in four patients with congenital dilatation. *American journal of neuroradiology*, 12(4), 733-737.
6. Matsubayashi, R., Uchino, A., Kato, A., Kudo, S., Sakai, S., & Murata, S. (1998). Cystic dilatation of ventriculus terminalis in adults: MRI. *Neuroradiology*, 40(1), 45-47.
7. de Moura Batista, L., Acioly, M. A., Carvalho, C. H., Ebner, F. H., & Tatagiba, M. (2008). Cystic lesion of the ventriculus terminalis: proposal for a new clinical classification: Case report. *Journal of Neurosurgery: Spine*, 8(2), 163-168.
8. Ciappetta, P., D'urso, P. I., Luzzi, S., Ingravallo, G., Cimmino, A., & Resta, L. (2008). Cystic dilation of the ventriculus terminalis in adults: Report of 2 cases. *Journal of Neurosurgery: Spine*, 8(1), 92-99.
9. Dhillon, R. S., McKelvie, P. A., Wang, Y. Y., Han, T., & Murphy, M. (2010). Cystic lesion of the ventriculus terminalis in an adult. *Journal of Clinical Neuroscience*, 17(12), 1601-1603.
10. Ganau, M., Talacchi, A., Cecchi, P. C., Ghimenton, C., Gerosa, M., & Faccioli, F. (2012). Cystic dilation of the ventriculus terminalis. *Journal of Neurosurgery: Spine*, 17(1), 86-92.
11. Kawanishi, M., Tanaka, H., Yokoyama, K., & Yamada, M. (2016). Cystic dilation of the ventriculus terminalis. *Journal of neurosciences in rural practice*, 7(04), 581-583.
12. Lotfinia, I., & Mahdkhah, A. (2018). The cystic dilation of ventriculus terminalis with neurological symptoms: three case reports and a literature review. *The journal of spinal cord medicine*, 41(6), 741-747.
13. Zeinali, M., Safari, H., Rasras, S., Bahrami, R., Arjipour, M., & Ostadrahimi, N. (2019). Cystic dilation of a ventriculus terminalis. Case report and review of the literature. *British journal of neurosurgery*, 33(3), 294-298.
14. Domingo, R. A., Bohnen, A. M., Middlebrooks, E. H., Quinones-Hinojosa, A., & Abode-Iyamah, K. (2020). T10-L3 Cystic Lesion of the Ventriculus Terminalis Presenting as Conus Medullaris Syndrome. *World neurosurgery*, 136, 146-149.
15. Tuleasca, C., Ducos, Y., Knafo, S., Levivier, M., Aghakhani, N., & Parker, F. (2020). Surgery for cyst dilatation of a ventriculus terminalis in adults: keep it simple!. *Neuro-chirurgie*.